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UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, DC 20549

Form 10-K

(Mark One)

☒ **ANNUAL REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934**
For the fiscal year ended December 31, 2018

or

☐ **TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934**
For the transition period from to

Commission file number: 001-35420

ChemoCentryx, Inc.

(Exact Name of Registrant as Specified in its Charter)

Delaware	**94-3254365**
(State or Other Jurisdiction of Incorporation or Organization)	**(I.R.S. Employer Identification No.)**
850 Maude Avenue Mountain View, California	**94043**
(Address of Principal Executive Offices)	**(Zip Code)**

(650) 210-2900
(Registrant's Telephone Number, Including Area Code)

Securities registered pursuant to Section 12(b) of the Act:

Title of Each Class	**Name of Each Exchange on Which Registered**
Common Stock, par value $0.001 per share	The Nasdaq Stock Market LLC

Securities registered pursuant to Section 12(g) of the Act:
None

Indicate by check mark if the registrant is a well-known seasoned issuer, as defined in Rule 405 of the Securities Act. Yes ☐ No ☒

Indicate by check mark if the registrant is not required to file reports pursuant to Section 13 or 15(d) of the Act. Yes ☐ No ☒

Indicate by check mark whether the registrant: (1) has filed all reports required to be filed by Section 13 or 15(d) of the Securities Exchange Act of 1934 during the preceding 12 months (or for such shorter period that the registrant was required to file such reports), and (2) has been subject to such filing requirements for the past 90 days. Yes ☒ No ☐

Indicate by check mark whether the registrant has submitted electronically every Interactive Data File required to be submitted pursuant to Rule 405 of Regulation S-T (§232.405 of this chapter) during the preceding 12 months (or for such shorter period that the registrant was required to submit such files). ☒ Yes ☐ No

Indicate by check mark if disclosure of delinquent filers pursuant to Item 405 of Regulation S-K is not contained herein, and will not be contained, to the best of the registrant's knowledge, in definitive proxy or information statements incorporated by reference in Part III of this Form 10-K or any amendment to this Form 10-K. ☒

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, a smaller reporting company, or emerging growth company. See definitions of "large accelerated filer", "accelerated filer", "smaller reporting company", and "emerging growth company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer ☐	Accelerated filer ☒
Non-accelerated filer ☐	Smaller reporting company ☐
	Emerging growth company ☐

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13 (a) of the Exchange Act. ☐

Indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Securities Exchange Act of 1934). Yes ☐ No ☒

The aggregate market value of the registrant's common stock held by non-affiliates of the registrant as of the last business day of the registrant's most recently completed second fiscal quarter was approximately $266.3 million, based on the closing price of the registrant's common stock on the Nasdaq Global Select Market of $13.17 per share.

The number of outstanding shares of the registrant's common stock, par value $0.001 per share, as of February 28, 2019 was 52,569,256.

DOCUMENTS INCORPORATED BY REFERENCE

Portions of the registrant's definitive proxy statement to be filed with the Securities and Exchange Commission pursuant to Regulation 14A in connection with the registrant's 2019 Annual Meeting of Stockholders, which will be filed subsequent to the date hereof, are incorporated by reference into Part III of this Annual Report on Form 10-K. Such proxy statement will be filed with the Securities and Exchange Commission not later than 120 days following the end of the registrant's fiscal year ended December 31, 2018.

CHEMOCENTRYX, INC.

FORM 10-K — ANNUAL REPORT

For the Fiscal Year Ended December 31, 2018

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PART I

Forward-Looking Statements and Market Data

This Annual Report on Form 10-K contains forward-looking statements that involve risks and uncertainties. All statements other than statements of historical facts contained in this Annual Report on Form 10-K are forward-looking statements. In some cases, you can identify forward-looking statements by terminology such as "may," "could," "will," "would," "should," "expect," "plan," "aim," "anticipate," "believe," "estimate," "intend," "predict," "seek," "contemplate," "potential" or "continue" or the negative of these terms or other comparable terminology. These forward-looking statements include, but are not limited to, statements about:

- the initiation, timing, progress and results of our preclinical studies and clinical trials, and our research and development programs;
- our ability to advance drug candidates into, and successfully complete, clinical trials;
- the commercialization of our drug candidates;
- the implementation of our business model, strategic plans for our business, drug candidates and technology;
- the scope of protection we are able to establish and maintain for intellectual property rights covering our drug candidates and technology;
- estimates of our expenses, future revenues, capital requirements and our needs for additional financing;
- the timing or likelihood of regulatory filings and approvals;
- our ability to maintain and establish collaborations or obtain additional government grant funding;
- our financial performance; and
- developments relating to our competitors and our industry.

These statements relate to future events or to our future financial performance and involve known and unknown risks, uncertainties and other factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by these forward-looking statements. Factors that may cause actual results to differ materially from current expectations include, among other things, those listed under "Item 1A. Risk Factors" and elsewhere in this Annual Report on Form 10-K.

Any forward-looking statement in this Annual Report on Form 10-K reflects our current views with respect to future events and is subject to these and other risks, uncertainties and assumptions relating to our operations, results of operations, industry and future growth. Given these uncertainties, you should not place undue reliance on these forward-looking statements. For all forward-looking statements, we claim the protection of the safe harbor for forward-looking statements contained in the Private Securities Litigation Reform Act of 1995. Except as required by law, we assume no obligation to update or revise these forward-looking statements for any reason, even if new information becomes available in the future.

This Annual Report on Form 10-K also contains estimates, projections and other information concerning our industry, our business, and the markets for certain drugs, including data regarding the estimated size of those markets, their projected growth rates, the incidence of certain medical conditions, statements that certain drugs, classes of drugs or dosages are the most widely prescribed in the United States or other markets, the perceptions and preferences of patients and physicians regarding certain therapies and other prescription, prescriber and patient data, as well as data regarding market research, estimates and forecasts prepared by our management. Information that is based on estimates, forecasts, projections, market research or similar methodologies is inherently subject to uncertainties and actual events or circumstances may differ materially from events and

circumstances reflected in this information. Unless otherwise expressly stated, we obtained this industry, business, market and other data from reports, research surveys, studies and similar data prepared by market research firms and other third parties, industry, medical and general publications, government data and similar sources. In particular, unless otherwise specified, all prescription, prescriber and patient data in this Annual Report on Form 10-K is from Datamonitor or Global Data. In some cases, we do not expressly refer to the sources from which this data is derived. In that regard, when we refer to one or more sources of this type of data in any paragraph, you should assume that other data of this type appearing in the same paragraph is derived from the same sources, unless otherwise expressly stated or the context otherwise requires.

ChemoCentryx®, the ChemoCentryx logo, Traficet™ and Traficet-EN™ are our trademarks in the United States, the European Community, Australia and Japan. EnabaLink® and RAM® are our trademarks in the United States. Each of the other trademarks, trade names or service marks appearing in this Annual Report on Form 10-K belongs to its respective holder.

Unless the context requires otherwise, in this Annual Report on Form 10-K the terms “ChemoCentryx,” “we,” “us” and “our” refer to ChemoCentryx, Inc., a Delaware corporation, and our subsidiaries taken as a whole unless otherwise noted.

Item 1. Business.

Overview

ChemoCentryx is a biopharmaceutical company developing new medications targeted at inflammatory disorders, autoimmune diseases and cancer. Each of our drug candidates is designed to selectively block a specific chemoattractant receptor, leaving the rest of the immune system intact. Our drug candidates are small molecules, which are orally administered, and, if approved, could address unmet medical needs, including improved efficacy, and offer significant quality of life benefits, since patients swallow a capsule or pill instead of having to visit a clinic for an infusion or undergo an injection.

In 2016, we executed on our strategy to form an alliance with a partner that could provide upfront fees and milestone payments to support the clinical development of our two leading drug candidates, avacopan and CCX140, to registration and pay us royalties upon sales in international markets, while we develop our own commercial infrastructure to sell directly in the United States.

To help communicate the breadth of our drug discovery platform, we have segmented our pipeline into early stage and late stage drug candidates.

Late Stage Drug Candidates

We have chosen to focus initially on orphan indications, where drug candidates tend to enjoy a faster path to market and better reimbursement. Our leading drug candidates address areas of clear unmet need, where the current standard of care, or SOC, is insufficient to halt progression of the disease and/or where today's treatment options come with serious side effects, such as those which accompany the prolonged use of steroids:

Avacopan (CCX168) — Inhibition of Complement-Mediated Pathways in Orphan Diseases

Avacopan (formerly CCX168) is an orally-administered complement inhibitor targeting the C5a receptor, or C5aR, and is being developed for orphan diseases, including (i) anti-neutrophil cytoplasmic auto-antibody associated vasculitis, or AAV, a devastating autoimmune disease that damages blood vessels and can lead to kidney failure, pulmonary failure and damage to other tissues; (ii) complement 3 glomerulopathy, or C3G, a debilitating disease that can lead to kidney failure; and (iii) moderate and severe hidradenitis suppurativa, or HS, a chronic, inflammatory, debilitating skin disease characterized by recurrent, painful, nodules and abscesses, ultimately leading to the formation of draining fistulas (also known as sinus tracts) as well as scarring.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV and C3G and by the European Medicines Agency, or EMA, for the treatment of C3G and microscopic polyangiitis and granulomatosis with polyangiitis, both forms of AAV. Additionally, avacopan has been granted PRIority MEdicines, or PRIME, designation from the EMA, to expedite its clinical development, and to potentially accelerate its marketing authorization.

Following completion of two Phase II clinical trials in patients with AAV, in which avacopan was well-tolerated and provided effective steroid-free control of the disease, we launched the Phase III ADVOCATE trial in December 2016. The FDA and the EMA concurred with the design of the study. ADVOCATE is a randomized, double-blind two-arm study which enrolled over 300 patients at over 200 sites in the United States, Canada, Europe, Australia, New Zealand and Japan. Patient enrollment of the Phase III ADVOCATE trial was completed in July 2018 and we expect to report topline data from this trial in the fourth quarter of 2019. Additionally, we launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G for which we aim to complete enrollment in 2019 and initiated a large placebo-controlled Phase IIb clinical trial, the AURORA trial, for the treatment of patients with moderate to severe HS in the fourth quarter of 2018.

CCX140 — Chronic and Orphan Kidney Diseases

CCX140, an orally-administered inhibitor of the chemokine receptor known as CCR2, has been in development for diabetic nephropathy, or DN, a form of chronic kidney disease, or CKD, and is now being developed for focal segmental glomerulosclerosis, or FSGS, a rare renal disease characterized by progressive proteinuria, excess protein in the urine, and impaired renal function. CCX140 has been granted orphan drug designation by the FDA for the treatment of FSGS.

A global Phase II clinical trial of CCX140 in patients with DN met its primary endpoint by demonstrating that CCX140 given orally once-daily added to a SOC renin-angiotensin-aldosterone system inhibitor treatment resulted in a statistically significant reduction in proteinuria, beyond that achieved with SOC alone, with the most pronounced effect shown in the patients with highest levels of proteinuria. Based on the safety and efficacy data related to reduction in proteinuria observed in the Phase II trial in DN, we launched two Phase IIb clinical trials, the LUMINA trials, of CCX140 for the treatment of primary FSGS, with and without nephrotic syndrome, for which there are currently no FDA-approved treatments.

Kidney Health Alliance with Vifor

In May 2016, we announced a partnership, which we refer to as the Avacopan Agreement, with Vifor (International) Ltd., and/or its affiliates, or collectively, Vifor, a European-based world leader specializing in kidney disease. While under this agreement we retained all rights to the United States and China, we granted Vifor exclusive commercialization rights to avacopan in Europe and certain other international markets. In December 2016, we entered into an additional agreement with Vifor, which we refer to as the CCX140 Agreement, relating to CCX140, our other late stage drug candidate. Under the CCX140 Agreement, we again retained all rights to the United States and China and we granted Vifor exclusive worldwide commercialization rights outside of the United States and China. In February 2017, we announced a further agreement with Vifor that harmonized the geographic commercialization rights underlying the agreements for both drug candidates, which we refer to as the Avacopan Amendment. In June 2018, we entered into additional agreements with Vifor to further expand Vifor's exclusive commercialization rights to include China under the Avacopan Agreement, or the Avacopan Letter Agreement, and the CCX140 Agreement, or the CCX140 Letter Agreement.

We have secured $215 million in upfront cash and milestone payments pursuant to our agreements with Vifor and are eligible for additional substantial milestone payments. Through our alliance, we maintain the commercialization rights to avacopan and CCX140 in the United States, and also retain control of the clinical development programs for orphan renal disease. Vifor gained the exclusive commercialization rights for all other international markets, and is obligated to pay us tiered royalties, with rates ranging from ten to the mid-twenties, on potential net sales.

At a future time defined in the CCX140 Agreement, Vifor has an option to solely develop and commercialize CCX140 in more prevalent forms of CKD. Should Vifor later exercise the CKD option, we would receive co-promotion rights for CKD in the United States, and we estimate that the clinical development and registration process for CKD would end at approximately the same time as Orphan Drug exclusivity.

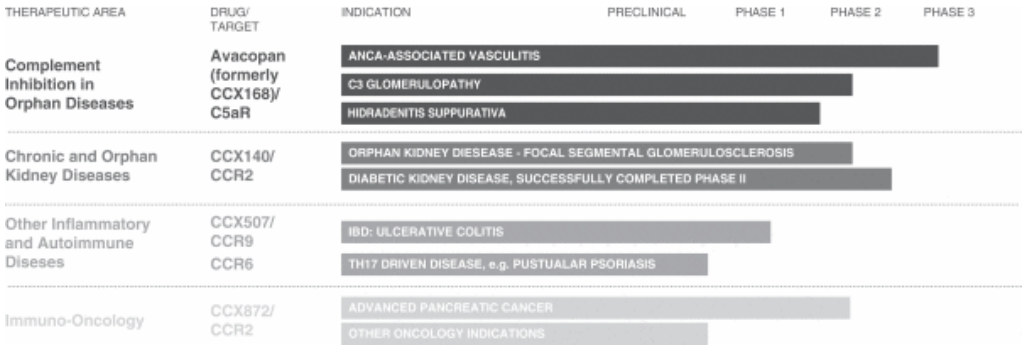
In October 2018, Vifor acquired 7,343,492 shares of our common stock from Glaxo Group Limited, bringing their aggregate holdings of our common stock to 10,676,825 as of December 31, 2018.

Early Stage Drug Candidates

While we have focused initially on orphan indications, our target-specific and selective approach designed to stop the spread of inflammatory disease-inducing cells shows promise in other disease areas. Over time we plan to bring forward drug candidates to treat a range of inflammatory and autoimmune disorders, as well as cancer, where our drug candidate CCX872 has shown promise in a Phase Ib trial for advanced pancreatic cancer. We expect that our ability to do so will grow as we increase our scale and to the extent that we start to earn revenues and royalties from the commercialization of our late stage kidney disease franchise.

Our Drug Candidate Pipeline

Broad Pipeline from Novel Discovery Platform



Late Stage Drug Candidates

Avacopan (CCX168) — Inhibition of Complement-Mediated Pathways in Orphan Diseases

In our complement inhibition orphan disease program, our lead drug candidate is avacopan.

Avacopan is a small molecule that selectively blocks the chemoattractant receptor known as C5aR, and is being developed for inflammatory and autoimmune diseases. Avacopan inhibits the activity of complement C5a, a component of the complement system and the natural ligand for C5aR. The complement system is a group of proteins that work together to regulate aspects of host defense against bacteria and viruses, trigger inflammation, and remove debris from cells and tissues. The complement system must be carefully regulated so it targets only unwanted materials and does not attack the body's healthy cells. In certain autoimmune diseases (including those in which we are engaged in clinical trials), components of the complement system have become dysregulated.

The FDA has granted avacopan orphan-drug designation for AAV and C3G. The European Commission has granted orphan medicinal product designation for avacopan for the treatment of two forms of AAV: microscopic polyangiitis and granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis), as well as for C3G.

In April 2016, we announced the award of an FDA Orphan Products Development grant of $500,000 to support the clinical development of avacopan for the treatment of patients with AAV in the United States.

Avacopan is in Phase III development for the treatment of patients with AAV in a pivotal trial called ADVOCATE. In May 2016, pursuant to the Avacopan Agreement, Vifor licensed the rights to commercialize avacopan for orphan renal diseases in Europe, certain other markets outside the United States and most of Asia. In February 2017, we entered into the Avacopan Amendment to expand the Vifor territories to include all markets outside the United States and China. In June 2018, we entered into the Avacopan Letter Agreement with Vifor to further expand Vifor's exclusive commercialization rights under the Avacopan Agreement to include China. We retain all rights to commercialize avacopan in the United States and also retain control of the clinical development programs for orphan renal disease. Additionally, we launched a potentially registration-supporting clinical trial to study avacopan for the treatment of patients with C3G for which we aim to complete enrollment in 2019 and initiated a large placebo-controlled Phase IIb clinical trial, the AURORA trial, for the treatment of patients with moderate to severe HS in the fourth quarter of 2018. All of these activities are designed to potentially support registration of avacopan in these indications.

AAV and C3G are orphan autoimmune diseases that are characterized by inflammation that often affects the kidneys, among other major organs. HS is a chronic, inflammatory, debilitating skin disease characterized by recurrent, painful, nodules and abscesses, ultimately leading to the formation of draining fistulas (also known as sinus tracts) as well as scarring.

ANCA-Associated Vasculitis (AAV)

AAV is an orphan, severe, and often fatal autoimmune disease that is characterized by elevated levels of autoantibodies called anti-neutrophil cytoplasmic autoantibodies and by inflammation that can affect many different organ systems, and commonly involves the kidneys. AAV affects approximately 40,000 people in the United States, with approximately 4,000 new cases each year; similarly, AAV affects approximately 50,000 people in Europe, with approximately 5,000 new cases each year.

Limitations of Current Therapies

AAV is currently treated with courses of immuno-suppressants (cyclophosphamide, or CYC, or rituximab, or RTX) combined with high-dose glucocorticoids (a type of steroid) administration. Complete remission is achieved in only 60-80% of patients and relapse is common. Following initial treatment, up to 30% of patients relapse within six to 18 months, and approximately 50% of all patients relapse within three to five years.

The current SOC for AAV is associated with significant safety risks. First year mortality is approximately 11% to 18%. The single greatest cause of premature mortality is not disease related adverse events, but rather infection and other side effects that are thought largely to be a consequence of steroid administration. The multiple adverse effects of courses of steroid treatment (both initial courses and those that are repeated as a consequence of relapse) are major causes of both short-term and long-term morbidity, damage and death. Such therapy-related adverse events contribute significantly to patient care costs, as well as to the diminution of quality of life for patients.

Role of C5a and C5aR in AAV

Complement C5a, acting through its receptor C5aR, sometimes called C5aR1 or CD88, is thought to play a pro-inflammatory role in AAV. Autoantibodies against neutrophil enzymes lead to the priming and activation of neutrophils and activation of the complement cascade. Activation of the complement cascade leads to production of C5a, one of the most potent pro-inflammatory mediators of the complement system. C5a, through binding to its receptor C5aR, induces expression of adhesion molecules and chemotactic migration of neutrophils and other white blood cells. These accumulating adhering neutrophils initiate an inflammatory cascade in the small blood vessels by secreting pro-inflammatory cytokines and chemoattractants that lead to necrotizing vasculitis.

Importantly, there are two distinct receptors for C5a: the pro-inflammatory C5a receptor known as C5aR, the target of avacopan, and the anti-inflammatory C5a-like receptor, or C5L2, which plays an important role in homeostasis. Accordingly, precisely inhibiting C5a at the level of the C5aR receptor is thought to block the pro-inflammatory effects of C5a, while leaving the protective effects of C5L2 functional. Avacopan does not bind into C5L2, thereby not interfering with the protective effects of C5L2.

Avacopan — A Novel C5a Receptor Inhibitor

Avacopan is a potent and highly specific inhibitor of C5aR, is orally bioavailable and has demonstrated an excellent preclinical safety profile, consistent with its intended chronic use in patients. Avacopan does not affect formation of the C5b-9 terminal complement complex (or MAC), unlike the anti-C5-antibody, eculizumab (Soliris®). Therefore, avacopan is believed not to increase the susceptibility to infections for which MAC is important in host defense, such as *Neisseria meningitidis*.

The efficacy of avacopan was demonstrated in a mouse model of the renal manifestations of AAV, which closely mimics many of the histological features of the human disease. In these studies, oral doses of avacopan completely blocked the glomerulonephritis induced by intravenous injection of anti-myeloperoxidase antibodies (one of the anti-neutrophil cytoplasmic antibodies that are implicated in AAV in humans). Levels of avacopan in the blood of these mice were comparable to levels in the blood of AAV patients who participated in our Phase II CLEAR and CLASSIC clinical trials with avacopan.

Clinical Development in AAV

Avacopan Phase I Clinical Trials

We have completed six Phase I clinical trials with avacopan in a total of 166 healthy subjects and 24 subjects with hepatic impairment. These studies evaluated the safety and tolerability, pharmacokinetic, or PK, and pharmacodynamic, or PD, profiles of avacopan, given orally at doses ranging from a single dose of 1 mg up to 100 mg given twice-daily for five days. Avacopan was well-tolerated and appeared to be safe in these studies. No serious adverse events or dropouts due to adverse events were observed in these studies. The most commonly reported adverse events in subjects receiving avacopan in these studies were headache, diarrhea, dizziness, sore throat, upper respiratory tract infections and decrease in white blood cells. These adverse events typically were mild and dosing was not stopped as a result.

Avacopan Phase II Clinical Trials

We have completed and reported positive clinical data from two Phase II clinical trials, known as the CLEAR and CLASSIC trials, with avacopan in patients with AAV.

CLEAR was a randomized, double-blind, placebo-controlled clinical trial in 67 patients with AAV in Europe. The aim of this trial was to provide effective therapy for AAV with an inhibitor of the C5a receptor while reducing toxicity associated with SOC therapy by eliminating or reducing exposure to high-dose systemic steroid use. The primary safety objective of this clinical trial was to evaluate the safety and tolerability of avacopan in patients with AAV on background CYC or RTX treatment. The primary efficacy objective was to evaluate the effect of avacopan based on the Birmingham Vasculitis Activity Score, or BVAS. BVAS measures AAV disease activity across all organ systems and is the most widely used and clinically validated outcome measure in AAV clinical trials. The higher the BVAS score, the higher the level of disease activity. The greater the reduction in BVAS score with treatment, the greater the disease improvement. The secondary objectives of this clinical trial included assessment of the feasibility of reducing or eliminating the use of steroids in the treatment of patients with AAV without the need for rescue steroid measures, assessment of changes in renal function based on estimated glomerular filtration rate, or eGFR, hematuria, and proteinuria with avacopan compared to SOC treatment, assessment of the effect of avacopan on health-related quality of life measurements, and evaluation of the PK and PD profiles of avacopan in patients with AAV.

The CLEAR trial met its primary endpoint based on the BVAS response at week 12 in patients receiving avacopan, compared to those patients receiving the high-dose steroid-containing SOC. Specifically, all treatment groups receiving avacopan demonstrated a statistically significant (P=0.002) non-inferior clinical efficacy outcome when compared to SOC. The study contained two avacopan-treated groups. One group received avacopan with a low dose of steroids (one third the steroid in the SOC group), in which the BVAS response was 86% at week 12 versus 70% for SOC (P=0.002 for non-inferiority). A separate group received avacopan without steroids; in this group the BVAS response was 81% (P=0.01 for non-inferiority). SOC treatment included a standard background immunosuppressant (CYC or RTX) given to all patients. The primary endpoint of BVAS response was prospectively defined as the proportion of patients with a decrease from baseline of at least 50% in BVAS plus no worsening in any body system.

Other beneficial changes were noted, including in pre-specified secondary endpoints:

(i) Avacopan exhibited a more rapid onset of improvement than SOC treatment, as evidenced by beneficial changes in proteinuria (measured as urinary albumin to creatinine ratio, or UACR); also

rapid beneficial reductions from baseline in BVAS, as well as reductions in the levels of monocyte chemoattractant protein-1, or MCP-1 (a marker of kidney inflammation), found in the urine;

(ii) Improvements in eGFR and hematuria were seen in all three treatment groups, indicating these disease activities did not require high-dose chronic steroid administration to be controlled; and

(iii) Improvements in "Quality of Life" (as defined by the visual analogue scale of the EuroQOL-5D-5L) and measurements, such as physical functioning, emotional role functioning, pain and vitality based on the Medical Outcomes Survey Short Form-36, or Short Form-36 were seen in avacopan treatment groups, but not in the SOC group.

CLASSIC was a randomized, double-blind, placebo-controlled Phase II clinical trial in patients in the United States and Canada with either newly diagnosed or relapsing AAV who required either CYC or RTX treatment. Eligible patients were randomized in a 1:1:1 ratio to receive either placebo plus CYC or RTX plus full dose starting steroids; 10mg avacopan twice-daily plus CYC or RTX plus full dose starting steroids; or 30mg avacopan twice-daily plus CYC or RTX plus full dose starting steroids. The treatment period was 12 weeks, with a 12-week follow-up period. The aim of the CLASSIC trial was different from the CLEAR trial. The CLASSIC trial was mainly a regulatory and safety trial. As such, the main goal of CLASSIC was to evaluate the safety of avacopan when given with high-dose steroid-containing SOC treatment, which also includes CYC or RTX. Therefore, the primary safety objective of this clinical trial was to evaluate the safety and tolerability of avacopan in patients with AAV on background CYC or RTX treatment. The primary efficacy objective was to evaluate the efficacy of avacopan based on BVAS. The study was not sized to formally evaluate efficacy. A total of 42 patients were enrolled in this trial.

The CLASSIC safety study met its objectives. Avacopan was shown to be well-tolerated in patients with AAV when added to the current SOC regimen. The incidence of serious adverse events was similar across treatment groups in the study. While the CLASSIC safety study was not designed or powered for inferential statistical analyses on efficacy, treatment response for each cohort was assessed at week 12 using the BVAS. Results showed that the BVAS response was numerically higher in patients receiving avacopan compared to control. The 30mg avacopan dose appeared to be most effective, based on a higher number of patients achieving early remission (based on BVAS of 0) at week four. The renal function, measured by eGFR appeared to improve most in the 30mg avacopan group, and renal response (based on improvement in hematuria, albuminuria, and eGFR) appeared to be highest in the 30mg avacopan group.

Taken together, these results suggest that avacopan, a target-specific C5aR inhibitor, may provide effective control of the disease while eliminating chronic steroids in the treatment of AAV. Avacopan also appeared safe and well-tolerated in the trials. There were no observations that would prevent further clinical development of avacopan. We also completed the long-term toxicology program with avacopan. The results provide support for chronic dosing of avacopan in future clinical trials.

We held End-of-Phase II meetings with the FDA and Protocol Assistance/Scientific Advice meetings with the EMA in 2016. Both the FDA and the EMA concurred with the design and scope of the Phase III registration clinical trial in AAV.

Avacopan Phase III Clinical Trial

In December 2016, we initiated the ADVOCATE, or Avacopan Development in Vasculitis to Obtain Corticosteroid elimination and Therapeutic Efficacy, Phase III clinical trial. ADVOCATE is a randomized, double-blind, placebo-controlled worldwide clinical trial which enrolled over 300 patients with newly diagnosed or relapsing AAV at over 200 sites in the United States, Canada, Europe, Australia, New Zealand and Japan. The aim of the trial is to assess the safety and efficacy of avacopan in inducing and sustaining remission in patients with AAV. The study includes two treatment arms: the test arm contains 30mg twice-daily oral doses of avacopan and eliminates corticosteroids, and the control arm contains an avacopan-matching placebo and

maintains a standard regimen of high-dose chronic steroids. All patients also receive a standard background immunosuppressant, either CYC followed by azathioprine or RTX. Primary endpoints will be measured by BVAS, assessing disease remission at weeks 26 and 52. Other key endpoints include early remission (BVAS of 0 at week 4), quality of life, and corticosteroid-related toxicities. Patient enrollment of the Phase III ADVOCATE trial was completed in July 2018 and we expect to report topline data from this trial in the fourth quarter of 2019 and believe, if successful, these data could form the basis of avacopan's registration for the treatment of AAV in the United States and Europe.

Avacopan Regulatory Matters

In addition to the earlier referenced orphan drug designations granted to avacopan by the FDA and the EMA, avacopan was also granted access to the EMA's PRIME initiative, which supports accelerated assessment of investigational therapies addressing unmet medical need. This was based on the assessment by the EMA that (i) AAV is a highly severe disease with high mortality; (ii) current standard therapies (including steroids) have partial efficacy and severe toxicity, indicating a high unmet medical need in AAV; and (iii) avacopan provides a new mechanism of action for the potential treatment of AAV and has the potential to significantly address the unmet medical need based on nonclinical and clinical data.

We filed a CMA application for avacopan in the treatment of patients with AAV with the EMA which was validated by the EMA in December 2017. In light of the upcoming availability of data from the pivotal Phase III ADVOCATE trial, we decided to withdraw the CMA application based on Phase II data. We intend to file integrated regulatory submissions in 2020 with the EMA and FDA for full (unconditional) marketing approval after the planned release of topline data from the Phase III ADVOCATE trial anticipated in the fourth quarter of 2019.

Complement 3 Glomerulopathy (C3G)

C3G disease is an ultra-rare disease of the kidney that is characterized by deposition of the complement fragment known as C3 in the glomeruli, or filtration units of the kidney, leading to inflammatory cell accumulation, profound kidney damage and eventual renal failure. The prevalence of C3G is estimated at two to three per million people or approximately 800 patients in the United States and approximately 2,000 in Europe.

Role of C5a and C5aR in C3G

While the disease name refers to complement 3, it is well known that the C5a receptor pathway, which is further downstream of C3 in the complement cascade and the target of avacopan, is an essential part of the disease causing pathology. Hence, C3 is a marker of more general complement activation.

Limitations of Current Therapies

There is currently no approved effective standard therapy for C3G. Typically, patients receive one or more non-specific immunosuppressants. Without treatment, C3G leads to kidney failure, and the current array of unapproved therapies at best only delays end stage renal disease, or ESRD. Kidney transplant is frequently the only option, and even after transplantation, the disease invariably returns.

Clinical Development in C3G

Avacopan Phase II Clinical Trial in C3G

We launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G. The clinical trial is expected to enroll approximately 88 patients with C3G, including both C3 Glomerulonephritis and Dense Deposit Disease. The primary objective is to evaluate the efficacy of avacopan

compared to placebo based on histologic changes in kidney biopsies taken at baseline and after 26 weeks of treatment. The primary endpoint will be based on the percent change from baseline in the C3G Histologic Index for disease activity.

The secondary objectives of this trial include evaluation of: (i) the safety of avacopan compared to placebo based on the incidence of adverse events, changes in clinical laboratory measurements, and vital signs; (ii) changes in laboratory parameters of renal disease including eGFR, proteinuria, and urinary excretion of MCP-1 with avacopan compared to placebo; (iii) health-related quality-of-life changes based on Short Form-36 version 2, or SF-36 v2, and EuroQOL-5D-5L, or EQ-5D-5L, with avacopan compared to placebo; and (iv) the PK profile of avacopan in patients with C3G.

Patients meeting inclusion criteria start study drug treatment on Day 1. Patients receive avacopan 30mg or matching placebo orally twice-daily. The placebo-controlled treatment period is 26 weeks (182 days). This will be followed by 26 weeks during which time all patients will receive avacopan. Thereafter, all patients will be followed for eight weeks (56 days) without study drug treatment.

Hidradenitis Suppurativa (HS)

HS is a chronic, inflammatory, debilitating skin disease characterized by recurrent, painful, nodules and abscesses, ultimately leading to the formation of draining fistulas (also known as sinus tracts) as well as scarring. The diseases originates from inflammation and occlusion of the hair follicle. Apart from pain, the nodules may rupture, and often extrude a purulent, foul-smelling discharge leading to substantial social embarrassment for these patients. Due to its chronic nature and frequently occurring relapses of the skin lesions, HS has a great impact on the patient's quality of life, deeply affecting social, working, and psychological aspects.

In the United States, moderate to severe HS has orphan designation with an estimated prevalence of up to 200,000 patients. In Europe, the number of affected patients is believed to be greater, with higher prevalence.

Role of C5a and C5aR in HS

Neutrophils are believed to play an important disease-promoting role, as well as certain cytokines and mediators commonly found in autoimmune diseases, such as TNF-alpha, IL-17, IL-1 and others such as C5a. C5a promotes inflammatory mediators and is a strong activator of neutrophils. HS is a neutrophil-driven skin disease and C5a has been found activated and significantly elevated in plasma of HS patients, as compared to healthy controls. In an open label Phase IIa study in 12 patients with moderate and severe HS, a specific intravenous anti-C5a antibody was shown to improve skin lesion in patients with moderate to severe HS.

With the role of C5a in HS, our C5aR antagonist avacopan could be effective in mediating the disease course of HS. Avacopan is a small molecule that is conveniently administered as an oral medication and could present itself as advantageous over intravenous or subcutaneous injections treatments for this condition.

Limitations of Current Therapies

Depending on the severity of disease, the current standard of care for HS patients includes topical, oral or intravenous antibiotic treatment, as well as surgery.

Adalimumab, an anti-TNF-alpha monoclonal antibody, is the only drug indicated for the treatment of patients with moderate to severe HS. Two pivotal adalimumab trials showed that approximately 50% of the patients who were treated with adalimumab achieved an improvement in their skin lesion, as measured by the widely accepted HiSCR (Hidradenitis Suppurativa Clinical Response) assessment instrument. There remains a high unmet medical need, however, as a very large proportion of the patients with moderate to severe HS do not adequately respond to adalimumab or other therapies used in the standard of care.

Avacopan Clinical Development in HS

Based on the role of C5a in the pathogenesis of HS, we believe that there is significant interest in the medical-scientific community to develop avacopan for the treatment of patients in HS. We initiated a clinical trial, the AURORA trial, of avacopan in patients with moderate to severe HS in the fourth quarter of 2018.

The AURORA trial is a randomized, double-blind, placebo-controlled, three arm Phase IIb trial in approximately 390 subjects with moderate to severe HS (Hurley stage II or III). Subjects will be randomized 1:1:1 to a treatment of 10 mg avacopan twice-daily, 30 mg avacopan twice-daily or placebo for 12 weeks. Subjects treated with 10 mg or 30 mg twice-daily during the blinded, placebo-controlled 12-week treatment period will be followed by an additional 24-week, active treatment period during which they will continue to receive the same dose regimen, either 10 mg or 30 mg avacopan twice-daily. Subjects on placebo who complete the blinded, placebo-controlled 12-week period will be re-randomized 1:1 to receive 10 mg or 30 mg avacopan twice-daily during the 24-week active treatment period. Thereafter, all subjects will be followed without study drug for eight weeks before they exit the study.

The primary efficacy objective is based on subjects achieving a HiSCR after 12 weeks of treatment. HiSCR is defined as at least a 50% reduction in abscess and inflammatory nodule count and no increase in abscess count and no increase in draining fistula count at Week 12 relative to baseline. The primary safety objective is to evaluate the safety of avacopan compared to placebo based on the adverse event incidence, changes from baseline in laboratory parameters, and vital signs. Secondary objectives include the evaluation of the subject's global assessment of skin pain numeric rating scale, the Sartorius score, International Hidradenitis Suppurativa Severity Score, Hidradenitis Suppurativa-Physician's Global Assessment, and the proportion of subjects who experienced flare or who received rescue therapy, and the duration of flare. Other secondary objectives include the assessment of subject reported outcomes including health-related quality-of-life changes based on the SF-36 v2, the EQ-5D-5L, and the Hidradenitis Suppurativa Quality of Life Index as well as the evaluation of the pharmacokinetic profile of avacopan in subjects with HS.

Avacopan Commercialization Strategy

We plan on building a sales infrastructure in the United States to commercialize our orphan disease drug candidates, including avacopan. Given that two of the target indications for which avacopan is being developed may have significant renal involvement, we expect that our future sales force will focus primarily on nephrologists. Other physician specialists such as rheumatologists, involved in the diagnosis and treatment of those diseases would also be targeted by our sales forces. In territories outside of the United States, our partner Vifor would be responsible for the commercialization of avacopan.

In May 2016, we entered into the Avacopan Agreement with Vifor to commercialize avacopan for orphan renal diseases in Europe and certain other markets. In connection with the Avacopan Agreement, we received a non-refundable upfront payment of $85.0 million, comprising $60.0 million in cash and $25.0 million in the form of an equity investment to purchase 3,333,333 shares of our common stock at a price of $7.50 per share. In February 2017, we and Vifor entered into the Avacopan Amendment to expand the licensed territory to include all markets outside the United States and China and we received an additional $20.0 million upfront cash commitment. Upon achievement of certain regulatory and sales based milestones with avacopan, we will receive additional payments under this agreement. In addition, we will receive royalties, with rates ranging from the teens to mid-twenties, on future potential net sales of avacopan by Vifor in the licensed territories. In December 2017, we achieved the first regulatory milestone under the Avacopan Agreement in the amount of $50.0 million, following the EMA's validation of the CMA application for avacopan for the treatment of patients with AAV. In June 2018, we and Vifor entered into the Avacopan Letter Agreement to further expand the Vifor territories under the Avacopan Agreement to provide Vifor with exclusive commercialization rights in China and we received a $5.0 million payment for the expanded rights. We retain control of ongoing and future development of avacopan (other than country-specific development in the licensed territories) and all commercialization rights to avacopan in the United States.

Under a prior development and commercialization agreement with Glaxo Group Limited, or GSK, an affiliate of GlaxoSmithKline, which ended in 2013, we are subject to reverse royalties to GSK of 3% on annual worldwide net sales of avacopan, not to exceed $50.0 million in total royalties.

CCX140 — Chronic and Orphan Kidney Diseases

Our second drug candidate in the orphan disease space is CCX140, an inhibitor of the chemokine receptor known as CCR2. CCX140 is an orally-administered small molecule that is a highly potent and selective inhibitor of the chemokine receptor known as CCR2. CCX140 has an excellent preclinical and clinical profile, including good safety and tolerability demonstrated in hundreds of healthy volunteers or patients with diabetes or diabetic nephropathy across seven clinical trials. These clinical trials include a successfully completed one-year dosing of CCX140 in a Phase II trial in CKD associated with diabetes. Preclinical data to date suggests CCR2 inhibition involves a unique mechanism of action in the kidney including a novel element of renal cellular protection at the level of the podocyte leading to rapid improvement in proteinuria. CCX140 has been granted orphan drug designation by the FDA for the treatment of FSGS.

Focal Segmental Glomerulosclerosis (FSGS)

FSGS is a histologic lesion that is associated with the clinical presentation, in children or adults, of proteinuria, nephrotic syndrome and progressive renal insufficiency. Nephrotic syndrome is the combination of nephrotic-range proteinuria (loss of more than three grams of protein per day into the urine) with a low serum albumin level and edema. Each kidney is made up of approximately one million tiny filters called “glomeruli.” Glomeruli filter blood, taking out the water-like part that becomes urine and leaving protein in the blood. When glomeruli or sections of the glomeruli become damaged or scarred (sclerosis), proteins leak into the urine (proteinuria). FSGS is understood to start with damage to podocytes, cells that wrap around capillaries of the glomerulus. Podocytes form part of the barrier that enable the glomerulus to filter the blood in a manner that retains large molecules such as proteins, while smaller molecules such as water, salts, and sugars are filtered as the first step in the formation of urine.

FSGS is classified as primary or idiopathic when the cause is not known, and secondary when it occurs in the setting of recognized genetic mutations or associated disease. The distinction between primary and secondary FSGS can be difficult, but it has been estimated that in 80% of the cases the etiology is unknown. Primary or idiopathic FSGS often presents with the nephrotic syndrome. Secondary FSGS, which most often presents with non-nephrotic proteinuria and some degree of renal insufficiency, can occur in the setting of genetic vulnerability, podocyte injury due to toxins or infections, or as an adaptive response to glomerular hypertrophy or hyper-filtration.

Symptoms or signs of FSGS may not be noticeable early in the course of disease, presenting only when sufficiently advanced to cause edema, or when physical examination and laboratory assessment reveal proteinuria, low blood albumin levels, high cholesterol and/or high blood pressure. FSGS is a disease characterized by progressive glomerular scarring and is life-threatening. In 20% of children and in 40% of adults, it is the underlying cause of nephrotic syndrome. When accompanied by high levels of proteinuria at the time of presentation, 50% of patients with FSGS will progress to ESRD within three to eight years. FSGS is causal for 4% of all ESRD cases. Furthermore, after kidney transplantation for primary FSGS, the recurrence rate is 40%.

FSGS is a rare form of CKD that affects approximately 80,000 patients in the United States, with 5,500 to 9,500 new cases each year. FSGS attacks the glomeruli causing scarring which leads to permanent kidney damage. Progressive FSGS can lead to ESRD, ultimately requiring kidney transplant or renal dialysis and total health expenditures of hundreds of thousands of dollars each year per patient.

Current Treatment Approaches

There are no approved drugs for the treatment of FSGS. Moreover, current treatment approaches are not very effective in halting the disease. Usually, treatments for FSGS include renin-angiotensin-aldosterone, or

RAAS, system blockers, corticosteroids, immunosuppressive drugs, diuretics and diet change (reducing sodium and protein intake).

RAAS blockade reduces proteinuria and slows progression in proteinuric kidney diseases, and is commonly used for treatment of secondary FSGS. Whether or not this is effective in primary FSGS is unknown. Patients with histologic evidence of primary FSGS who have nephrotic syndrome are usually offered disease-modifying therapy with glucocorticoids and other immunosuppressive drugs. However, in the absence of nephrotic range proteinuria (>3.5 g/day), administration of steroids or other immunosuppressive drugs is generally not recommended. In many cases, an overall course of treatment of at least six months is required and complete remission may not be attained for 12 months or longer. Shorter courses (two months or less) result in much lower remission rates (20% to 30%).

Patients with little or no reduction in protein excretion at 12 to 16 weeks are considered steroid resistant. Initial therapy of steroid-dependent or steroid-resistant FSGS consists of a calcineurin inhibitor (cyclosporine or tacrolimus) with or without low-dose prednisone. Among those unresponsive to this combination, or among those with substantially reduced eGFR (<40 mL/min per 1.73 m^2), mycophenolate mofetil in combination with glucocorticoids is recommended. In addition, in patients at increased risk for glucocorticoid-associated toxicity (e.g., obese patients, diabetic patients, patients with severe osteoporosis, patients >70 years of age), cyclosporine or tacrolimus with or without low-dose prednisone has been recommended, although data evaluating this strategy are limited. Calcineurin inhibitors must be used with caution in patients with impaired renal function because of the nephrotoxicity of these drugs, and some authors recommend avoiding these in subjects whose kidney function approaches renal failure, i.e., subjects with an eGFR of <30 mL/min/1.73 m^2.

Limitations of Current Therapies

As described above, there are no effective therapies available for FSGS. Control of hypertension, particularly with angiotensin inhibitors, is supportive but does not address the underlying pathology. Glucocorticoids and immunosuppressants are also used, but the results have been inconsistent. The increased risk of infection associated with these agents is a significant concern. Histologic recurrence in renal transplants is high, with high levels of proteinuria portending a poor renal prognosis.

Role of CCR2 inhibition in FSGS

There is evidence that the chemokine receptor known as CCR2 plays a role in the pathogenesis of FSGS. CCR2 is a major driver of monocyte migration and activation, and has been shown to mediate renal interstitial inflammation and tubular atrophy in a number of chronic renal diseases by recruiting monocytes to the renal interstitium. Further, studies have shown that the degree of protein excretion correlates with urine MCP-1 levels, one of the signature ligands of CCR2 and a biomarker of inflammation, and the infiltration of immune cells called macrophages into the kidney in patients with CKD. Experiments performed in vitro have added to the mechanistic rationale for the notion that CCR2 is an important driver of FSGS. Proteinuria is the hallmark characteristic of FSGS, and in vitro experiments have found that tubular epithelial cells release MCP-1 when exposed to serum proteins on the inside of the tubules. Clinically, in children with FSGS, urinary MCP-1 levels correlate with the degree of proteinuria.

Blocking CCR2 provided significant and rapid renal protection in two distinct models of FSGS, as measured both by reduction in proteinuria and improvement in multiple histological parameters, and it thus represents a novel and mechanistically-distinct approach for the treatment of FSGS.

In the 5/6 remnant kidney model, mice had a rapid reduction in protein excretion when treated with a CCR2 inhibitor and a RAAS inhibitor. Combining a CCR2 inhibitor with a RAAS inhibitor reduced the protein excretion by 91%. The protective effects were evident within one week of treatment and were maintained for the duration of the study (six weeks). The same renal protective effects of CCR2 blockade were seen in the

adriamycin nephropathy model. Administration of adriamycin caused significant proteinuria, which was significantly reduced by the combination of a CCR2 inhibitor and a RAAS inhibitor after two weeks of treatment. Histological parameters also improved with the combination of the CCR2 inhibitor and a RAAS inhibitor; these included reduced glomerular hypertrophy, glomerular sclerosis, kidney fibrosis, and mesangial expansion and increased podocyte density. Further, we demonstrated marked histological improvements in an FSGS animal model through inhibition of CCR2, including increased density of podocytes. The data suggest CCR2 inhibition involves a unique mechanism of action in the kidney including a novel element of renal cellular protection at the level of the podocyte.

Clinical Development

Our clinical development strategy was to first assess the safety and tolerability of CCX140 in healthy subjects, then in patients with type 2 diabetes and normal renal function, and finally to evaluate the drug in patients with DN. As a precursor to our clinical trials in patients with DN, we completed a 159-patient randomized Phase II clinical trial to assess the safety and tolerability of CCX140 in patients with type 2 diabetes, one of the most common causes of nephropathy. We also subsequently completed a 332-patient randomized Phase II clinical trial to assess the efficacy, safety, and tolerability of CCX140 in patients with DN.

Based on safety and encouraging efficacy signals related to reduction in proteinuria and stabilization/improvement of renal function observed in the Phase II study in patients with DN, we launched our clinical-development program of CCX140 for the treatment of patients with FSGS.

CCX140 Phase I Clinical Trials

We completed four Phase I clinical trials in 118 healthy volunteers. A CCX140 dose range of 0.05 to 15mg was studied. CCX140 was generally well-tolerated with no serious adverse events observed in these Phase I clinical trials. The PK profile was supportive of once-daily oral dosing of CCX140 in the Phase II clinical trials in patients with type 2 diabetes and in patients with DN.

CCX140 Phase II Clinical Trial in Type 2 Diabetes

Our Phase II clinical trial was designed to demonstrate safety of CCX140 in patients with type 2 diabetes and normal renal function, and to examine the effect of CCX140 on glycemic indices. We conducted a randomized, double-blind, placebo and active controlled clinical trial in 159 patients with type 2 diabetes on a stable dose of metformin for at least eight weeks, with 32 patients receiving placebo, 32 receiving pioglitazone hydrochloride (an approved therapeutic for type 2 diabetes serving as the active control), 63 receiving 5mg of CCX140 and 32 receiving 10mg of CCX140 orally once-daily for 28 days.

The clinical trial met its primary objective by demonstrating the safety and tolerability of CCX140 in these patients. In addition, CCX140 showed encouraging signs of biological activity based on a statistically significant decrease in HbA1c, a marker of glycemic control, for the 10mg dose group.

CCX140 Phase II Clinical Trial in Diabetic Nephropathy

We completed a Phase II clinical trial in patients with DN. A total of 332 patients were enrolled in a randomized, double-blind, placebo-controlled clinical trial. The goals of this clinical trial were to evaluate the efficacy, safety and tolerability of CCX140 in patients with DN. The primary efficacy objective was evaluation of the effect of CCX140 on albuminuria. Secondary efficacy objectives were evaluation of the effect of CCX140 on HbA1c and eGFR. The three treatment groups consisted of (i) SOC, (ii) a RAAS inhibitor, plus placebo (control group), and (iii) 5mg and 10mg of CCX140 once-daily plus SOC. The treatment duration was up to 52 weeks, with a four-week follow-up period. Patients with residual albuminuria, despite being on a stable therapeutic dose of a RAAS inhibitor, were included in this clinical trial. The key efficacy endpoint was a change from baseline in first morning UACR, a major indicator of renal health.

The Phase II trial met its primary endpoint by demonstrating that treatment with 5mg of CCX140 given orally once-daily added to a SOC regimen of RAAS inhibitor treatment resulted in a statistically significant (p=0.01) improvement in UACR, beyond that achieved with SOC alone. The maximum treatment effect (24% reduction) was reached at 12 weeks, and sustained reduction in albuminuria induced by CCX140 relative to SOC alone was observed over the full year (UACR at each one of the ten time points over the 52-week treatment period in the patients who received 5mg CCX140 continuously for 52 weeks, were below those of the SOC alone group). A dose of 10mg CCX140 per day did not provide more improvement in albuminuria as compared to the 5mg dose. CCX140 did not affect systematic blood pressure, suggesting that the beneficial effect of CCX140 is mediated locally in the kidney micro-environment, possibly through a beneficial reduction in renal inflammation. CCX140 was well-tolerated with a low overall dropout rate over the 52-week treatment period (10%). No safety issues were observed that would prevent further clinical development of CCX140 in DN.

CCX140 Phase II Clinical Trials in Primary Focal Segmental Glomerulosclerosis

The successfully completed CCX140 Phase II clinical trial in DN, which demonstrated a statistically significant reduction in proteinuria compared to SOC, showed the most pronounced effect in the highest proteinuric patients. In addition, preclinical data to date suggest CCR2 inhibition involves a unique mechanism of action in the kidney including a novel element of renal cellular protection at the level of the podocyte, leading to rapid improvement in proteinuria. Building upon our orphan kidney disease franchise, we launched our clinical development program of CCX140 in patients with primary FSGS. Our clinical development program will comprise two populations of patients:

(i) Primary FSGS patients with sub-nephrotic (moderate) proteinuria (>1 g/g) – LUMINA 1 trial; and

(ii) Primary FSGS patients with nephrotic syndrome (heavy proteinuria>3.5 g/g) – LUMINA 2 trial.

The first trial is a randomized controlled, dose ranging study with the aim to evaluate the effect of treatment with CCX140 in subjects with sub-nephrotic primary FSGS. The second trial is an open label, intra-subject dose escalation study in subjects with primary FSGS and nephrotic syndrome. The primary efficacy objective of both trials is to evaluate the effect of CCX140 on proteinuria, as assessed by factors including reduction in proteinuria through the 12-week treatment period. Other study objectives will include the assessment of partial or complete remission and the time to, and proportion of subjects with, partial or complete remission and other variables. Changes in quality-of-life changes (SF-36 v2, EQ-5D-5L) will also be assessed.

CCX140 Commercialization Strategy

We plan on building a sales infrastructure in the Unites States to commercialize our orphan disease drug candidates such as CCX140. FSGS patients are primarily being treated by nephrologists in so-called centers of excellence in renal diseases. Similar to avacopan, we plan on establishing a commercial presence focused on the nephrology arena. In territories outside of the United States, our partner Vifor will be responsible for the commercialization of CCX140.

In December 2016, we entered into a second collaboration and license agreement with Vifor, the CCX140 Agreement, pursuant to which we granted Vifor exclusive rights to commercialize CCX140 in rare renal diseases in markets outside the United States and China. We are responsible for the clinical development of CCX140 in rare renal diseases, while sharing the cost of such development with Vifor. In connection with the CCX140 Agreement, we received a non-refundable upfront commitment totaling $50.0 million and are eligible to receive additional payments upon the achievement of certain regulatory and sales based milestones, as well as tiered double-digit royalties on potential net sales of CCX140 in the licensed territories. Under the CCX140 Agreement, Vifor retains an option to solely develop and commercialize CCX140 in more prevalent forms of CKD. Should Vifor later exercise the CKD option, we would receive co-promotion rights in CKD in the United States. In June 2018, we and Vifor entered into the CCX140 Letter Agreement to further expand the Vifor territories under the CCX140 Agreement to provide Vifor with exclusive commercialization rights in China and we

received a non-refundable $5.0 million payment for the expanded rights. Additionally, in June 2018, we and Vifor entered into an amendment to the CCX140 Agreement, which we refer to as the CCX140 Amendment, to clarify the timing of certain payments with respect to development funding of the CCX140 program by Vifor, and we received a non-refundable payment of $11.5 million. We retain control of ongoing and future development of CCX140 (other than country-specific development in the licensed territories) and all commercialization rights to CCX140 in the United States.

Early Stage Drug Candidates

Immuno-Oncology and Other Therapeutic Areas

In oncologic disease, tumors can profoundly subvert inflammatory and effector immune responses. In the tumor cellular microenvironment, CCR2 bearing cells are thought to largely have an immunosuppressive behavior. These are the so-called myeloid derived suppressor cells, or MDSCs. These cells effectively help tumors hide from the body's cytotoxic immune response to tumor cells. Inhibiting CCR2, and thus the MDSCs controlled by CCR2, could therefore lead to the liberation of the cytotoxic immune response against the tumor cells, tumor shrinkage, and improved patient survival. We have an ongoing clinical development program for the treatment of patients with advanced pancreatic cancer with our drug candidate CCX872, our second inhibitor of the chemokine receptor known as CCR2.

Understanding Pancreatic Cancer

Pancreatic cancer is a rare but deadly cancer. It is the 15th most common cancer worldwide but the fourth highest cause of cancer-related death. In the United States in 2019, approximately 56,770 people are expected to develop pancreatic cancer, and within five years of diagnosis, about 80% of those patients are expected to succumb to the disease. Primarily due to the aging of the population, the incidence of pancreatic cancer is predicted to increase to 62,000 new cases per year by 2030. Pancreatic adenocarcinoma, which represents 85% of all pancreatic cancers, is characterized by rapid progression and a dismal prognosis. Because of the deep location of the pancreas in the abdomen and the lack of markers of early disease, most cancers remain asymptomatic until they obstruct the biliary tract, which usually occurs with tumors of the pancreatic head, or until they become metastatic. Hence, less than 15% of patients initially present with a resectable cancer (stage 1 or 2), while the majority of patients have either a locally advanced, nonresectable, stage 3 cancer or a metastatic, stage 4 cancer at the time of diagnosis. Even with the best current treatment, the median overall survival of these patients is less than one year, an outlook that has remained largely unchanged over the last few decades.

The dismal prognosis of this cancer results from the combination of the late diagnosis, the early metastatic dissemination, and resistance to most chemotherapies. The main factors explaining this resistance to treatment include a very high rate of activation of the Kirsten rat sarcoma viral oncogene, mutations, a propensity for both local extension and distal spreading, the presence of a dense stromal tissue surrounding the tumor that results in a hypoxic, hypovascularized environment with high interstitial pressure, which may impede drug delivery, and ultimately the loss of immune control. Therapeutic interventions that improve the prognosis of patients with pancreatic cancer are urgently needed.

Limitations of Current Therapies

Current SOC regimens are not only limited by modest efficacy but also by significant toxicity. For patients with nonresectable cancer (stage 3 or 4), FOLFIRINOX (oxaliplatin, irinotecan, leucovorin, and 5-fluorouracil) or a combination of gemcitabine and nanoparticle albumin-bound-, or nab-, paclitaxel are considered standard treatments, but the median overall survival of patients remains less than one year. Further, these treatments often are poorly tolerated. FOLFIRINOX is associated with a high rate of Grade 3-4 adverse events, can rarely be administered for more than six months, and is mostly prescribed to patients with excellent performance status. Frail, elderly patients usually receive palliative treatment. Extensive research is ongoing to identify novel agents with improved efficacy and a reduced toxicity profile, including chemotherapies with improved formulations of currently available agents, therapies targeted against specific oncogenic pathways, or cancer vaccines.

Role of CCR2 in Pancreatic Cancer

Human pancreatic tumors are characterized by a highly immunosuppressive microenvironment. In the tumor cellular microenvironment, CCR2 bearing cells are thought to be largely of an immunosuppressive behavior; these are the so-called MDSCs. These cells effectively help tumors hide from the body's cytotoxic immune response to tumor cells. Inhibiting CCR2, and thus the MDSCs controlled by CCR2, could therefore lead to the liberation of the cytotoxic immune response against the tumor cells, and improved patient survival.

Clinical Development

CCX872 is a potent and selective inhibitor of CCR2. The objective of using a CCR2 inhibitor such as CCX872 is to reduce the suppressive myeloid cell presence in the tumor and, in doing so, slow the progression of disease in these patients. We believe that CCX872 may represent a promising novel immunotherapeutic approach. Drugs that block CCR2 have shown evidence of activity in patients with pancreatic cancer as well as in a mouse orthotopic pancreatic cancer model.

Phase I Clinical Trials

We completed a first-in-human Phase I clinical trial in healthy subjects. This clinical trial was a combined single-and-multiple-ascending dose clinical trial in 40 subjects. The clinical trial was conducted in the Netherlands. CCX872 doses of 3mg, 10mg, 30mg, 100mg and 300mg were given as a single dose in the first study period and once-daily doses for seven days in the second study period. Data showed that CCX872 was well-tolerated and appeared to be safe in healthy volunteers at all dose levels studied. There were no serious adverse events or dropouts due to adverse events in the trial. The most common adverse events reported by subjects receiving CCX872 in the multi-dose period were dizziness, diarrhea, and headache. These events typically were mild in intensity and did not result in dosing discontinuation. The results showed that CCX872 was safe and well-tolerated. CCX872 was able to block CCR2 in the circulation, and it had a predictable dose-linear PK profile.

Our Phase Ib study for CCX872 explores a novel approach (CCR2 inhibition) for the treatment of patients with stage 3 and 4 pancreatic cancer. Beyond the field of pancreatic cancer, the results of this study will also advance our understanding of the role of chemokines in solid tumors and of the potential for chemokine receptor inhibitors as therapeutic options in cancer patients when combined with SOC regimens. The primary aim of this study is to evaluate the safety and efficacy of orally-administered CCX872 with respect to disease progression in patients with nonresectable pancreatic cancer being treated with FOLFIRINOX, one of the current SOC treatments for this disease. Enrollment in the trial occurred in two stages, Part A (single dose) and Part B (multiple dose). Part A has been completed. Results showed that a single oral dose of 150-mg CCX872 was well-tolerated and safe in this study. The PK profile in patients with pancreatic cancer was in line with the PK profile observed in healthy volunteers in the previous clinical trial. CCX872 was effective in blocking CCR2 in circulating cells as measured by CCR2 occupancy and internalization assays, as well as migration assays. Successful completion of Part A led to initiation of Part B.

Enrollment of 50 patients in Part B was completed in 2016. In January 2017, we reported 24-week progression-free survival, or PFS, data, 12-week objective response rate, or ORR, data, and initial overall survival data. PFS rate was 57% at week 24 in the primary analysis population and median PFS was 179 days. ORR was 37% at week 12 in the primary analysis population. Overall survival rate was 52% at week 48 in the primary analysis population and median survival time was 11.5 months. The longest ongoing CCX872 treatment period for a patient in the study to date is 73 weeks (and continuing). CCX872 has been well-tolerated in the clinical trial. There has been no apparent additional safety burden of combining CCX872 with FOLFIRINOX, as evidenced by an incidence and rate of adverse events in the trial to date consistent with data reported historically for FOLFIRINOX on its own.

In January 2018, we further reported overall survival data at 18 months of 29% for the primary analysis population and 28% in patients with metastatic pancreatic disease. This compares favorably with previously

published data of overall survival of 18.6% at 18 months for FOLFIRINOX regimen alone in metastatic pancreatic cancer. Overall, circulating monocytic myeloid derived cells likely including myeloid suppressor cells were reduced by treatment.

Preclinical Development in Immuno-Oncology

One of the most exciting advances in oncology in decades is the recent observation that modifiers of the activity of the patient's own immune system can profoundly enhance their response to chemotherapy.

A critical cellular component of this response are the MDSCs, which inhibit the activity of the effector T cells, and thus dampen the immune response of the body to the tumor. These MDSCs express chemokine and chemoattractant receptors that they use to migrate to the tumor microenvironment. We believe that blocking these chemokine receptors with small molecule antagonists could be effective either as stand-alone therapies for certain cancers or by synergistic effect when given in combination with traditional chemotherapies or other immunotherapies.

We have discovered small molecule inhibitors that target these chemoattractant receptors, and one or more of them may be developed in certain oncology indications targeting both solid and liquid tumors.

In our preclinical research, we are conducting studies with various chemokine receptor inhibitors in combination with check point inhibitors, such as those inhibiting the programmed cell death-1, or PD-1, programmed death-ligand 1, or PD-L1, pathway, that we believe may result in a greater anti-tumor effect, than with check-point inhibition alone.

A growing body of data suggests that a number of chemokine receptors, including, but not limited to, CCR1, CCR2, CCR4, CCR5, CXCR7 and CXCR2, may play diverse roles in cancer growth, cancer metastasis, cancer angiogenesis, or the composition of the tumor microenvironment. Given the potential role of chemokine receptors in cancer cell survival, the combination of chemokine receptor antagonists with traditional chemotherapeutic agents or with immunotherapy, such as PD-1 or PD-L1 inhibitors is an attractive strategy because it may result in greater efficacy and/or allow dose reductions of the chemotherapeutic drugs and therefore limit systemic side effects.

In March 2018, we presented data from *in vivo* model of colorectal cancer with a selective orally-administered CCR2 inhibitor. We reported that the therapeutic effects of PD-1 therapy are appreciably enhanced by specific blockade of CCR2 via a small molecule inhibitor, CCX598. In this mouse model, the anti-tumor response is specific and long-term survivors are resistant to re-inoculation with the CT26 tumor (even without further dosing of either drug). CCR2 inhibition alters the tumor microenvironment by reducing the number of monocytic-MDSC per gram of tumor. Reduction in tumor size is inversely proportional to the ratio of CD8 T cells to monocytic-MDSCs.

In November and December 2018, we presented data from animal models of pancreatic cancer (KCM model) and colorectal cancer (CT26 model) with a selective orally-administered CCR4 inhibitor at two international Cancer Immunology conferences. We reported that specific blockade of CCR4 with a small molecule inhibitor CCX6239 significantly reduced tumor burden and also enhanced the therapeutic effects of anti-cytotoxic T-lymphocyte-associated protein 4 (anti-CTLA-4) antibody in the KCM orthotopic pancreatic cancer model. In the colorectal cancer model, CCR4 inhibitor in combination with anti-CTLA-4 antibody also decreased tumor growth and increased the proportion of long-term survivors. In this model, the anti-tumor response was CT26-specific; mice with tumor regression exhibited a high proportion of CD8 T cells that recognized a CT26-specific neoantigen, and long term survivors were resistant to re-inoculation with CT26 cells (without further dosing of either drug).

At these same conferences, we for the first time reported the development of a unique class of human PD-L1 small molecule inhibitors. These inhibitors exhibited activity in blocking the PD-1/PD-L1 interaction in a variety

of biochemical and cell-based assays. This inhibitory effect was at least partially through PD-L1 reduction on the cell surface, a potentially beneficial feature not exhibited by anti-PD-L1 antibodies. In an animal tumor model, our current lead compound CCX4503 reduced tumor growth to a similar extent as the positive control anti-human PD-L1 antibody. The anti-tumor activity was completely dependent on the presence of human peripheral blood mononuclear cells, and was accompanied by a significantly higher CD8+ T-Cell to CD4+ T-cell ratio. With further optimization, we believe these novel PD-L1 inhibitors would provide a valuable alternative to the current antibody-based PD-1/PD-L1 therapeutics.

Other Therapeutic Areas

CCX872, a selective orally-administered inhibitor of the chemokine receptor known as CCR2, demonstrated significant reductions in liver fibrosis *in vivo* models of NASH, a severe type of non-alcoholic fatty liver disease caused by chronic inflammation that can lead to fibrosis (scarring) of the liver, when compared to either placebo or a separate compound which is a dual inhibitor of the chemokine receptors CCR2 and CCR5 currently in clinical development by another party. The data suggest a potential application of CCX872 for the treatment of patients with NASH. NASH affects 3% to 5% of the U.S. population.

Other Inflammatory and Autoimmune Diseases

Th17 Driven Diseases and CCR6

One of the most intriguing areas of current research in immunology involves a relatively recently discovered type of helper T cells known as Th17 cells. There is a large amount of preclinical and clinical data that implicate Th17 cells, as well as Interleukin 17, or IL-17, in the development of a large number of autoimmune diseases, including psoriasis, rheumatoid arthritis, asthma, and multiple sclerosis.

Activated Th17 cells isolated from chronically inflamed human tissues produce high levels of TNF-α and other cytokines. A hallmark of Th17 cells is that they express high levels of the chemokine receptor known as CCR6, which is not found on Th1 and Th2 cells. High levels of the CCR6 chemokine ligand, CCL20, have been found in psoriatic skin, in rheumatoid arthritis joint biopsies, and in asthmatic lungs.

We believe that these are potential therapeutic opportunities for a CCR6 inhibitor. We have produced several unique CCR6 inhibitor leads, which are now being optimized through medicinal chemistry approaches and undergoing further evaluation in preclinical pharmacology models.

We have shown in preclinical models that an orally bioavailable, small molecule inhibitor of the chemokine receptor known as CCR6 confers protection against IL17-mediated inflammation. We have generated potent orally bioavailable CCR6 inhibitors that inhibit CCL20-mediated chemotaxis of both human and mouse CCR6-positive cells. The utility of CCR6 inhibition was tested in preclinical models of psoriasis, and demonstrated that animals treated with our CCR6 inhibitor were protected against imiquimod induced skin thickening. Histological analysis of the skin confirmed the protective effect of our CCR6 inhibitor compared to an aqueous vehicle control and significantly reduced ear-thickening induced by intradermal injections of Interleukin 23, or IL-23, a cytokine that is important for the terminal differentiation and pathogenicity of Th17 cells.

The mechanism of action for CCR6 inhibitors is different from other therapeutics targeting IL-17, because inhibition of CCR6 disrupts the recruitment of infiltrating leukocytes into the epidermis upon skin damage, thereby protecting against epidermal hyperplasia, or an abnormal increase in the number of cells on the skin. Thus, pharmacological inhibition of CCR6 with an orally bioavailable small molecule inhibitor mitigates IL-17-driven inflammation in psoriasis models, and its distinct mechanism of action suggests it may offer additional efficacy when added to current SOC.

In April and May 2018, we presented data from *in vivo* models of psoriasis with a selective orally-administered CCR6 inhibitor. Genetically modified mice demonstrate that psoriatic lesions do not progress in

mice lacking chemokine receptor CCR6. CCL20, the only known chemokine ligand for CCR6, is highly expressed in psoriatic plaques. Our potent, orally bioavailable small-molecule inhibitor of CCR6 ameliorated skin inflammation in the IL-23 and imiquimod induced models of psoriasis, and in the IL-36 induced model representative of rare form of psoriasis referred to as generalized pustular psoriasis. Results from this work were submitted and accepted to be published as a peer reviewed journal article in the Journal of Immunology in January 2019. CCR6 antagonists present a novel therapeutic approach to treating multiple forms of psoriasis.

Intellectual Property

Our commercial success depends in part on our ability to obtain and maintain proprietary protection for our drug candidates, novel biological discoveries, screening and drug development technology and other know-how, to operate without infringing on the proprietary rights of others and to prevent others from infringing our proprietary rights. Our policy is to seek to protect our proprietary position by, among other methods, filing U.S. and foreign patent applications related to our proprietary technology, inventions and improvements that are important to the development and implementation of our business. We also rely on trade secrets, know-how, continuing technological innovation and potential in-licensing opportunities to develop and maintain our proprietary position.

As for the pharmaceutical products we develop and commercialize, as a normal course of business, we intend to pursue composition-of-matter patents, where possible, manufacturing, salts and polymorphs, dosage, combinations and formulation patents, as well as method of use patents on novel indications for known compounds. We also seek patent protection with respect to novel biological discoveries, including new targets and applications as well as adjuvant and vaccine candidates. We have also pursued patents with respect to our proprietary screening and drug development processes and technology. We have sought patent protection, either alone or jointly with our collaborators, as our collaboration agreements may dictate.

As of December 31, 2018, our patent estate, on a worldwide basis, included approximately 849 issued or allowed patents and approximately 362 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. As of December 31, 2018, there were approximately 95 issued or allowed patents and 33 patent applications pending for avacopan, our lead drug candidate in the C5aR program. As of December 31, 2018, with respect to our drug candidates in the CCR2 program, we had approximately 78 issued or allowed patents and 67 patents pending worldwide relating to their chemical composition or use thereof. As of December 31, 2018, with respect to the CCR1 and CCR9 chemokine receptors, we had approximately 495 issued or allowed patents and 124 patents pending worldwide relating to their chemical composition or use thereof. As of December 31, 2018, we had approximately 120 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs..

Avacopan, our lead drug candidate in the C5aR program, is covered by an issued patent in the United States for the composition-of-matter of avacopan and pharmaceutical compositions thereof, which will expire in 2031 (not including patent term extension that may be available to extend the term of the patent). Avacopan is also covered by an additional issued patent in the United States with an expiration date of 2029. Avacopan is covered by an issued patent in Europe (covering avacopan's composition-of-matter, compositions and certain methods of use) with an expiration date of 2029 (not including a supplementary protection certificate that may be available to extend the term of the patent). Additionally, avacopan is covered by issued patents in several jurisdictions including Australia, Canada, China, Hong Kong, India, Israel, Japan, Mexico, Singapore, South Africa, South Korea and Taiwan. These issued patents will expire in 2029 (not including patent term extensions or supplementary protection certificates that may be available in some countries). Patent applications are pending in other countries including Brazil which, if issued, are anticipated to expire in 2029 (not including patent term extensions or supplementary protection certificates that may be available). We have patent applications pending covering certain synthetic methods related to making avacopan, which, if issued, are anticipated to expire in 2035. More recent patent application filings in the avacopan family are directed to formulations and methods of use.

CCX140 is covered by three issued patents in the United States for the composition-of-matter of CCX140 and pharmaceutical compositions thereof that will expire in 2026, 2028 and 2029, respectively (not including patent term extension that may be available to extend the term of the granted patents). CCX140 is also covered by two additional issued patents in the United States (covering certain methods of use) that will expire in 2026 and 2028 respectively. CCX140 is also covered by certain issued patents in Europe (covering CCX140 composition-of-matter and certain methods of use) that will expire in 2026 and 2028, respectively (not including a supplementary protection certificate that may be available to extend the term of the patents). CCX140 is covered by certain issued patents in several jurisdictions including Australia, Canada, China, Hong Kong, India, Israel, Japan, Mexico, South Africa and South Korea, covering CCX140 composition-of-matter. These issued patents will expire in 2028 (not including patent term extensions or supplementary protection certificates that may be available in some countries). We have patent applications pending covering certain methods of use, which, if issued, are anticipated to expire in 2037.

CCX872 is covered by two issued patents in the United States for the composition-of-matter of CCX872 and pharmaceutical compositions thereof that will expire in 2026, respectively (not including patent term extension that may be available to extend the term of the granted patents). We have patent applications pending covering certain methods of use, which, if issued, are anticipated to expire in 2037.

Nonetheless, the actual protection afforded by a patent varies on a product by product basis, from country to country and depends upon many factors, including the type of patent, the scope of its coverage, the availability of regulatory related extensions, the availability of legal remedies in a particular country and the validity and enforceability of the patent.

In addition to patents, we rely upon unpatented trade secrets and know-how and continuing technological innovation to develop and maintain our competitive position. We seek to protect our proprietary information, in part, using confidentiality agreements with our commercial partners, collaborators, employees and consultants and invention assignment agreements with our employees. We also have confidentiality agreements or invention assignment agreements with our commercial partners and selected consultants. These agreements are designed to protect our proprietary information and, in the case of the invention assignment agreements, to grant us ownership of technologies that are developed through a relationship with a third party.

Our commercial success will also depend in part on not infringing upon the proprietary rights of third parties. It is uncertain whether the issuance of any third party patent would require us to alter our development or commercial strategies, or our drugs or processes, obtain licenses or cease certain activities. Our breach of any license agreements or failure to obtain a license to proprietary rights that we may require to develop or commercialize our future drugs may have a material adverse impact on us. If third parties prepare and file patent applications in the United States that also claim technology to which we have rights, we may have to participate in interference proceedings in the U.S. Patent and Trademark Office, or USPTO, to determine priority of invention.

Competition

We compete in the pharmaceutical, biotechnology and other related markets that address AAV, HS, C3G, FSGS and other renal diseases, rheumatoid arthritis, psoriasis, other autoimmune diseases and inflammatory disorders, and cancer. We face significant competition from many pharmaceutical and biotechnology companies that are also researching and selling products designed to address these markets. Many of our competitors have materially greater financial, manufacturing, marketing, research, and drug development resources than we do. Large pharmaceutical companies in particular have extensive expertise in preclinical and clinical testing and in obtaining regulatory approvals for drugs. In addition, academic institutions, government agencies, and other public and private organizations conducting research may seek patent protection with respect to potentially competitive products or technologies. These organizations may also establish exclusive collaborative or licensing relationships with our competitors.

It is possible that our competitors will develop and market drugs that are less expensive and more effective than our drug candidates, or that will render our drug candidates obsolete. It is also possible that our competitors will commercialize competing drugs before we or our partners can launch any drugs developed from our drug candidates.

Avacopan, our C5aR inhibitor, if approved for marketing by the FDA or other regulatory agencies for the treatment of AAV, might compete with current treatments, such as steroids, CYC, RTX, azathioprine, methotrexate, and mycophenolate mofetil. If avacopan were approved for the treatment of aHUS, it would potentially compete with eculizumab (Soliris®). If Avacopan is approved for marketing by the FDA or other regulatory agencies for the treatment of C3G, avacopan might compete with treatments that are in development. If avacopan were approved for the treatment of HS, it would potentially compete with adalimumab (Humira®) or other TNF-alpha antibodies which physician sometimes prescribe off-label for the treatment of HS.

CCX140, our first CCR2 inhibitor, if approved for marketing by the FDA or other regulatory agencies for the treatment of FSGS, might compete with treatments commonly used for type 2 diabetes and hypertension patients, RAAS inhibitors, are commonly prescribed treatments used to reduce blood pressure and preserve kidney function, reducing the progression of the disease. Other commercially available treatment options for FSGS include steroids, RAAS inhibitors, cyclosporine, mycophenolate mofetil (Cellcept®, Myortic®), tacrolimus (Prograf®), CYC (Cytoxan®), RTX, ACTHAR®, sirolimus (Rapamycin®, Rapamune®), and Liposorber® LA-15 System. If CCX140 is approved for marketing by the FDA or other regulatory agencies for the treatment of FSGS, CCX140 might compete with treatments that are in development.

CCX872, our second CCR2 inhibitor, if approved by the FDA or other regulatory agencies for the treatment of pancreatic cancer, might compete with treatments that are currently available, such as chemotherapeutic drugs including gemcitabine and nab-paclitaxel, or new treatments in development.

Many of these currently approved treatments have notable and common adverse events including liver and bone marrow toxicity, renal toxicity, pneumonitis, immunosuppression, allergic reactions, autoimmune diseases and infections.

We expect that competition among any of our drugs approved for sale will be based on various factors, including drug safety and efficacy, prevalence of negative side effects, reliability, ease of administration, availability, price, insurance coverage and reimbursement status and patent position. We believe that our ability to compete depends largely upon our ability to research, develop and commercialize our existing and future drug candidates. Further, we need to continue to attract and retain qualified personnel, obtain patent protection, develop proprietary technology or processes and secure sufficient capital resources for the substantial time period between technological conception and commercial sales of drugs. Our ability to compete will also be affected by the speed at which we are able to identify and develop, conduct clinical testing and obtain regulatory approvals of our drug candidates. Potential competitors may develop treatments that are more effective and/or safer than our drug candidates or that would make our technology and drug candidates obsolete or non-competitive.

Established pharmaceutical companies that currently sell or are developing drugs in our markets of interest include, but are not limited to, AbbVie, Alexion, Amgen, AstraZeneca, Biogen, Bayer, Elan, GlaxoSmithKline, Johnson & Johnson, Mallinckrodt, Merck, Merck Serono, Novartis, Pfizer, Retrophin, Roche/Genentech, Sanofi, Takeda and Teva. In addition, in some instances we may face competition from companies that sell generic versions of approved drugs that are part of the current standard of care. Many or all of these established competitors are also involved in research and drug development regarding various chemokine receptors. Pharmaceutical and biotechnology companies which are known to be involved in chemokine and chemoattractant research and related drug development include, but are not limited to, Pfizer, GlaxoSmithKline, Bristol-Myers Squibb, Merck, Takeda, Sanofi, Incyte, Achillion, Alexion, Allergan, Appellis, Omeros, InflaRX, Dimerix, X4 Pharmaceuticals, Mitsubishi Tanabe, Biolinerx, Akari Therapeutics and UCB Pharma, among others. These companies and others also compete with us in recruiting and retaining qualified scientific and management personnel, and in acquiring technologies complementary to, or necessary for, our programs.

Manufacturing

Our current drug candidates are manufactured using commonly used chemical synthetic and engineering processes using readily available or made to order raw materials. We rely on contract manufacturing organizations to produce our drug candidates in accordance with the FDA's current good manufacturing practices, or cGMP, for use in our clinical trials. We currently rely on a single source supplier for our active pharmaceutical ingredient, or API, manufacturing requirements for each of our drug candidates and for the manufacturing of drug product. The manufacture of pharmaceutical products is subject to extensive cGMP regulations, which impose various procedural and documentation requirements and govern all areas of record keeping, production processes and controls, personnel and quality control. We expect to continue to rely on contract manufacturers for the manufacture of clinical and commercial supplies of our compounds.

We purchase quantities of our drug candidates from our contract manufacturers pursuant to purchase orders that we place with them. If we were unable to obtain sufficient quantities of drug supply or receive raw materials in a timely manner, or secure the manufacturing and release of drug product by the contract manufacturer, we could be required to delay our ongoing clinical trials as we seek, engage and enable alternative manufacturers, which would be costly and time-consuming.

Government Regulation

The FDA and comparable regulatory authorities in state and local jurisdictions and in other countries impose substantial and burdensome requirements upon companies involved in the clinical development, manufacture, marketing and distribution of drugs. These agencies and other federal, state and local entities regulate research and development activities and the testing, manufacture, quality control, safety, effectiveness, labeling, storage, record keeping, approval, advertising and promotion, export and import of our drug candidates.

In the United States, the FDA regulates drug products under the Federal Food, Drug, and Cosmetic Act and the FDA's implementing regulations. If we fail to comply with applicable FDA or other requirements at any time during the drug development process, clinical testing, the approval process or after approval, we may become subject to administrative or judicial sanctions. These sanctions could include the FDA's refusal to approve pending applications, license suspension or revocation, withdrawal of an approval, warning letters, product recalls, product seizures, total or partial suspension of production or distribution, injunctions, fines, civil penalties or criminal prosecution. Any FDA enforcement action could have a material adverse effect on us. The process required by the FDA before our drug candidates may be marketed in the United States generally involves the following:

- completion of extensive preclinical laboratory tests, preclinical animal studies and formulation studies all performed in accordance with the FDA's good laboratory practices, or GLP, regulations;
- submission to the FDA of an investigational new drug, or IND, application which must become effective before human clinical trials in the United States may begin;
- performance of adequate and well-controlled human clinical trials to establish the safety and efficacy of the drug candidate for each proposed indication;
- submission to the FDA of a new drug application, or NDA;
- satisfactory completion of an FDA inspection of the manufacturing facility or facilities at which the drug is produced to assess compliance with cGMP regulations; and
- FDA review and approval of the NDA prior to any commercial marketing, sale or shipment of the drug.

The testing and approval process requires substantial time, effort and financial resources, and we cannot be certain that any approvals for our drug candidates will be granted on a timely basis, if at all.

Once a pharmaceutical drug candidate is identified for development, it enters the preclinical testing stage. Preclinical studies include laboratory evaluations of drug chemistry, formulation and stability, as well as studies

to evaluate toxicity in animals. The results of the preclinical studies, together with manufacturing information and analytical data, are submitted to the FDA as part of an IND application. The IND automatically becomes effective 30 days after receipt by the FDA, unless the FDA, within the 30-day time period, raises concerns or questions about the conduct of the clinical trial, including concerns that human research subjects will be exposed to unreasonable health risks. In such a case, the IND sponsor and the FDA must resolve any outstanding concerns before the clinical trial can begin. Submission of an IND may result in the FDA not allowing the clinical trials to commence or not allowing the clinical trials to commence on the terms originally specified in the IND. A separate submission to an existing IND must also be made for each successive clinical trial conducted during drug development, and the FDA must grant permission, either explicitly or implicitly by not objecting, before each clinical trial can begin.

Clinical trials involve the administration of the investigational drug to human subjects under the supervision of qualified investigators. Clinical trials are conducted under protocols detailing, among other things, the objectives of the clinical trial, the parameters to be used in monitoring safety and the effectiveness criteria to be used. Each protocol must be submitted to the FDA as part of the IND. An independent institutional review board, or IRB, for each medical center proposing to conduct a clinical trial must also review and approve a plan for any clinical trial before it can begin at that center and the IRB must monitor the clinical trial until it is completed. The FDA, the IRB, or the sponsor may suspend or discontinue a clinical trial at any time on various grounds, including a finding that the subjects are being exposed to an unacceptable health risk. Clinical testing also must satisfy extensive Good Clinical Practice, or GCP, requirements, including the requirements for informed consent.

All clinical research performed in the United States in support of an NDA must be authorized in advance by the FDA under the IND regulations and procedures described above. However, a sponsor who wishes to conduct a clinical trial outside the United States may, but need not, obtain FDA authorization to conduct the clinical trial under an IND. If a foreign clinical trial is not conducted under an IND, the sponsor may submit data from the clinical trial to the FDA in support of an NDA so long as the clinical trial is conducted in accordance with GCP, and so long as the FDA is able to validate the data from the study through an onsite inspection if necessary. We conducted our PROTECT-1 clinical trial solely at foreign clinical research sites, and we did not have authorization from the FDA under an IND to conduct that clinical trial in the United States. We designed the clinical trial to comply with FDA regulatory requirements for the use of foreign clinical data in support of an NDA. We are pursuing a similar development strategy for CCX140 which has completed a Phase II clinical trial in patients with DN in Europe. One of the Phase II clinical trials with avacopan in patients with AAV has also been conducted in Europe. The second clinical trial with avacopan was conducted in North America. We have open INDs in the United States for avacopan, CCX140, and CCX872. All of our clinical trials are designed to comply with FDA regulatory requirements so that the data from all trials can be used to support a regulatory filing in the United States. We are including the United States, Europe, Japan, Australia, and New Zealand in our Phase III study of avacopan in AAV. Other planned studies with avacopan and CCX140 will likely include the United States and Europe, and potentially other geographies.

Clinical Trials

For purposes of NDA submission and approval, clinical trials are typically conducted in three sequential phases, which may overlap or be combined.

- Phase I clinical trials are initially conducted in a limited population of subjects to test the drug candidate for safety, dose tolerance, absorption, metabolism, distribution and excretion in healthy humans or, on occasion, in patients with severe problems or life-threatening diseases to gain an early indication of its effectiveness.
- Phase II clinical trials are generally conducted in a limited patient population to:
    - evaluate dosage tolerance and appropriate dosage;
    - identify possible adverse effects and safety risks; and

- evaluate preliminarily the efficacy of the drug for specific targeted indications in patients with the disease or condition under study.
- Phase III clinical trials, commonly referred to as pivotal studies, are typically conducted when Phase II clinical trials demonstrate that a dose range of the drug candidate is effective and has an acceptable safety profile. Phase III clinical trials are generally undertaken with large numbers of patients, such as groups of several hundred to several thousand, to further evaluate dosage, to provide substantial evidence of clinical efficacy and to further test for safety in an expanded and diverse patient population at multiple, geographically-dispersed clinical trial sites. An exception might be drugs developed for an orphan indication, where smaller clinical trials might be acceptable to the FDA and the EMA.

In some cases, the FDA may condition approval of an NDA on the sponsor's agreement to conduct additional clinical trials to further assess the drug's safety and effectiveness after NDA approval. Such post-approval clinical trials are typically referred to as Phase IV clinical trials.

Concurrent with clinical trials, companies usually complete additional animal studies and must also develop additional information about the chemistry and physical characteristics of the drug and finalize a process for manufacturing the drug in commercial quantities in accordance with cGMP requirements. The manufacturing process must be capable of consistently producing quality batches of the drug candidate and, among other things, the manufacturer must develop methods for testing the identity, strength, quality and purity of the final drug product. Additionally, appropriate packaging must be selected and tested and stability studies must be conducted to demonstrate that the drug candidate does not undergo unacceptable deterioration over its shelf life.

New Drug Applications

The results of preclinical studies and of the clinical trials, together with other detailed information, including extensive manufacturing information and information on the composition of the drug, are submitted to the FDA in the form of an NDA requesting approval to market the drug for one or more specified indications. The FDA reviews an NDA to determine, among other things, whether a drug is safe and effective for its intended use. Under the Prescription Drug User Fee Act, or PDUFA, guidelines that are currently in effect, the FDA has a goal of ten months from the date of "filing" of a standard NDA for a new molecular entity to review and act on the submission. This review typically takes twelve months from the date the NDA is submitted to FDA because the FDA has approximately two months to make a "filing" decision.

The FDA conducts a preliminary review of all NDAs within the first 60 days after submission, before accepting them for filing, to determine whether they are sufficiently complete to permit substantive review. The FDA may request additional information rather than accept an NDA for filing. In this event, the application must be resubmitted with the additional information. The resubmitted application is also subject to review before the FDA accepts it for filing. Once the submission is accepted for filing, the FDA begins an in-depth substantive review. The FDA reviews an NDA to determine, among other things, whether the drug is safe and effective and whether the facility in which it is manufactured, processed, packaged or held meets standards designed to assure the product's continued safety, quality and purity.

Before approving an NDA, the FDA typically will inspect the facility or facilities where the product is manufactured. The FDA will not approve an application unless it determines that the manufacturing processes and facilities are in compliance with cGMP requirements and adequate to assure consistent production of the product within required specifications. Additionally, before approving an NDA, the FDA may inspect one or more clinical trial sites to assure compliance with GCP requirements.

The FDA may refer the application to an advisory committee for review, evaluation and recommendation as to whether the application should be approved. The FDA is not bound by the recommendation of an advisory

committee, but it generally follows such recommendations. The FDA may deny approval of an NDA if the applicable statutory and regulatory criteria are not satisfied, or it may require additional clinical data or an additional Phase III clinical trial. Even if such data are submitted, the FDA may ultimately decide that the NDA does not satisfy the criteria for approval. Data from clinical trials are not always conclusive and the FDA may interpret data differently than we interpret data. Even if the FDA approves a product, it may limit the approved indications for use of the product, require that contraindications, warnings or precautions be included in the product labeling, require that post-approval studies, including Phase IV clinical trials, be conducted to further assess a drug's safety after approval, require testing and surveillance programs to monitor the product after commercialization, or impose other conditions, including distribution and use restrictions or other risk management mechanisms under a risk evaluation and mitigation strategy, or REMS, which can materially affect the potential market and profitability of the product. The FDA may prevent or limit further marketing of a product based on the results of post-marketing studies or surveillance programs. After approval, some types of changes to the approved product, such as adding new indications, manufacturing changes, and additional labeling claims, are subject to further testing requirements and FDA review and approval.

Once the FDA approves an NDA, or supplement thereto, the FDA may withdraw the approval if ongoing regulatory requirements are not met or if safety problems are identified after the drug reaches the market. Where a withdrawal may not be appropriate, the FDA still may seize existing inventory of such drug or require a recall of any drug already on the market. In addition, the FDA may require testing, including Phase IV clinical trials and surveillance programs to monitor the effect of approved drugs which have been commercialized. The FDA has the authority to prevent or limit further marketing of a drug based on the results of these post-marketing programs.

Expedited Development and Review Programs

A sponsor may also seek approval of its drug candidates under programs designed to accelerate the FDA's review and approval of NDAs. For instance, a sponsor may seek FDA designation of a drug candidate as a "fast track product." Fast track products are those products intended for the treatment of a serious or life-threatening disease or condition and which demonstrate the potential to address unmet medical needs for such disease or condition. If fast track designation is obtained, the FDA may initiate review of sections of an NDA before the application is complete. This "rolling review" is available if the applicant provides and the FDA approves a schedule for submission to the FDA of the remaining information. In some cases, products studied for their safety and effectiveness in treating serious or life-threatening illnesses and that provide meaningful therapeutic benefit over existing treatments may be approved on the basis of either a surrogate endpoint that is reasonably likely to predict clinical benefit, or on a clinical endpoint that can be measured earlier than irreversible morbidity or mortality, that is reasonably likely to predict an effect on irreversible morbidity or mortality or other clinical benefit, taking into account the severity, rarity, or prevalence of the condition and the availability or lack of alternative treatments. Approvals of this kind, referred to as accelerated approvals, typically include requirements for appropriate post-approval Phase IV clinical trials to validate the surrogate endpoint or otherwise confirm the effect of the clinical endpoint.

In addition, the Food and Drug Administration Safety and Innovation Act, or FDASIA, which was enacted and signed into law in 2012, established a category of drugs referred to as "breakthrough therapies" that may be subject to accelerated approval. A sponsor may seek FDA designation of a drug candidate as a "breakthrough therapy" if the drug is intended, alone or in combination with one or more other drugs, to treat a serious or life-threatening disease or condition and preliminary clinical evidence indicates that the drug may demonstrate substantial improvement over existing therapies on one or more clinically significant endpoints, such as substantial treatment effects observed early in clinical development. Drug candidates designed to prevent, diagnose, or treat serious diseases or conditions may also be eligible for "priority review," or review within a six-month timeframe from the date a complete NDA for a new molecular entity is accepted for filing, if a sponsor shows that its drug candidate, if approved, would provide a significant improvement in safety or effectiveness over existing therapies. Fast track designation, accelerated approval, breakthrough therapy designation and

priority review do not change the standards for approval, but may expedite the development or approval process. When appropriate, we intend to seek fast track designation, accelerated approval, breakthrough therapy designation and priority review, as applicable, for our drug candidates.

Orphan Drug Designation

In the United States, under the Orphan Drug Act, the FDA may grant orphan designation to a drug or biological product intended to treat a rare disease or condition. Such diseases and conditions are those that affect fewer than 200,000 individuals in the United States, or if they affect more than 200,000 individuals in the United States, there is no reasonable expectation that the cost of developing and making a drug available in the United States for these types of diseases or conditions will be recovered from sales of the drug. Orphan drug designation must be requested before submitting an NDA. If the FDA grants orphan drug designation, the identity of the therapeutic agent and its potential orphan use are disclosed publicly by that agency. Orphan drug designation does not convey any advantage in or shorten the duration of the regulatory review and approval process, but it can lead to financial incentives, such as opportunities for grant funding toward clinical trial costs, tax advantages and user-fee waivers.

If a drug that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the drug is entitled to orphan drug marketing exclusivity for a period of seven years. Orphan drug marketing exclusivity generally prevents the FDA from approving another application, including a full NDA, to market the same drug or biological product for the same indication for seven years, except in limited circumstances, including if the FDA concludes that the later drug is safer, more effective or makes a major contribution to patient care. For purposes of small molecule drugs, the FDA defines "same drug" as a drug that contains the same active chemical entity and is intended for the same use as the drug in question. A designated orphan drug may not receive orphan drug marketing exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. Orphan drug marketing exclusivity rights in the United States may be lost if the FDA later determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantity of the drug to meet the needs of patients with the rare disease or condition.

The criteria for designating an orphan medicinal product in the European Union, or EU, are similar in principle to those in the United States. Under Article 3 of Regulation (EC) 141/2000, a medicinal product may be designated as orphan if (i) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (ii) either (a) such condition affects no more than five in 10,000 persons in the EU when the application is made, or (b) the product, without the benefits derived from orphan status, would not generate sufficient return in the EU to justify investment; and (iii) there exists no satisfactory method of diagnosis, prevention or treatment of such condition authorized for marketing in the EU, or if such a method exists, the product will be of significant benefit to those affected by the condition, as defined in Regulation (EC) 847/2000. Orphan medicinal products are eligible for financial incentives such as reduction of fees or fee waivers and are, upon grant of a marketing authorization, entitled to ten years of market exclusivity for the approved therapeutic indication. The application for orphan designation must be submitted before the application for marketing authorization. The applicant will receive a fee reduction for the marketing authorization application if the orphan designation has been granted, but not if the designation is still pending at the time the marketing authorization is submitted.

The ten-year market exclusivity in the EU may be reduced to six years if, at the end of the fifth year, it is established that the product no longer meets the criteria for orphan designation, for example, if the product is sufficiently profitable not to justify maintenance of market exclusivity. Additionally, marketing authorization may be granted to a similar product for the same indication at any time if:

- the second applicant can establish that its product, although similar, is safer, more effective or otherwise clinically superior;

- the applicant consents to a second orphan medicinal product application; or
- the applicant cannot supply enough orphan medicinal product.

Post-Approval Requirements

Drugs manufactured or distributed pursuant to FDA approvals are subject to pervasive and continuing regulation by the FDA, including, among other things, requirements relating to recordkeeping, periodic reporting, product sampling and distribution, advertising and promotion and reporting of adverse experiences with the product. After approval, most changes to the approved product, such as adding new indications or other labeling claims are subject to prior FDA review and approval. There also are continuing, annual program user fee requirements for any marketed products.

The FDA may impose a number of post-approval requirements as a condition of approval of an NDA. For example, the FDA may require post-marketing testing, including Phase IV clinical trials, and surveillance to further assess and monitor the product's safety and effectiveness after commercialization.

In addition, drug manufacturers and other entities involved in the manufacture and distribution of approved drugs are required to register their establishments with the FDA and state agencies, and are subject to periodic unannounced inspections by the FDA and these state agencies for compliance with cGMP requirements. Changes to the manufacturing process are strictly regulated and often require prior FDA approval before being implemented. FDA regulations also require investigation and correction of any deviations from cGMP requirements and impose reporting and documentation requirements upon the sponsor and any third-party manufacturers that the sponsor may decide to use. Accordingly, manufacturers must continue to expend time, money, and effort in the area of production and quality control to maintain cGMP compliance.

Once an approval of a drug is granted, the FDA may withdraw the approval if compliance with regulatory requirements and standards is not maintained or if problems occur after the product reaches the market. Later discovery of previously unknown problems with a product, including adverse events of unanticipated severity or frequency, or with manufacturing processes, or failure to comply with regulatory requirements, may result in mandatory revisions to the approved labeling to add new safety information; imposition of post-market studies or clinical trials to assess new safety risks; or imposition of distribution or other restrictions under a REMS program. Other potential consequences include, among other things:

- restrictions on the marketing or manufacturing of the product, complete withdrawal of the product from the market or product recalls;
- fines, warning letters or holds on post-approval clinical trials;
- refusal of the FDA to approve pending NDAs or supplements to approved NDAs, or suspension or revocation of product approvals;
- product seizure or detention, or refusal to permit the import or export of products; or
- injunctions or the imposition of civil or criminal penalties.

The FDA closely regulates the post-approval marketing and promotion of drugs, including standards and regulations for direct-to-consumer advertising, off-label promotion, industry-sponsored scientific and educational activities and promotional activities involving the Internet. A company can make only those claims relating to safety and efficacy that are approved by the FDA. Failure to comply with these requirements can result in adverse publicity, warning letters, corrective advertising and potential civil and criminal penalties. Physicians may prescribe legally available drugs for uses that are not described in the product's labeling and that differ from those tested by us and approved by the FDA. Such off-label uses are common across medical specialties. Physicians may believe that such off-label uses are the best treatment for many patients in varied circumstances. The FDA does not regulate the behavior of physicians in their choice of treatments. The FDA does, however, impose stringent restrictions on manufacturers' communications regarding off-label use.

Healthcare Reform

In March 2010, President Barack Obama signed the Patient Protection and Affordable Care Act, as amended by the Health Care and Education Reconciliation Act, collectively known as the Affordable Care Act. The Affordable Care Act substantially changes the way healthcare is financed by both governmental and private insurers, and significantly impacts the pharmaceutical industry. The Affordable Care Act contained a number of provisions, including those governing enrollment in federal healthcare programs, reimbursement changes and fraud and abuse, which have impacted existing government healthcare programs and resulted in the development of new programs, including Medicare payment for performance initiatives and improvements to the physician quality reporting system and feedback program. Additionally, the Affordable Care Act:

- imposed a non-deductible annual fee on pharmaceutical manufacturers or importers who sell "branded prescription drugs" to specified federal government programs;
- increased the minimum level of Medicaid rebates payable by manufacturers of brand-name drugs from 15.1% to 23.1%;
- required collection of rebates for drugs paid by Medicaid managed care organizations;
- required manufacturers to participate in a coverage gap discount program, under which they must agree to offer 50% point-of-sale discounts off negotiated prices of applicable brand drugs to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient drugs to be covered under Medicare Part D;
- mandated a further shift in the burden of Medicaid payments to the states;
- created the Independent Payment Advisory Board, which, once empaneled, will have authority to recommend certain changes to the Medicare program that could result in reduced payments for prescription drugs; and
- established a Center for Medicare Innovation at the Department of Health and Human Services Centers for Medicare and Medicaid Services, or CMS, to test innovative payment and service delivery models to lower Medicare and Medicaid spending.

We expect that the current presidential administration and U.S. Congress will seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Since taking office, President Trump has continued to support the repeal of all or portions of the Affordable Care Act. There have been judicial and Congressional challenges to certain aspects of the Affordable Care Act, and we expect additional challenges and amendments in the future. Moreover, the Trump Administration and the U.S. Congress may take further action regarding the Affordable Care Act, including, but not limited to, repeal or replacement. Most recently, the Tax Cuts and Jobs Act of 2017, or the Jobs Act, was enacted, which, among other things, removes penalties for not complying with the individual mandate to carry health insurance. There is still uncertainty with respect to the impact President Trump's administration and the U.S. Congress may have, if any, and any changes will likely take time to unfold, and could have an impact on coverage and reimbursement for healthcare items and services covered by plans that were authorized by the Affordable Care Act. However, we cannot predict the ultimate content, timing or effect of any healthcare reform legislation or the impact of potential legislation on us.

Other legislative changes have been proposed and adopted since the Affordable Care Act was enacted. These changes include aggregate reductions to Medicare payments to providers of 2% per fiscal year, which went into effect in April 2013 and, due to subsequent legislative amendments, will remain in effect through 2025 unless additional Congressional action is taken. In January 2013, American Taxpayer Relief Act of 2012, or the ATRA, was enacted, which, among other things, further reduced Medicare payments to several providers, including hospitals, imaging centers and cancer treatment centers, and increased the statute of limitations period for the government to recover overpayments to providers from three to five years. Recently, there has also been heightened governmental scrutiny over the manner in which manufacturers set prices for their marketed products, which has resulted in several Congressional inquiries and proposed bills designed to, among other things, bring

more transparency to product pricing, review the relationship between pricing and manufacturer patient programs and reform government program reimbursement methodologies for drug products. The full impact on our business of the Affordable Care Act and other new laws is uncertain. Nor is it clear whether other legislative changes will be adopted, if any, or how such changes would affect the demand for our drugs if commercialized.

Third-Party Payor Coverage and Reimbursement

Although none of our drug candidates has been commercialized for any indication, if they are approved for marketing, commercial success of our drug candidates will depend, in part, upon the availability of coverage and reimbursement from third-party payors at the federal, state, and private levels. Government payor programs, including Medicare and Medicaid, private health care insurance companies, and managed-care plans have attempted to control costs by limiting coverage and the amount of reimbursement for particular procedures or drug treatments. The U.S. Congress and state legislatures from time to time propose and adopt initiatives aimed at cost-containment. Ongoing federal and state government initiatives directed at lowering the total cost of health care will likely continue to focus on health care reform, the cost of prescription pharmaceuticals and on the reform of the Medicare and Medicaid payment systems. Examples of how limits on drug coverage and reimbursement in the United States may cause reduced payments for drugs in the future include:

- changing Medicare reimbursement methodologies;
- fluctuating decisions on which drugs to include in formularies;
- revising drug rebate calculations under the Medicaid program; and
- reforming drug importation laws.

Some third-party payors also require pre-approval of coverage for new or innovative devices or drug therapies before they will reimburse health care providers who use such therapies. While we cannot predict whether any proposed cost-containment measures will be adopted or otherwise implemented in the future, the announcement or adoption of these proposals could have a material adverse effect on our ability to obtain adequate prices for our drug candidates and operate profitably.

Other Healthcare Laws and Regulations

We are also subject to healthcare regulation and enforcement by the federal government and the states and foreign governments in which we conduct our business. The laws that may affect our ability to operate include:

- the federal Anti-Kickback Statute, which prohibits, among other things, persons from knowingly and willfully soliciting, receiving, offering or paying remuneration, directly or indirectly, in exchange for or to induce either the referral of an individual for, or the purchase, order or recommendation of, any good or service for which payment may be made under federal healthcare programs such as the Medicare and Medicaid programs. A person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. In addition, the government may assert that a claim including items or services resulting from a violation of the federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the False Claims Act. Violations of the federal Anti-Kickback Statute may result in civil monetary penalties up to $100,000 for each violation, plus up to three times the remuneration involved. Civil penalties for such conduct can further be assessed under the federal False Claims Act. Violations can also result in criminal penalties, including criminal fines of up to $100,000 and imprisonment of up to ten years. Similarly, violations can result in exclusion from participation in government healthcare programs, including Medicare and Medicaid;
- federal false claims laws which prohibit, among other things, individuals or entities from knowingly presenting, or causing to be presented, claims for payment from Medicare, Medicaid, or other federal healthcare programs that are false or fraudulent. Private individuals can bring False Claims Act “qui

tam" actions, on behalf of the government and such individuals, commonly known as "whistleblowers," may share in amounts paid by the entity to the government in fines or settlement. When an entity is determined to have violated the federal civil False Claims Act, the government may impose civil fines and penalties ranging from $11,181 to $22,363 for each false claim, plus treble damages, and exclude the entity from participation in Medicare, Medicaid and other federal healthcare programs;

- federal criminal laws that prohibit executing a scheme to defraud any healthcare benefit program or making false statements relating to healthcare matters. Similar to the federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation;
- the federal Physician Sunshine Act, which requires certain applicable manufacturers of drugs, devices, biologics and medical supplies for which payment is available under Medicare, Medicaid or the Children's Health Insurance Program, or CHIP, to report annually to CMS, information related to payments and other transfers of value to physicians, which is defined broadly to include other healthcare providers and teaching hospitals, and applicable manufacturers and group purchasing organizations, to report annually ownership and investment interests held by physicians and their immediate family members. Applicable manufacturers are required to submit annual reports to CMS. Failure to submit required information may result in civil monetary penalties of $11,278 per failure up to an aggregate of $169,170 per year (or up to an aggregate of $1.127 million per year for "knowing failures"), for all payments, transfers of value or ownership or investment interests that are not timely, accurately, and completely reported in an annual submission, and may result in liability under other federal laws or regulations;
- the federal Health Insurance Portability and Accountability Act of 1996, or HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act, and their respective implementing regulations, which impose requirements on certain covered healthcare providers, health plans and healthcare clearinghouses as well as their business associates that perform services for them that involve individually identifiable health information, relating to the privacy, security and transmission of individually identifiable health information without appropriate authorization, including mandatory contractual terms as well as directly applicable privacy and security standards and requirements. Failure to comply with the HIPAA privacy and security standards can result in civil monetary penalties up to $57,051 per violation, not to exceed $1.71 million per calendar year for non-compliance of an identical provision, and, in certain circumstances, criminal penalties with fines up to $250,000 per violation and/or imprisonment. State attorneys general can also bring a civil action to enjoin a HIPAA violation or to obtain statutory damages on behalf of residents of his or her state; and
- state law equivalents of each of the above federal laws, such as anti-kickback and false claims laws which may apply to items or services reimbursed by any third-party payor, including commercial insurers; state laws that require pharmaceutical companies to comply with the industry's voluntary compliance guidelines and the applicable compliance guidance promulgated by the federal government, or otherwise restrict payments that may be made to healthcare providers and other potential referral sources; state laws that require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers or marketing expenditures; and state laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and may not have the same effect, thus complicating compliance efforts.

International Regulation

In addition to regulations in the United States, we will be subject to a variety of foreign regulations governing clinical trials and commercial sales and distribution of our future drugs. Whether or not we obtain

FDA approval for a drug, we must obtain approval of a drug by the comparable regulatory authorities of foreign countries before we can commence clinical trials or marketing of the drug in those countries. The approval process varies from country to country, and the time may be longer or shorter than that required for FDA approval. The requirements governing the conduct of clinical trials, product licensing, pricing and reimbursement vary greatly from country to country.

Under the European Economic Area, or EEA (which is comprised of the 28 member states of the European Union plus Norway, Iceland and Liechtenstein), regulatory systems, marketing authorizations may be submitted either under the Centralized, Mutual Recognition, Decentralized or national EEA member state procedures. The Centralized Procedure provides for the grant of a single marking authorization that is valid for all member states of the EEA. The Mutual Recognition Procedure provides for mutual recognition of national approval decisions. Under this procedure, the holder of a national marking authorization may submit an application to the remaining Member States. Under the Decentralized Procedure, if the product has not received a marketing authorization in any EEA member state at the time of application, the applicant can file an application to various EEA member states (choosing once as the so-called reference member states) of its choice which will be reviewed and approved simultaneously by them.

In addition to regulations in Europe and the United States, we will be subject to a variety of foreign regulations governing clinical trials and commercial distribution of our future drugs.

Employees

As of December 31, 2018, we had 76 full-time employees, 31 of whom hold Ph.D.s, M.D.s or both. Of our total workforce, 58 employees are engaged in research and development, and 18 employees are engaged in business development, finance, legal, human resources, facilities, information technology administration and general management. We have no collective bargaining agreements with our employees and we have not experienced any work stoppages. We believe that our relations with our employees are good.

About ChemoCentryx

We commenced operations in 1997. Our principal offices are located at 850 Maude Avenue, Mountain View, California 94043, and our telephone number is (650) 210-2900. Our website address is www.chemocentryx.com. The information contained in, or that can be accessed through, our website is not part of this Annual Report on Form 10-K. We have two wholly owned inactive subsidiaries, ChemoCentryx Limited, organized under the laws of the United Kingdom and ChemoCentryx Ireland Limited, organized under the laws of Ireland.

Available Information

We file electronically with the Securities and Exchange Commission, or SEC, our annual reports on Form 10-K, quarterly reports on Form 10-Q and current reports on Form 8-K pursuant to Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended. We make available on our website at www.chemocentryx.com, free of charge, copies of these reports, as soon as reasonably practicable after we electronically file such material with, or furnish it to, the SEC. The SEC maintains a website that contains reports, proxy and information statements, and other information regarding issuers that file electronically with the SEC. The address of that website is www.sec.gov. The information in or accessible through the SEC and our website are not incorporated into, and are not considered part of, this filing. Further, our references to the URLs for these websites are intended to be inactive textual references only.

Item 1A. Risk Factors.

The following section includes the most significant factors that may adversely affect our business and operations. You should carefully consider the risks and uncertainties described below and all information contained in this Annual Report on Form 10-K before deciding to invest in our common stock. If any of the following risks actually occur, our business, financial condition, results of operations and growth prospects would likely be materially and adversely affected. In that event, the trading price of our common stock could decline, and you could lose all or part of your investment.

Risks Related to Our Business

We anticipate that we will continue to incur significant losses for the foreseeable future, and if we are unable to achieve and sustain profitability, the market value of our common stock will likely decline.

We are a clinical-stage biopharmaceutical company. We do not currently have any products approved for sale, and we continue to incur significant research and development and general and administrative expenses related to our operations. Our net income (loss) for the years ended December 31, 2018, 2017 and 2016 was $(38.0) million, $17.9 million and $(40.0) million, respectively. As of December 31, 2018, we had an accumulated deficit of $374.5 million. We expect to continue to incur significant losses for the foreseeable future. We expect these losses and our cash utilization to increase in the near term as we continue to conduct clinical trials for CCX140, avacopan, and CCX872 and conduct research and development of our other drug candidates. To date, we have derived all of our revenues from upfront fees and milestone payments, other payments pursuant to our collaboration agreements and government grants and contracts for research and development. For example, in May 2016 and December 2016, we entered into collaboration and license agreements with Vifor (International) Ltd. and/or its affiliates, or collectively, Vifor, for the commercialization of avacopan and CCX140, respectively. We do not anticipate that we will generate revenue from the sale of our products for the foreseeable future. In addition, if approved, we expect to incur significant costs to commercialize our drug candidates and our drugs may never gain market acceptance. If our drug candidates fail to demonstrate safety and efficacy in clinical trials, do not gain regulatory approval, or do not achieve market acceptance, we may never become profitable. Even if we achieve profitability in the future, we may not be able to sustain profitability in subsequent periods. If we are unable to achieve and sustain profitability, the market value of our common stock will likely decline. Because of the numerous risks and uncertainties associated with developing pharmaceutical products, we are unable to predict the extent of any future losses or whether we will become profitable.

The development of new drugs is a highly risky undertaking which involves a lengthy process, and our drug discovery and development activities therefore may not result in products that are approved for marketing and sale by the applicable regulatory authorities on the time schedule we have planned, or at all, or result in substantial payments to us.

Our drug candidates are in the early stages of drug discovery or clinical trials and are prone to the risks of failure inherent in drug development. As of the date of this Annual Report on Form 10-K, nine of our drug candidates have been tested in human beings. We will need to conduct significant additional preclinical studies and clinical trials before we can demonstrate that any of our drug candidates is safe and effective to the satisfaction of the FDA, the EMA and other regulatory authorities. Preclinical studies and clinical trials are expensive and uncertain processes that take years to complete. For example, we incurred significant expenses related to the IND filing and the completed single ascending dose Phase I clinical trial for CCX915, our first generation CCR2 drug candidate, which did not advance into Phase II clinical trials because its pharmacokinetic, or PK, properties in humans did not meet our expectations. Failure can occur at any stage of the process, and we cannot assure you that any of our drug candidates will demonstrate safety and efficacy in clinical trials or result in commercially successful products. For instance, we filed a CMA application for avacopan in the treatment of patients with AAV with the EMA, which was validated by the EMA in December 2017. However, in light of the upcoming availability of data from the pivotal Phase III ADVOCATE trial, we decided to withdraw the CMA

application based on Phase II data. While we intend to file integrated regulatory submissions in 2020 with the EMA and FDA for full (unconditional) marketing approval after the planned release of topline data from the Phase III ADVOCATE trial anticipated in the fourth quarter of 2019, we can provide no assurance that we will receive such approval.

We cannot assure you that our ongoing clinical trials or any future clinical trial of any of our other drug candidates will be completed on schedule, or at all, or whether our planned clinical trials will start in a timely manner. The commencement of our planned clinical trials could be substantially delayed or prevented by a number of factors, including:

- delays or failures in obtaining sufficient quantities of the active pharmaceutical ingredient, or API, and/or drug product;
- delays or failures in reaching agreement on acceptable clinical trial agreement terms or clinical trial protocols with prospective sites;
- delays or failures in obtaining institutional review board, or IRB, or ethics committee approval to conduct a clinical trial at a prospective site;
- the need to successfully complete, on a timely basis, preclinical safety pharmacology or toxicology studies;
- the limited number of, and competition for, suitable sites to conduct the clinical trials;
- the limited number of, and competition for, suitable patients for enrollment in the clinical trials; and
- delays or failures in obtaining regulatory approval to commence a clinical trial.

The completion of our clinical trials could also be substantially delayed or prevented by a number of factors, including:

- slower than expected rates of patient recruitment and enrollment;
- failure of patients to complete the clinical trials;
- failure of our third party vendors to timely or adequately perform their contractual obligations relating to the clinical trials;
- inability or unwillingness of patients or medical investigators to follow our clinical trial protocols;
- inability to monitor patients adequately during or after treatment;
- termination of the clinical trials by one or more clinical trial sites;
- unforeseen safety issues;
- lack of efficacy demonstrated during clinical trials;
- lack of adequate funding to continue the clinical trials;
- the need for unexpected discussions with the FDA, EMA or other foreign regulatory agencies regarding the scope or design of our clinical trials or the need to conduct additional trials;
- unforeseen delays by the FDA, EMA or other foreign regulatory agencies after submission of our results;
- an unfavorable FDA or EMA inspection of our contract manufacturers of API or drug product; and
- inspection of the clinical investigation records, operations or trial sites by the FDA or other regulatory authorities resulting in the imposition of a clinical hold.

Any failure or significant delay in completing clinical trials for our drug candidates would harm the commercial prospects for our drug candidates and adversely affect our financial results.

Additionally, changes in regulatory requirements and guidance may occur and we may need to amend clinical trial protocols to reflect these changes. Amendments may require us to resubmit our clinical trial protocols to regulatory agencies and ethics committees for reexamination, which may impact the costs, timing or successful completion of a clinical trial. If we experience delays in completion of, or if we terminate, any of our clinical trials, the commercial prospects for our drug candidates may be harmed and our ability to generate product revenues will be delayed. In addition, many of the factors that cause, or lead to, a delay in the commencement or completion of clinical trials may also ultimately lead to the denial of regulatory approval of a drug candidate.

If we are required to suspend or discontinue clinical trials due to side effects or other safety risks, or if we are required to conduct studies on the long-term effects associated with the use of our drug candidates, our efforts to commercialize our products could be delayed or halted.

Our clinical trials may be suspended or terminated at any time for a number of safety-related reasons. For example, we may voluntarily suspend or terminate our clinical trials if at any time we believe that our drug candidates present an unacceptable safety risk to the clinical trial patients. In addition, IRBs or regulatory agencies may order the temporary discontinuation or termination of our clinical trials at any time if they believe that the clinical trials are not being conducted in accordance with applicable regulatory requirements, including if they present an unacceptable safety risk to patients. Administering any drug candidate to humans may produce undesirable side effects. The existence of undesirable side effects resulting from our drug candidates could cause us or regulatory authorities, such as the FDA, to interrupt, delay or halt clinical trials of our drug candidates and could result in the FDA or other regulatory agencies denying further development or approval of our drug candidates for any or all targeted indications.

Further, chemokine receptors and chemoattractant receptors are a novel class of targets. As a result, we may experience unforeseen adverse side effects with our existing and future drug candidates, including avacopan and CCX140. As of the date of this Annual Report on Form 10-K, nine of our drug candidates have been tested in human beings. Although we have not observed significant harmful side effects in prior studies of our drug candidates, later trials could reveal such side effects. The PK profile of preclinical studies may not be indicative of results in any clinical trial. For example, prior to commencing our preclinical studies of our CCX140 drug candidate, we studied another drug candidate that targeted CCR2, which we abandoned after PK results were not as favorable in humans as in earlier preclinical animal studies. We have not completed studies on the long-term effects associated with the use of our drug candidates. Completion of studies of these long-term effects may be required for regulatory approval and would delay our introduction of our drug candidates into the market. These studies could also be required at any time after regulatory approval of any of our drug candidates. Absence of long-term data may also limit the approved uses of our products, if any, to short-term use. Some or all of our drug candidates may prove to be unsafe for human use.

Undesirable side effects caused by our drug candidates could cause us or regulatory authorities to interrupt, delay or halt clinical trials and could result in a more restrictive label or the delay or denial of regulatory approval by the FDA, EMA or other comparable regulatory authorities. Drug-related side effects could affect patient recruitment or the ability of enrolled patients to complete a trial or result in potential product liability claims. Any of these occurrences may harm our business, financial condition and prospects significantly. Given the serious nature of the conditions we are treating in our clinical trials, and the multiple concomitant medications including our active drug candidates that our patients are treated with, side effects (such as nausea, diarrhea, infections, hepatic enzyme elevations, and possible allergic reactions) have been reported in our ongoing blinded clinical studies. While such disorders may be found to be not related to our drug candidates, such events may create a negative safety perception. Even if any of our drug candidates receives marketing approval, as greater numbers of patients use a drug following its approval, an increase in the incidence or severity of side effects or the incidence of other post-approval problems that were not seen or anticipated during pre-approval clinical trials could result in a number of potentially significant negative consequences, including that regulatory authorities may withdraw their approval of the product, regulatory authorities may require the addition of labeling

statements, such as "black box" warnings or contraindications, or impose additional safety monitoring or reporting requirements, we may be required to change the way the product is administered or conduct additional clinical trials, we may be required to implement a REMS or create a medication guide outlining the risks of such side effects for distribution to patients, we could be sued and held liable for harm caused to patients, and our reputation may suffer. Any of these events could substantially increase the costs and expenses of developing, commercializing and marketing any such drug candidates or could harm or prevent sales of any approved products.

Even if our drug candidates do obtain regulatory approval they may never achieve market acceptance or commercial success.

Even if we obtain FDA or other regulatory approvals, our drug candidates may not achieve market acceptance among physicians, patients and third-party payors and, ultimately, may not be commercially successful. Market acceptance of our drug candidates for which we receive approval depends on a number of factors, including:

- the efficacy and safety as demonstrated in clinical trials;
- the clinical indications for which the drug is approved;
- acceptance by physicians, major operators of clinics and patients of the drug as a safe and effective treatment;
- the potential and perceived advantages of our drug candidates over alternative treatments;
- the safety of drug candidates seen in a broader patient group, including its use outside the approved indications;
- the cost of treatment in relation to alternative treatments;
- the availability of adequate reimbursement and pricing by third parties and government authorities;
- relative convenience and ease of administration;
- the prevalence and severity of adverse side effects; and
- the effectiveness of our sales and marketing efforts.

Any failure by our drug candidates that obtain regulatory approval to achieve market acceptance or commercial success would adversely affect our financial results.

The commercial success of our drug candidates depends on our ability to develop and market such drug candidates or co-develop and commercialize such drug candidates, and if we fail in these initiatives, our ability to generate future revenue could be significantly reduced.

We may retain commercialization rights to certain of our drug candidates or find partners for their co-development and commercialization. Any of the following events or factors could have a material adverse effect on our ability to generate revenue from the commercialization of our drug candidates:

- We may be unable to successfully complete the clinical development of our drug candidates;
- Our lack of experience in commercializing and marketing drug products;
- We may not have or be able to obtain sufficient financial resources to develop and commercialize our drug candidates;

- We may not be able to identify a suitable co-development partner;
- We, our partners or any of our future partners may fail to fulfill responsibilities in a timely manner or fail to commit sufficient resources to the development, regulatory approval, and commercialization efforts related to our drug candidates;
- We, our partners or any of our future partners must comply with additional requests and recommendations from the FDA, including additional clinical trials;
- We, our partners or any of our future partners may not obtain all necessary approvals from the FDA and similar foreign regulatory agencies;
- Our drug candidates must be manufactured in compliance with requirements of the FDA and similar foreign regulatory agencies and in commercial quantities sufficient to meet market demand;
- Our drug candidates may not achieve market acceptance by physicians, patients and third-party payors;
- Our drug candidates may not compete successfully against alternative products and therapies; and
- We or any pharmaceutical company may independently develop products that compete with our drug candidates.

We rely on third parties to conduct all our preclinical studies and clinical trials. If these third parties do not successfully carry out their contractual duties or meet expected deadlines, we may be unable to obtain regulatory approval for or commercialize any of our drug candidates.

We currently do not have the ability to independently conduct preclinical studies or clinical trials. We rely on medical institutions, clinical investigators, contract laboratories, collaborative partners and other third parties, such as clinical research organizations, or CROs, to conduct clinical trials on our drug candidates. The third parties with which we contract for execution of our clinical trials play a significant role in the conduct of these trials and the subsequent collection and analysis of data. These third parties are not our employees, and except for contractual duties and obligations, we have limited ability to control the amount or timing of resources that they devote to our programs. Although we rely on these third parties to conduct our preclinical studies and clinical trials, we remain responsible for ensuring that each of our preclinical studies and clinical trials is conducted in accordance with its investigational plan and protocol. Moreover, the FDA and foreign regulatory authorities require us to comply with GCP requirements for conducting, monitoring, recording and reporting the results of clinical trials to ensure that the data and results are scientifically credible and accurate and that the trial subjects are adequately informed of the potential risks of participating in clinical trials.

In addition, the execution of preclinical studies and clinical trials, and the subsequent compilation and analysis of the data produced, requires coordination among various parties. In order for these functions to be carried out effectively and efficiently, it is imperative that these parties communicate and coordinate with one another. Moreover, these third parties may also have relationships with other commercial entities, some of which may compete with us. In most cases, these third parties may terminate their agreements with us upon 30 days prior written notice of a material breach by us that is not cured within 30 days. Many of these agreements may also be terminated by such third parties under certain other circumstances, including our insolvency or our failure to comply with applicable laws. In general, these agreements require such third parties to reasonably cooperate with us at our expense for an orderly winding down of services of such third parties under the agreements. If the third parties conducting our clinical trials do not perform their contractual duties or obligations, experience work stoppages, do not meet expected deadlines, terminate their agreements with us or need to be replaced, or if the quality or accuracy of the clinical data they obtain is compromised due to the failure to adhere to our clinical trial protocols or GCPs, or for any other reason, we may need to enter into new arrangements with alternative third parties, which could be costly, and our clinical trials may be extended, delayed or terminated or may need to be repeated, and we may not be able to obtain regulatory approval for or commercialize the drug candidate being tested in such trials.

If any of our drug candidates receives marketing approval and we or others later identify undesirable side effects caused by the drug candidate, our ability to market and derive revenue from the drugs could be compromised.

If we or others identify undesirable side effects caused by one of our drugs, any of the following adverse events could occur:

- regulatory authorities may withdraw their approval of the drug or seize the drug;
- we may be required to recall the drug or change the way the drug is administered;
- additional restrictions may be imposed on the marketing of the particular drug or the manufacturing processes;
- we may be subject to fines, injunctions or the imposition of civil or criminal penalties;
- regulatory authorities may require the addition of labeling statements, such as a "black box" warning or a contraindication;
- we may be required to create a Medication Guide outlining the risks of such side effects for distribution to patients;
- we could be sued and held liable for harm caused to patients;
- the drug may become less competitive; and
- our reputation may suffer.

Any of these could result in the loss of significant revenues, which would materially and adversely affect our results of operations and business.

We will need additional financing and may be unable to raise capital on acceptable terms, or at all, when needed, which would force us to delay, reduce or eliminate our research and development programs and other operations or commercialization efforts.

We are advancing multiple drug candidates through discovery and development and will require substantial funds to conduct development, including preclinical studies and clinical trials, of our drug candidates. Commercialization of any drug candidate will also require substantial expenditures. Our ability to develop and commercialize our drug candidates will depend upon our ability to identify financing or collaboration arrangements and there can be no assurance that we will be successful in identifying or implementing any such arrangement.

As of December 31, 2018, we had approximately $177.0 million in cash, cash equivalents and investments. Additionally, in the first quarter of 2019, we sold 1,666,367 shares of our common stock pursuant to our "at-the market" equity offering program for net proceeds of $19.4 million. We believe that our available cash, cash equivalents and investments will be sufficient to fund our anticipated level of operations for at least 12 months following our financial statement issuance date, March 11, 2019. Our future financing requirements will depend on many factors, some of which are beyond our control, including:

- the rate of progress and cost of our clinical trials, preclinical studies and other discovery and research and development activities;
- the timing of, and costs involved in, seeking and obtaining FDA and other regulatory approvals;
- the success of any strategic alliance with collaboration partners and potential future collaboration partners;
- the costs of preparing, filing, prosecuting, maintaining and enforcing any patent claims and other intellectual property rights, including litigation costs and the results of such litigation;

- our ability to enter into additional collaboration, licensing, government or other arrangements and the terms and timing of such arrangements;
- potential acquisition or in-licensing of other products or technologies; and
- the emergence of competing technologies or other adverse market developments.

Future capital requirements will also depend on the extent to which we acquire or invest in additional complementary businesses, products and technologies. We currently have no understandings, commitments or agreements relating to any of these types of transactions.

Until we can generate a sufficient amount of product revenue to finance our cash requirements, which we may never do, we expect to finance future cash needs primarily through public or private equity offerings, debt financings, our credit facility, our "at-the market" equity offering program, government grants and contracts and/or strategic collaborations. Additional financing may not be available to us when we need it or it may not be available on favorable terms, if at all. If we are unable to obtain adequate financing when needed, we may have to delay, reduce the scope of or eliminate one or more of our clinical trials or research and development programs or our commercialization efforts. We may be required to enter into collaborative partnerships for one or more of our drug candidate programs at an earlier stage of development or on less favorable terms, which may require us to relinquish rights to some of our drug candidates that we would otherwise have pursued on our own.

The terms of our credit facility place restrictions on our operating and financial flexibility.

We have entered into a loan and security agreement, or credit facility, with Hercules Capital, Inc., or Hercules, which is secured by substantially all of our assets, excluding intellectual property, pursuant to which we may borrow up to an aggregate principal amount of $50.0 million, subject to certain terms and conditions. The outstanding principal balance under the credit facility was $20.0 million at December 31, 2018.

The credit facility also includes customary affirmative and negative covenants and events of default, the occurrence and continuance of which provide Hercules with the right to demand immediate repayment of all principal and unpaid interest under the credit facility, and to exercise remedies against us and the collateral securing the credit facility. These events of default include, among other things: (i) insolvency, liquidation, bankruptcy or similar events; (ii) failure to pay any debts due under the credit facility or other loan documents on a timely basis; (iii) failure to observe any covenant or secured obligation under the credit facility, which failure, in most cases, is not cured within 15 days; (iv) occurrence of an event that could reasonably be expected to have a material adverse effect on us; (v) material misrepresentations; (vi) occurrence of any default under any other agreement to which we are a party involving indebtedness in excess of $750,000 or the occurrence of a default under any agreement to which we are a party that could reasonably be expected to have a material adverse effect on us; and (vii) certain money judgments being entered against us or if any portion of our assets are attached or seized.

Our ability to make scheduled payments on or to refinance our indebtedness depends on our future performance and ability to raise additional sources of cash, which is subject to economic, financial, competitive and other factors beyond our control. If we are unable to generate sufficient cash to service our debt, we may be required to adopt one or more alternatives, such as selling assets, restructuring our debt or obtaining additional equity capital on terms that may be onerous or highly dilutive. If we desire to refinance our indebtedness, our ability to do so will depend on the capital and lending markets and our financial condition at such time. We may not be able to engage in any of these activities or engage in these activities on desirable terms, which could result in a default on our debt obligations.

Any orphan drug designations we receive may not confer marketing exclusivity or other benefits.

In the United States, under the Orphan Drug Act, the FDA may grant orphan designation to a drug or biological product intended to treat a rare disease or condition. Such diseases and conditions are those that affect

fewer than 200,000 individuals in the United States, or if they affect more than 200,000 individuals in the United States, there is no reasonable expectation that the cost of developing and making a drug available in the United States for these types of diseases or conditions will be recovered from sales of the drug. Orphan drug designation must be requested before submitting an NDA. If the FDA grants orphan drug designation, the identity of the therapeutic agent and its potential orphan use are disclosed publicly by that agency. Orphan drug designation does not convey any advantage in or shorten the duration of the regulatory review and approval process, but it can lead to financial incentives, such as opportunities for grant funding toward clinical trial costs, tax advantages and user-fee waivers.

If a drug that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the drug is entitled to orphan drug marketing exclusivity for a period of seven years. Orphan drug marketing exclusivity generally prevents the FDA from approving another application, including a full NDA, to market the same drug or biological product for the same indication for seven years, except in limited circumstances, including if the FDA concludes that the later drug is safer, more effective or makes a major contribution to patient care. For purposes of small molecule drugs, the FDA defines "same drug" as a drug that contains the same active chemical entity and is intended for the same use as the drug in question. A designated orphan drug may not receive orphan drug marketing exclusivity if it is approved for a use that is broader than the indication for which it received orphan designation. Orphan drug marketing exclusivity rights in the United States may be lost if the FDA later determines that the request for designation was materially defective or if the manufacturer is unable to assure sufficient quantity of the drug to meet the needs of patients with the rare disease or condition.

The criteria for designating an orphan medicinal product in the EU are similar in principle to those in the United States. Under Article 3 of Regulation (EC) 141/2000, a medicinal product may be designated as orphan if (i) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (ii) either (a) such condition affects no more than five in 10,000 persons in the EU when the application is made, or (b) the product, without the benefits derived from orphan status, would not generate sufficient return in the EU to justify investment; and (iii) there exists no satisfactory method of diagnosis, prevention or treatment of such condition authorized for marketing in the EU, or if such a method exists, the product will be of significant benefit to those affected by the condition, as defined in Regulation (EC) 847/2000. Orphan medicinal products are eligible for financial incentives such as reduction of fees or fee waivers and are, upon grant of a marketing authorization, entitled to ten years of market exclusivity for the approved therapeutic indication. The application for orphan designation must be submitted before the application for marketing authorization. The applicant will receive a fee reduction for the marketing authorization application if the orphan designation has been granted, but not if the designation is still pending at the time the marketing authorization is submitted. Orphan designation does not convey any advantage in, or shorten the duration of, the regulatory review and approval process.

The ten-year market exclusivity in the EU may be reduced to six years if, at the end of the fifth year, it is established that the product no longer meets the criteria for orphan designation, for example, if the product is sufficiently profitable not to justify maintenance of market exclusivity. Additionally, marketing authorization may be granted to a similar product for the same indication at any time if:

- the second applicant can establish that its product, although similar, is safer, more effective or otherwise clinically superior;
- the applicant consents to a second orphan medicinal product application; or
- the applicant cannot supply enough orphan medicinal product.

The FDA granted orphan drug designation for avacopan for the treatment of C3G and AAV, including granulomatosis with polyangiitis or Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome. In November 2014, the European Commission granted orphan drug designation for avacopan for the

treatment of granulomatosis with polyangiitis or Wegener's granulomatosis and microscopic polyangiitis, and, in June 2017, for the treatment of C3G. However, we cannot assure you that we will be able to obtain or maintain orphan drug exclusivity for avacopan, if it is approved for the treatment of C3G and/or AAV in any jurisdiction, in a timely manner or at all, or that a competitor will not obtain orphan drug exclusivity that could block the regulatory approval of avacopan for several years. If we are unable to obtain or maintain orphan drug exclusivity in the United States or the EU, our ability to generate sufficient revenues may be negatively affected. If a competitor is able to obtain orphan drug exclusivity that would block avacopan's regulatory approval, our ability to generate revenues would be significantly reduced, which would harm our business prospects, financial condition and results of operations.

We may form additional strategic alliances in the future with respect to our programs, and we may not realize the benefits of such alliances.

We may form additional strategic alliances, create joint ventures or collaborations or enter into licensing arrangements with third parties with respect to our programs that we believe will complement or augment our existing business. For example, we entered into collaboration and license agreements with Vifor for the development and commercialization of CCX140 and avacopan. We face significant competition in seeking appropriate strategic partners or other alternative arrangements and the negotiation process is time-consuming and complex. Moreover, we may not be successful in our efforts to establish a strategic partnership or other alternative arrangements for any current or future drug candidates and programs because our research and development pipeline may be insufficient, our drug candidates and programs may be deemed to be at too early of a stage of development for collaborative effort and/or third parties may not view our drug candidates and programs as having the requisite potential to demonstrate safety and efficacy. We cannot be certain that, following a strategic transaction or license, we will achieve the revenues or specific net income that justifies such transaction. For example, Vifor has the right to terminate the Avacopan Agreement and the CCX140 Agreement at its convenience, in which case we would not receive payments under such agreements. Any delays in entering into new strategic partnership agreements related to our drug candidates could also delay the development and commercialization of our drug candidates and reduce their competitiveness even if they reach the market.

Key elements of our proprietary suite of drug discovery technologies, known as EnabaLink, including our RAM screening technology, are proprietary approaches to the discovery and development of new drug candidates and may not result in the discovery of any small molecule compounds of commercial value.

We must continue to identify and develop compounds that target the chemokine network and expand our portfolio of drug candidates. Research programs to identify new disease targets and drug candidates require substantial technical, financial and human resources. We have limited resources to study the more than 50 known chemokine ligands, as described in a February 2006 article in the New England Journal of Medicine, and approximately 25 identified chemokine receptors as described in a January 2014 publication by the nomenclature committee of the International Union of Pharmacology. Two structural biology papers published during 2016 in Nature describe crystal structures of two different chemokine receptors in complex with small molecule inhibitors and provides insight to the function and respective modulation through multiple binding pockets. We expect that this pivotal work will assist in the development of novel small inhibitors of chemokine receptors. EnabaLink represents a new approach to the development of new drug candidates and we cannot assure you that EnabaLink will result in the discovery of new drug candidates. EnabaLink has only resulted in a limited number of clinical and preclinical-stage programs to date, and we may not identify any therapeutic small molecule compounds of commercial value using EnabaLink or other commercially available drug discovery technologies.

If our Reverse Activation of Migration, or RAM, screening technology or any other screening technologies fail to identify highly specific "hits" that lead to the development of new drug candidates, our business may be materially and adversely affected. Our scientists may be unable to optimize the chemical "hits" identified by our RAM screening technology and develop the identified starting material into a candidate for further development that meets the desired product criteria. Our research and development programs may initially show promise in

identifying chemokine receptors and their impact on the body's immune system, yet fail to yield drug candidates that are suitable for preclinical and clinical development. We cannot assure you that our current efforts will be successful or that we will not abandon any of our efforts in the future related to a particular chemokine receptor or small molecule program.

We rely on third party contract manufacturing organizations to manufacture and supply our drug candidates for us. If one of our suppliers or manufacturers fails to perform adequately or fulfill our needs, we may be required to incur significant costs and devote significant efforts to find new suppliers or manufacturers. We may also face delays in the development and commercialization of our drug candidates.

We currently have limited experience in, and we do not own facilities for, manufacturing our drug candidates. We rely upon third party contract manufacturing organizations to manufacture and supply larger quantities of these other drug candidates. The manufacture of pharmaceutical products in compliance with cGMP requires significant expertise and capital investment, including the development of advanced manufacturing techniques and process controls. Manufacturers of pharmaceutical products often encounter difficulties in production, including difficulties with production costs and yields, quality control, including stability of the drug candidate and quality assurance testing, shortages of qualified personnel, as well as compliance with strictly enforced FDA cGMP requirements, other federal and state regulatory requirements, and foreign regulations. Raw materials for the synthesis of our API are sourced globally. If the manufacturers of our raw materials and pharmaceutical products were to encounter any difficulties or otherwise fail to comply with their obligations to us or under applicable regulations, our ability to provide study drugs in our preclinical studies and clinical trials would be jeopardized. Any delay or interruption in the supply of preclinical study or clinical trial materials could delay the completion of our preclinical studies and clinical trials, increase the costs associated with maintaining our preclinical study and clinical trial programs and, depending upon the period of delay, require us to commence new trials at significant additional expense or terminate the studies and trials completely.

All manufacturers of our drug candidates must comply with cGMP requirements enforced by the FDA through its facilities inspection program. These requirements include, among other things, quality control, quality assurance and the maintenance of records and documentation. Manufacturers of our component materials may be unable to comply with these cGMP requirements and with other FDA, state and foreign regulatory requirements. The FDA or similar foreign regulatory agencies at any time may also implement new standards, or change their interpretation and enforcement of existing standards for manufacture, packaging or testing of products. We have little control over our manufacturers' compliance with these regulations and standards. A failure to comply with these requirements may result in fines and civil penalties, suspension of production, suspension or delay in product approval, product seizure or recall, or withdrawal of product approval. If the safety of any product supplied is compromised due to our manufacturers' failure to adhere to applicable laws or for other reasons, we may not be able to obtain regulatory approval for or successfully commercialize our products, and we may be held liable for any injuries sustained as a result. Any of these factors could cause a delay of clinical trials, regulatory submissions, approvals or commercialization of our drug candidates or entail higher costs or impair our reputation.

We currently rely on a single source supplier for API and drug product for each of our drug candidates. In the event that we and our suppliers cannot agree to the terms and conditions for them to provide some or all of our API clinical and commercial supply needs, or if any single source supplier terminates the agreement in response to a breach by us, we would not be able to manufacture the API on a commercial scale until a qualified alternative supplier is identified, which could also delay the development of, and impair our ability to commercialize, drug candidates.

Although alternative sources of supply exist, the number of third-party suppliers with the necessary manufacturing and regulatory expertise and facilities is limited, and it could be expensive and take a significant amount of time to arrange for alternative suppliers, which could have a material adverse effect on our business. New suppliers of any API would be required to qualify under applicable regulatory requirements and would need

to have sufficient rights under applicable intellectual property laws to the method of manufacturing such ingredients. Obtaining the necessary FDA approvals or other qualifications under applicable regulatory requirements and ensuring non-infringement of third-party intellectual property rights could result in a significant interruption of supply and could require the new manufacturer to bear significant additional costs which may be passed on to us.

We do not currently have a fully established sales and marketing staff or distribution organization. If we are unable to develop a sales and marketing and distribution capability on our own or through collaborations with other marketing partners, we will not be successful in commercializing our future products.

We do not currently have fully established sales, marketing or distribution capabilities or experience. In order to market any products that may be approved by the FDA, EMA or other comparable regulatory authorities, we must build our sales, marketing, managerial and other non-technical capabilities, or make arrangements with third parties to perform these services. We have entered into the Avacopan Agreement and the CCX140 Agreement with Vifor for development and commercialization of avacopan and CCX140 outside of the United States. We retain commercialization rights to avacopan and CCX140 in the United States. To the extent we rely on third parties such as Vifor for marketing and distributing our approved products, any revenue we receive will depend upon the efforts of third parties, which may not be successful and are only partially within our control and our product revenue is likely to be lower than if we directly marketed or sold our products. Future collaborators may fail to develop or effectively commercialize our drug candidates because they cannot obtain necessary regulatory approvals, development or commercialization is not commercially reasonable, they elect to pursue competitive products outside of the collaboration, or for other reasons. If we are unable to enter into arrangements with third parties to commercialize any approved products on acceptable terms or at all, we may not be able to successfully commercialize our future products or we will have to market these products ourselves, which will be expensive and require us to build our own sales force, which we do not have experience doing. We cannot assure you we will be successful in any of these initiatives. If we are not successful in commercializing our future products, either on our own or through collaborations with third parties, any future product revenue will be materially adversely affected.

We will need to increase the size of our organization, and we may experience difficulties in managing growth.

As of December 31, 2018, we had 76 full-time employees. We will need to continue to expand our commercial, managerial, operational, financial and other resources in order to manage our operations and clinical trials, continue our development activities and commercialize our drug candidates. Our management and personnel, systems and facilities currently in place may not be adequate to support this future growth. Our need to effectively execute our growth strategy requires that we:

- build our sales, marketing and distribution capabilities;
- manage our clinical trials effectively, including our Phase III clinical trial for avacopan, which is being conducted at numerous trial sites throughout the world;
- manage our internal development efforts effectively while carrying out our contractual obligations to licensors, contractors, collaborators, government agencies and other third parties;
- continue to improve our operational, financial and management controls, reporting systems and procedures; and
- identify, recruit, maintain, motivate and integrate additional employees.

We face substantial competition and our competitors may discover, develop or commercialize products faster or more successfully than us.

The biotechnology and pharmaceutical industries are highly competitive, and we face significant competition from companies in the pharmaceutical, biotechnology and other related markets that are researching

and marketing products designed to address autoimmune diseases, inflammatory disorders, and cancer. Established pharmaceutical companies that currently sell or are developing drugs in our markets of interest include, but are not limited to, AbbVie, Alexion, Amgen, AstraZeneca, Biogen, Bayer, Elan, GlaxoSmithKline, Johnson & Johnson, Mallinckrodt, Merck, Merck Serono, Novartis, Pfizer, Retrophin, Roche/Genentech, Sanofi, Takeda and Teva. In addition, in some instances we may face competition from companies that sell generic versions of approved drugs that are part of the current standard of care. Many or all of these established competitors are also involved in research and drug development regarding various chemokine receptors. Pharmaceutical and biotechnology companies which are known to be involved in chemokine and chemoattractant research and related drug development include, but are not limited to, Pfizer, GlaxoSmithKline, Bristol-Myers Squibb, Merck, Takeda, Sanofi, Incyte, Achillion, Alexion, Allergan, Appellis, Omeros, InflaRX, Dimerix, X4 Pharmaceuticals, Mitsubishi Tanabe, Biolinerx, Akari Therapeutics and UCB Pharma, among others.

We are developing small molecule therapeutics that will compete with other drugs and alternative therapies that are currently marketed or are being developed to treat AAV, C3G, HS, FSGS and other renal disease, other autoimmune diseases, metabolic diseases, inflammatory disorders, and cancer. Similarly, other future drug candidates we are pursuing would compete against numerous existing and established drugs and potentially against other novel drugs and therapies that are currently in development. See “Item 1. Business—Competition.” We also anticipate that we will face increased competition in the future as new companies enter into our target markets and scientific developments surrounding the chemokine system continue to develop.

Many of our competitors have materially greater name recognition and financial, manufacturing, marketing, research and drug development resources than we do. Additional mergers and acquisitions in the biotechnology and pharmaceutical industries may result in even more resources being concentrated in our competitors. Large pharmaceutical companies in particular have extensive expertise in preclinical and clinical testing and in obtaining regulatory approvals for drugs. In addition, academic institutions, government agencies, and other public and private organizations conducting research may seek patent protection with respect to potentially competitive products or technologies. These organizations may also establish exclusive collaborative or licensing relationships with our competitors.

We may be subject to costly product liability claims related to our clinical trials and drug candidates and, if we are unable to obtain adequate insurance or are required to pay for liabilities resulting from a claim excluded from, or beyond the limits of, our insurance coverage, a material liability claim could adversely affect our financial condition.

Because we conduct clinical trials with human patients, we face the risk that the use of our drug candidates may result in adverse side effects to patients and to otherwise healthy volunteers in our clinical trials. We face even greater risks upon any commercialization of our drug candidates. Although we have product liability insurance for clinical trials for up to $10.0 million, our insurance may be insufficient to reimburse us for any expenses or losses we may suffer, and we will be required to increase our product liability insurance coverage for our advanced clinical trials that we plan to initiate. We do not know whether we will be able to continue to obtain product liability coverage and obtain expanded coverage on acceptable terms, or at all. We may not have sufficient resources to pay for any liabilities resulting from a claim excluded from, or beyond the limits of, our insurance coverage. There is also a risk that third parties that we have agreed to indemnify could incur liability. An individual may bring a product liability claim against us if one of our drug candidates or products causes, or is claimed to have caused, an injury or is found to be unsuitable for consumer use. Any product liability claim brought against us, with or without merit, could result in:

- withdrawal of clinical trial volunteers, investigators, patients or trial sites;
- the inability to commercialize our drug candidates;
- decreased demand for our drug candidates;
- regulatory investigations that could require costly recalls or product modifications;

- loss of revenues;
- substantial costs of litigation;
- liabilities that substantially exceed our product liability insurance, which we would then be required to pay ourselves;
- an increase in our product liability insurance rates or the inability to maintain insurance coverage in the future on acceptable terms, if at all;
- the diversion of management's attention from our business; and
- damage to our reputation and the reputation of our products.

Our business involves the use of hazardous materials and we and our third-party manufacturers must comply with environmental laws and regulations, which may be expensive and restrict how we do business.

Our third-party manufacturers' activities and our own activities involve the controlled storage, use, handling and disposal of hazardous materials, including the components of our pharmaceutical products, test samples and reagents, biological materials and other hazardous compounds. We and our manufacturers are subject to federal, state and local and foreign laws and regulations governing the use, generation, manufacture, storage, handling and disposal of these hazardous materials. We currently carry no insurance specifically covering environmental claims relating to the use of hazardous materials. Although we believe that our safety procedures for handling and disposing of these materials and waste products comply with the standards prescribed by these laws and regulations, we cannot eliminate the risk of accidental injury or contamination from the use, storage, handling or disposal of hazardous materials. In the event of an accident, state or federal or other applicable authorities may curtail our use of these materials and/or interrupt our business operations. In addition, if an accident or environmental discharge occurs, or if we discover contamination caused by prior operations, including by prior owners and operators of properties we acquire, we could be liable for cleanup obligations, damages and fines. The substantial unexpected costs we may incur could significantly harm our financial condition and results of operations.

Future financings may adversely affect our stockholders or impose additional restrictions on our assets or operations, which may harm our business.

If we raise additional capital by issuing equity securities or convertible debt securities, then our existing stockholders' ownership will be diluted and the terms of any new equity securities may have preferences over our common stock. If we raise additional capital through the issuance of debt securities, the debt will have rights senior to the holders of our common stock and may contain covenants that restrict our operational flexibility or impose liens or other restrictions on our assets, in addition to the restrictions imposed by our credit facility with Hercules. In addition, the terms of future financings may restrict our ability to raise additional capital, which would delay or prevent the further development or commercialization of our drug candidates. If we raise additional funds through collaboration, licensing or other similar arrangements, it may be necessary to relinquish potentially valuable rights to our current drug candidates, potential products or proprietary technologies, or grant licenses on terms that are not favorable to us. If adequate funds are not available, our ability to achieve profitability or to respond to competitive pressures would be significantly limited and we may be required to delay, significantly curtail or eliminate the development of one or more of our drug candidates.

We are highly dependent on the services of our founder, President and Chief Executive Officer, Dr. Thomas J. Schall, and if we are not able to retain Dr. Schall or other members of our management or recruit additional management, clinical and scientific personnel, our business will suffer.

We may not be able to attract or retain qualified management and scientific and clinical personnel in the future due to the intense competition for qualified personnel among biotechnology, pharmaceutical and other

businesses, particularly in the San Francisco Bay area. Our industry has experienced a high rate of turnover of management personnel in recent years. If we are not able to attract, retain and motivate necessary personnel to accomplish our business objectives, we may experience constraints that will significantly impede the achievement of our development objectives, our ability to raise additional capital and our ability to implement our business strategy.

We are highly dependent on the principal members of our management and scientific staff. The loss of service of any of our management could harm our business. In addition, we are dependent on our continued ability to attract, retain and motivate highly qualified additional management, clinical and scientific personnel. The competition for qualified personnel in the pharmaceutical industry is intense. Due to our limited resources, we may not be able to effectively attract and recruit additional qualified personnel. If we are not able to retain our management, particularly our founder, President and Chief Executive Officer, Dr. Schall, and attract, on acceptable terms, additional qualified personnel necessary for the continued development of our business, we may not be able to sustain our operations or grow our business. Although we have executed employment agreements with each member of our current executive management team, including Dr. Schall, these agreements are terminable at will with or without notice and, therefore, we may not be able to retain their services as expected. In addition to the competition for personnel, the San Francisco Bay area in particular is characterized by a high cost of living. As such, we could have difficulty attracting experienced personnel to our company and may be required to expend significant financial resources in our employee recruitment and retention efforts.

In addition, we have scientific and clinical advisors who assist us in formulating our product development and clinical strategies. These advisors are not our employees and may have commitments to, or consulting or advisory contracts with, other entities that may limit their availability to us, or may have arrangements with other companies to assist in the development of products that may compete with ours.

We are required to maintain compliance with Section 404 of the Sarbanes-Oxley Act of 2002 or we may be subject to sanctions by regulatory authorities.

Section 404(a) of the Sarbanes-Oxley Act of 2002 requires that we evaluate and determine the effectiveness of our internal controls over financial reporting and provide a management report on the internal control over financial reporting. We have performed the system and process evaluation and testing required to comply with the management certification. We are also required to comply with auditor attestation requirements of Section 404(b) of the Sarbanes-Oxley Act of 2002. If we do not properly implement the requirements of Section 404 with adequate compliance, and maintain such compliance, we may be subject to sanctions or investigation by regulatory authorities, such as the SEC or The Nasdaq Stock Market LLC, or Nasdaq. Any such action could adversely affect our financial results or investors' confidence in us and could cause our stock price to fall. If we have a material weakness in our internal controls over financial reporting, we may not detect errors on a timely basis and our consolidated financial statements may be materially misstated. If we or our independent registered public accounting firm identifies deficiencies in our internal controls that are deemed to be material weaknesses, we could be subject to sanctions or investigations by Nasdaq, the SEC or other regulatory authorities, which would entail expenditure of additional financial and management resources and could materially adversely affect our stock price.

We may be adversely affected by the economic environment.

Our ability to attract and retain collaboration partners or customers, invest in and grow our business and meet our financial obligations depends on our operating and financial performance, which, in turn, is subject to numerous factors, including the prevailing economic conditions and financial, business and other factors beyond our control, such as the rate of unemployment, the number of uninsured persons in the United States and inflationary pressures. We cannot anticipate all the ways in which the current economic climate and financial market conditions could adversely impact our business.

We are exposed to risks associated with reduced profitability and the potential financial instability of our collaboration partners or customers, many of which may be adversely affected by volatile conditions in the financial markets. For example, unemployment and underemployment, and the resultant loss of insurance, may decrease the demand for healthcare services and pharmaceuticals. If fewer patients are seeking medical care because they do not have insurance coverage, our collaboration partners or customers may experience reductions in revenues, profitability and/or cash flow that could lead them to reduce their support of our programs or financing activities. If collaboration partners or customers are not successful in generating sufficient revenue or are precluded from securing financing, they may not be able to pay, or may delay payment of, accounts receivable that are owed to us. This, in turn, could adversely affect our financial condition and liquidity. In addition, if economic challenges in the United States result in fewer individuals pursuing or being able to afford our products once commercialized, our business, results of operations, financial condition and cash flows could be adversely affected.

Our internal computer systems, or those of our CROs or other contractors or consultants, may fail or suffer security breaches, which could result in a material disruption of our drug development programs.

Despite the implementation of security measures, our internal computer systems and those of our CROs and other contractors and consultants are vulnerable to damage from computer viruses, unauthorized access, natural disasters, terrorism, war and telecommunication and electrical failures. While we have not experienced any such system failure, accident or security breach to date, if such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our drug development programs, adverse publicity, and fines or penalties. For example, the loss of clinical trial data from completed or ongoing clinical trials for any of our drug candidates could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any disruption or security breach were to result in a loss of or damage to our data or applications, or inappropriate disclosure of confidential or proprietary information, we could incur liability and the further development of our drug candidates could be delayed.

Our employees, independent contractors, principal investigators, CROs, consultants, vendors and collaboration partners may engage in misconduct or other improper activities, including noncompliance with regulatory standards and requirements.

We are exposed to the risk of that our employees, independent contractors, principal investigators, CROs, consultants, vendors and collaboration partners may engage in fraudulent or other illegal activity. Misconduct by these parties could include intentional, reckless and/or negligent conduct or disclosure of unauthorized activities to us that violate: FDA regulations, including those that require the reporting of true, complete and accurate information to the FDA; manufacturing standards we have established; federal and state healthcare fraud and abuse laws and regulations; and laws that require the reporting of true, complete and accurate financial information or data. In particular, sales, marketing and business arrangements in the healthcare industry are subject to extensive laws and regulations intended to prevent fraud, kickbacks, self-dealing and other abusive practices. These laws and regulations may restrict or prohibit a wide range of pricing, discounting, marketing and promotion, sales commission, customer incentive programs and other business arrangements. These activities could also include the improper use of information obtained in the course of clinical trials, which could result in regulatory sanctions and serious harm to our reputation. It is not always possible to identify and deter misconduct by employees and other third parties, and the precautions we take to detect and prevent this activity may not be effective in controlling unknown or unmanaged risks or losses or in protecting us from governmental investigations or other actions or lawsuits stemming from a failure to be in compliance with such laws or regulations. If any such actions are instituted against us, and we are not successful in defending ourselves or asserting our rights, those actions could have a significant impact on our business, including the imposition of significant civil, criminal and administrative penalties, damages, monetary fines, possible exclusion from participation in Medicare, Medicaid and other U.S. federal healthcare programs, contractual damages, reputational harm, diminished profits and future earnings, and curtailment of our operations.

Our ability to utilize our net operating loss carryforwards and certain other tax attributes may be limited.

Under Section 382 of the Internal Revenue Code of 1986, as amended, if a corporation undergoes an "ownership change" (generally defined as a greater than 50% change, by value, in its equity ownership over a three year period), the corporation's ability to use its pre-change net operating loss carryforwards and other pre-change tax attributes to offset its post-change taxable income and taxes may be limited. We previously determined that we had ownership changes, which limit our ability to use our then existing tax attributes. Future changes in our stock ownership, many of the causes of which are outside our control, could result in additional ownership changes. Any such ownership changes could further limit our ability to use net operating loss carryforwards and other pre-change tax attributes. Furthermore, under U.S. tax legislation enacted in 2017, the treatment of tax losses generated before December 31, 2017 has generally not changed but tax losses generated in calendar year 2018 and beyond may be used to offset only 80% of taxable income and carryforward indefinitely. This change may require us to pay federal income taxes in future years despite generating a loss for federal income tax purposes in prior years.

Recent U.S. tax legislation may materially adversely affect our financial condition, results of operations and cash flows.

The Tax Cuts and Jobs Act of 2017 significantly changed the U.S. federal income taxation of U.S. corporations, including by reducing the U.S. corporate income tax rate, and revising the rules governing net operating losses and foreign tax credits. Many of these changes were effective immediately, without any transition periods or grandfathering for existing transactions. The legislation is unclear in many respects and could be subject to potential amendments and technical corrections, as well as interpretations and implementing regulations by the Treasury and Internal Revenue Service, or IRS, any of which could lessen or increase certain adverse impacts of the legislation. In addition, it is unclear how these U.S. federal income tax changes will affect state and local taxation, which often uses federal taxable income as a starting point for computing state and local tax liabilities. As of December 31, 2017, we recorded a provisional impact to write down the deferred income tax assets (including the value of our net operating loss carryforwards and our tax credit carryforwards) of $36.0 million with an equal offset to the valuation allowance. As of December 31, 2018, we have finalized our computation of the impact of the Tax Cuts and Jobs Act and no material adjustments were recorded. Further, there may be material adverse effects resulting from the legislation that we have not yet identified. While some of the changes made by the tax legislation may adversely affect us in one or more reporting periods and prospectively, other changes may be beneficial on a going forward basis. We will continue to evaluate the impact of the Tax Cuts and Jobs Act as we receive further guidance and notices and will make adjustments to the financial statements or related disclosures accordingly.

Business disruptions could seriously harm our future revenues and financial condition and increase our costs and expenses.

Our operations could be subject to earthquakes, power shortages, telecommunications failures, floods, hurricanes, typhoons, fires, extreme weather conditions, medical epidemics and other natural or manmade disasters or business interruptions. The occurrence of any of these business disruptions could seriously harm our operations and financial condition and increase our costs and expenses. Our corporate headquarters is located in California and certain clinical sites for our drug candidates, operations of our existing and future partners and suppliers are or will be located in California near major earthquake faults and fire zones. The ultimate impact on us, our significant partners, suppliers and our general infrastructure of being located near major earthquake faults and fire zones and being consolidated in certain geographical areas is unknown, but our operations and financial condition could suffer in the event of a major earthquake, fire or other natural or manmade disaster.

Risks Related to Intellectual Property

Our proprietary rights may not adequately protect our technologies and drug candidates. If we are unable to protect our drug candidates and our intellectual property rights, it may materially adversely affect our position in the market.

Our commercial success will depend on our ability to obtain patents and maintain adequate protection for our technologies, intellectual property and drug candidates in the United States and other countries. Our patent estate, on a worldwide basis, includes approximately 849 issued or allowed patents and approximately 362 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. There are approximately 95 issued or allowed patents and 33 patent applications pending for avacopan, our lead drug candidate in the C5aR program. With respect to our drug candidates in the CCR2 programs, we have approximately 78 issued or allowed patents and 67 patents pending worldwide relating to their chemical composition or use thereof. With respect to the CCR1 and CCR9 chemokine receptors, we have approximately 495 issued or allowed patents and 124 patents pending worldwide relating to their chemical composition or use thereof. We have approximately 120 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs. We cannot assure you that any of our patent applications will result in issued patents. We will be able to protect our proprietary rights from unauthorized use by third parties only to the extent that our proprietary technologies and future products are covered by valid and enforceable patents or are effectively maintained as trade secrets. If third parties disclose or misappropriate our proprietary rights, it may materially and adversely impact our position in the market.

We apply for patents covering both our technologies and drug candidates, as we deem appropriate. However, we may fail to apply for patents on important technologies or drug candidates in a timely fashion, or at all. Our existing patents and any future patents we obtain may not be sufficiently broad to prevent others from using our technologies or from developing competing products and technologies. Composition-of-matter patents on the chemical active pharmaceutical ingredient are generally considered to be the strongest form of intellectual property protection for pharmaceutical products, as such patents provide protection without regard to any method of use. We cannot be certain that the claims in our patent applications covering composition-of-matter of our drug candidates will be considered patentable by the USPTO and courts in the United States or by the patent offices and courts in other countries, nor can we be certain that the claims in our issued composition-of-matter patents will not be found invalid or unenforceable if challenged. Method-of-use patents protect the use of a product for the specified method. This type of patent does not prevent a competitor from making and marketing a product that is identical to our product for an indication that is outside the scope of the patented method. Moreover, even if competitors do not actively promote their product for our targeted indications, physicians may prescribe these products "off-label." Although off-label prescriptions may infringe or contribute to the infringement of method-of-use patents, the practice is common and such infringement is difficult to prevent or prosecute.

The issuance of a patent is not conclusive as to its inventorship, scope, validity or enforceability, and our patents or the patent rights that we license from others, may be challenged in the courts or patent offices in the United States and abroad. Once granted, patents may remain open to opposition, interference, re-examination, post-grant review, inter partes review, nullification or derivation action or similar proceedings in court or before patent offices in the United States or foreign jurisdictions for a given period after allowance or grant, during which time third parties can raise objections against such patents. Such challenges may result in loss of exclusivity or in patent claims being narrowed, invalidated or held unenforceable, all of which could limit our ability to stop others from using or commercializing similar or identical product candidates, or limit the duration of the patent protection of our drug candidates.

Moreover, the patent positions of numerous biotechnology and pharmaceutical companies are highly uncertain and involve complex legal and factual questions for which important legal principles remain

unresolved. As a result, the validity and enforceability of our patents cannot be predicted with certainty. In addition, we cannot assure you that:

- we were the first to make the inventions covered by each of our issued patents and pending patent applications;
- we were the first to file patent applications for these inventions;
- others will not independently develop similar or alternative technologies or duplicate any of our technologies by inventing around our claims;
- any of our pending patent applications will result in issued patents;
- a third party will not challenge our proprietary rights or that a court will hold that our patents are valid and enforceable;
- any patents issued to us or our collaboration partners will provide us with any competitive advantages, or will not be challenged by third parties;
- we will develop additional proprietary technologies that are patentable; or
- the patents of others will not have an adverse effect on our business.

Changes in patent law in the United States or in other countries could diminish the value of patents in general, thereby impairing our ability to protect our product candidates.

Our patent rights may be affected by developments or uncertainty in the United States' or other jurisdictions' patent statutes, patent case law, USPTO rules and regulations or the rules and regulations of other jurisdictions' patent offices.

There are a number of recent changes to United States patent laws that may have a significant impact on our ability to protect our technology and enforce our intellectual property rights. For example, on September 16, 2011, the Leahy-Smith America Invents Act, or Leahy-Smith Act, was signed into law. The Leahy-Smith Act includes a number of significant changes to United States patent law. These include provisions that affect the way patent applications will be prosecuted and may also affect patent litigation. In particular, under the Leahy-Smith Act, the United States transitioned in March 2013 to a "first to file" system in which the first inventor to file a patent application is typically entitled to the patent. Third parties are allowed to submit prior art before the issuance of a patent by the USPTO, and may become involved in post-grant proceedings including opposition, derivation, reexamination, inter partes review or interference proceedings challenging our patent rights or the patent rights of others. An adverse determination in any such submission, proceeding or litigation could reduce the scope or enforceability of, or invalidate, our patent rights, which could adversely affect our competitive position. In addition, the United States Congress may pass additional patent reform legislation that is unfavorable to us.

The Supreme Court has ruled on several patent cases in recent years, either narrowing the scope of patent protection available in certain circumstances or weakening the rights of patent owners in certain situations. In addition to increasing uncertainty with regard to our ability to obtain patents in the future, this combination of events has created uncertainty with respect to the value of patents once obtained. Depending on decisions by the United States Congress, the federal courts and the USPTO, the laws and regulations governing patents could change in unpredictable ways that would weaken our ability to obtain new patents or to enforce our existing patents and patents we might obtain in the future. Similarly, statutory or judicial changes to the patent laws of other countries may increase the uncertainties and costs surrounding the prosecution of patent applications and the enforcement or defense of issued patents.

We may become subject to third parties' claims alleging infringement of patents and proprietary rights or seeking to invalidate our patents or proprietary rights, which would be costly, time consuming and, if successfully asserted against us, delay or prevent the development and commercialization of our products.

Intellectual property litigation and patent litigation in particular, is expensive, complex and lengthy and its outcome is difficult to predict. There has been substantial litigation and other proceedings regarding patent and other intellectual property rights in the pharmaceutical and biotechnology industries. We may be subject to third-party claims in the future against us or our collaborators that would cause us to incur substantial expenses and, if successful against us, could cause us to pay substantial damages, including treble damages and attorney's fees if we are found to be willfully infringing a third party's patents. Further, if a patent infringement suit were brought against us or our collaborators, we or they could be forced to stop or delay research, development, manufacturing or sales of the product or drug candidate that is the subject of the suit. As a result of patent infringement claims, or in order to avoid potential claims, we or our collaborators may choose to seek, or be required to seek, a license from the third party and would most likely be required to pay license fees or royalties or both. These licenses may not be available on acceptable terms, or at all. Even if we or our collaborators were able to obtain a license, the rights may be nonexclusive, which would give our competitors access to the same intellectual property. Ultimately, we could be prevented from commercializing a product, or forced to redesign it, or to cease some aspect of our business operations if, as a result of actual or threatened patent infringement claims, we or our collaborators are unable to enter into licenses on acceptable terms. This could harm our business significantly.

Furthermore, the scope of a patent claim is determined by an interpretation of the law, the written disclosure in a patent and the patent's prosecution history and can involve other factors such as expert opinion. Our analysis of these issues, including interpretation the relevance or the scope of claims in a patent or a pending application, determining applicability of such claims to our proprietary technologies or drug candidates, predicting whether a third party's pending patent application will issue with claims of relevant scope, and determining the expiration date of any patent in the United States or abroad that we consider relevant may be incorrect, which may negatively impact our ability to develop and market our drug candidates. We do not always conduct independent reviews of pending patent applications of and patents issued to third parties.

Additionally, patent applications in the United States and elsewhere are typically published approximately 18 months after the earliest filing for which priority is claimed, with such earliest filing date being commonly referred to as the priority date. Certain United States applications that will not be filed outside the United States can remain confidential until patents issue. In addition, patent applications in the United States and elsewhere can be pending for many years before issuance, or unintentionally abandoned patents or applications can be revived. Furthermore, pending patent applications that have been published can, subject to certain limitations, be later amended in a manner that could cover our technologies, our drug candidates or the use of our drug candidates. These applications may later result in issued patents, or the revival of previously abandoned patents, that will prevent, limit or otherwise interfere with our ability to make, use or sell our products. As a result, we may be unaware of third-party patents that may be infringed by commercialization of our drug candidates, and cannot be certain that we were the first to file a patent application related to a drug candidate or proprietary technology. In addition, identification of third-party patent rights that may be relevant to our technology is difficult because patent searching is imperfect due to differences in terminology among patents, incomplete databases and the difficulty in assessing the meaning of patent claims.

In addition to infringement claims against us, third parties may challenge or infringe upon our existing or future patents. Proceedings involving our patents or patent applications or those of others could result in adverse decisions regarding the patentability of our inventions relating to our drug candidates, and/or the enforceability, validity or scope of protection offered by our patents relating to our drug candidates. Even if we are successful in these proceedings, we may incur substantial costs and divert management time and attention in pursuing these proceedings, which could have a material adverse effect on us. Or, if third parties prepare and file patent applications in the United States that also claim technology to which we have rights, we may have to participate in interference proceedings in the USPTO to determine the priority of invention. We may also become involved

in similar opposition proceedings in the European Patent Office regarding our intellectual property rights with respect to our products and technology.

The cost to us of any intellectual property litigation or other proceedings could be substantial. Some of our competitors may be able to sustain the costs of such litigation or proceedings more effectively than we can because of their substantially greater financial resources. Discovery proceedings in the United States might lead to the disclosure of some of our proprietary confidential information. Uncertainties resulting from the initiation and continuation of patent litigation or other proceedings could have a material adverse effect on our ability to compete in the marketplace. Intellectual property litigation and other proceedings may also absorb significant management and technical staff's time which may materially and adversely impact our financial position and results of operations.

Restrictions on our patent rights relating to our drug candidates may limit our ability to prevent third parties from competing against us.

Our success will depend, in part, on our ability to obtain and maintain patent protection for our drug candidates, preserve our trade secrets, prevent third parties from infringing upon our proprietary rights and operate without infringing upon the proprietary rights of others. Composition-of-matter patents on APIs are generally considered to be the strongest form of intellectual property protection for pharmaceutical products as they apply without regard to any method of use. Entirely new individual chemical compounds, often referred to as new chemical entities, are typically entitled to composition-of-matter coverage. However, we cannot be certain that the current law will remain the same, or that our drug candidates will be considered novel and non-obvious by the USPTO and courts.

In addition to composition-of-matter patents and patent applications, we also have filed method-of-use patent applications. This type of patent protects the use of the product only for the specified method. However, this type of patent does not prevent a competitor from making and marketing a product that is identical to our product for an indication that is outside the scope of the patented method. Moreover, even if these competitors do not actively promote their product for our targeted indication, physicians may prescribe these products "off-label." Although off-label prescriptions may infringe or contribute to the infringement of method-of-use patents, the practice is common and such infringement is difficult to prevent or prosecute.

Patent applications in the United States and most other countries are confidential for a period of time until they are published, and publication of discoveries in scientific or patent literature typically lags actual discoveries by several months or more. As a result, we cannot be certain that we and the inventors of the issued patents and applications that we may in-license were the first to conceive of the inventions covered by such patents and pending patent applications or that we and those inventors were the first to file patent applications covering such inventions. Also, we have numerous issued patents and some patent applications pending before the USPTO and the patent protection may lapse before we manage to obtain commercial value from them, which might result in increased competition and materially affect our position in the market.

We may be subject to claims that we or our employees or consultants have wrongfully used or disclosed alleged trade secrets of our employees' or consultants' former employers or their clients. These claims may be costly to defend and if we do not successfully do so, we may be required to pay monetary damages and may lose valuable intellectual property rights or personnel.

Many of our employees were previously employed at universities or biotechnology or pharmaceutical companies, including our competitors or potential competitors. Although no claims against us are currently pending, we may be subject to claims that these employees or we have inadvertently or otherwise used or disclosed trade secrets or other proprietary information of their former employers. Litigation may be necessary to defend against these claims. If we fail in defending such claims, in addition to paying monetary damages, we may lose valuable intellectual property rights or personnel. A loss of key research personnel or their work

product could hamper our ability to commercialize, or prevent us from commercializing our drug candidates, which could severely harm our business. Even if we are successful in defending against these claims, litigation could result in substantial costs and be a distraction to management.

Some of our intellectual property which is discovered through government funded programs is subject to federal regulation such as "march-in" rights, certain reporting requirements, and a preference for U.S. industry. Compliance with such regulations may limit our exclusive rights, subject us to expenditure of resources with respect to reporting requirements, and limit our ability to contract with foreign manufacturers.

Some of our existing drug candidates, including CCX140, and some of our research and development work were funded, at least in part, by the U.S. government and are therefore subject to certain federal regulations. For example, some of our research and development work on vaccine adjuvants and immunomodulation for biothreat applications was funded by government research grants. In addition, as noted on several of our patents including U.S. Patent Nos. 7,884,110; 7,622,583; 7,776,877; 8,198,309 and 8.093,247, inventions covering various CCR9 and CCR2 inhibitors were supported at least in part by National Institutes of Health funding (U19-AI056690-01). Under the "march-in" provisions of the Bayh-Dole Act, the government may have the right under limited circumstances to require us to grant exclusive, partially exclusive or non-exclusive rights to third parties for intellectual property discovered through the government funded program. The government can exercise its march-in rights if it determines that action is necessary because we fail to achieve practical application of the new invention or because action is necessary to alleviate health or safety needs of the public. Intellectual property discovered under the government funded program is also subject to certain reporting requirements, compliance with which may require us to expend substantial resources. Such intellectual property is also subject to a preference for U.S. industry, which may limit our ability to contract with foreign product manufacturers for products covered by such intellectual property. We plan to apply for additional U.S. government funding, and it is possible that we may discover compounds or drug candidates as a result of such funding. Intellectual property under such discoveries would be subject to the applicable provisions of the Bayh-Dole Act.

Risks Related to Government Regulation

The regulatory approval process is expensive, time consuming and uncertain and may prevent us from obtaining approvals for the commercialization of some or all of our drug candidates.

The research, testing, manufacturing, labeling, approval, selling, import, export, marketing and distribution of drug products are subject to extensive regulation by the FDA and other regulatory authorities in the United States, the EMA, the EU institutions (e.g., the European Commission) and the EU Member State Competent Authorities, as well as equivalent authorities and regulatory bodies in other countries, which regulations differ from country to country. We are not permitted to market our drug candidates in the United States until we receive approval of a new drug application, or NDA, from the FDA and in the EU until we have received approval from the European Commission or EU Member State Competent Authorities. We have not submitted an application for or received marketing approval for any of our drug candidates, except in the EU where we have applied to the EMA for a CMA, which we subsequently withdrew, for avacopan in the treatment of patients with AAV. Obtaining approval of an NDA or CMA can be a lengthy, expensive and uncertain process. In addition, failure to comply with FDA, EMA and other applicable U.S., EU and foreign regulatory requirements may subject us to administrative or judicially imposed sanctions, including:

- warning letters;
- civil and criminal penalties;
- injunctions;
- withdrawal of approved products;
- product seizure or detention;
- product recalls;

- total or partial suspension of production; and
- refusal to approve pending NDAs or supplements to approved NDAs, pending CMA or marketing authorization applications, or MAAs.

Prior to receiving approval to commercialize any of our drug candidates in the United States, the EU, or abroad, we must demonstrate with substantial evidence from well controlled clinical trials, and to the satisfaction of the FDA, the EMA, and other regulatory authorities abroad, that such drug candidates are safe and effective for their intended uses. Results from preclinical studies and clinical trials can be interpreted in different ways. Even if we believe the preclinical or clinical data for our drug candidates are promising, such data may not be sufficient to support approval by the FDA, the EMA, and other regulatory authorities. Administering any of our drug candidates to humans may produce undesirable side effects, which could interrupt, delay or halt clinical trials of our drug candidates and result in the FDA, the EMA, or other regulatory authorities denying approval of our drug candidates for any or all targeted indications.

Regulatory approval of an NDA or NDA supplement, or of a CMA, MAA, or of their respective extensions and variations, is not guaranteed, and the approval process is expensive and may take several years. The FDA and the EMA also have substantial discretion in the approval process. Despite the time and expense exerted, failure can occur at any stage, and we could encounter problems that cause us to abandon or repeat clinical trials, or perform additional preclinical studies and clinical trials. The number of preclinical studies and clinical trials that will be required for FDA or EMA approval varies depending on the drug candidate, the disease or condition that the drug candidate is designed to address, and the regulations applicable to any particular drug candidate. The FDA or EMA can delay, limit or deny approval of a drug candidate for many reasons, including, but not limited to, the following:

- a drug candidate may not be deemed safe or effective;
- FDA or EMA officials may not find the data from preclinical studies and clinical trials sufficient;
- the FDA or EMA might not approve our or our third-party manufacturer's processes or facilities; or
- the FDA or EMA may change its approval policies or adopt new regulations.

If any of our drug candidates fails to demonstrate safety and efficacy in clinical trials or does not gain regulatory approval, our business and results of operations will be materially and adversely harmed.

Even if we receive regulatory approval for a drug candidate, we will be subject to ongoing regulatory obligations and continued regulatory review, which may result in significant additional expense and subject us to penalties if we fail to comply with applicable regulatory requirements.

Once regulatory approval has been granted, the approved product and its manufacturer are subject to continual review by the FDA, the EMA and EU Member State Competent Authorities, and/or non-U.S./non-EU regulatory authorities. Any regulatory approval that we receive for our drug candidates may be subject to limitations on the indicated uses for which the product may be marketed or contain requirements for potentially costly post-marketing follow-up studies to monitor the safety and efficacy of the product. The FDA and the EMA also have authority to require a risk evaluation and mitigation strategy, or REMS, or risk management plan, as part of an NDA, CMA, MAA, or after approval, which may impose further requirements or restrictions on the distribution or use of an approved drug, such as limiting prescribing to certain physicians or medical centers that have undergone specialized training, limiting treatment to patients who meet certain safe-use criteria or requiring treated patients to enroll in a registry. In addition, if the FDA, the EMA, EU Member State Competent Authorities, and/or non-U.S./non-EU regulatory authorities approve any of our drug candidates, we will be subject to extensive and ongoing regulatory requirements by the FDA, the EMA, EU Member State Competent Authorities, and other regulatory authorities with regard to the labeling, packaging, adverse event reporting, storage, advertising, promotion and recordkeeping for our products. The FDA and the EMA, the European

institutions and the EU Member State Competent Authorities, strictly regulate the promotional claims that may be made about prescription products. In particular, a product may not be promoted for uses that are not approved by the FDA or the European Commission as reflected in the product's approved labeling. If we receive marketing approval for any of our drug candidates, physicians may nevertheless prescribe our products to their patients in a manner that is inconsistent with the approved label. If we are found to have promoted such off-label uses, we may become subject to significant liability and government fines.

In addition, manufacturers of our drug products are required to comply with cGMP regulations, which include requirements related to quality control and quality assurance as well as the corresponding maintenance of records and documentation. Further, regulatory authorities must approve manufacturing facilities before they can be used to manufacture our drug products, and such facilities are subject to continual review and periodic inspections by the FDA, the EMA, EU Member State Competent Authorities, and other regulatory authorities for compliance with cGMP regulations. If we or a regulatory authority discovers previously unknown problems with a product, such as adverse events of unanticipated severity or frequency, or problems with the facility where the product is manufactured, a regulatory authority may impose restrictions on that product, the manufacturer or us, including imposition of a REMS, or similar risk management measures, or requesting recall or withdrawal of the product from the market or suspension of manufacturing. If we, our drug candidates or the manufacturing facilities for our drug candidates fail to comply with regulatory requirements of the FDA, the EMA, the EU institutions, the EU Member State Competent Authorities and/or other non-U.S./non-EU regulatory authorities, we could be subject to administrative or judicially imposed sanctions, including:

- warning letters;
- civil or criminal penalties;
- injunctions;
- suspension of or withdrawal of regulatory approval;
- suspension of any ongoing clinical trials;
- voluntary or mandatory product recalls and publicity requirements;
- refusal to approve pending applications for marketing approval of new drugs or supplements to approved applications filed by us;
- restrictions on operations, including costly new manufacturing requirements; or
- seizure or detention of our products or import bans.

The regulatory requirements and policies may change and additional government regulations may be enacted for which we may also be required to comply. For example, the 21st Century Cures Act, or the Cures Act, was signed into law. The Cures Act is intended, among other things, to modernize the regulation of drugs and spur innovation. If we are slow or unable to adapt to changes in existing requirements or the adoption of new requirements or policies, or if we are not able to maintain regulatory compliance, we will not be permitted to market our future products and our business will suffer.

We cannot predict the likelihood, nature or extent of government regulation that may arise from future legislation or administrative action, either in the United States, the EU or in other countries or jurisdictions. For example, certain policies of President Trump's administration may impact our business and industry. Namely, the current administration has taken several executive actions, including the issuance of a number of Executive Orders, that could impose significant burdens on, or otherwise materially delay, the FDA's ability to engage in routine regulatory and oversight activities such as implementing statutes through rulemaking, issuance of guidance and review and approval of marketing applications. It is difficult to predict how these changes will be implemented, and the extent to which they will impact the FDA's ability to exercise its regulatory authority. If these executive actions impose constraints on the FDA's ability to engage in oversight and implementation activities in the normal course, our business may be negatively impacted.

Changes in funding for the FDA and other government agencies could hinder their ability to hire and retain key leadership and other personnel, or otherwise prevent new products and services from being developed or commercialized in a timely manner, which could negatively impact our business.

The ability of the FDA to review and approve new products can be affected by a variety of factors, including government budget and funding levels, ability to hire and retain key personnel and accept the payment of user fees, and statutory, regulatory, and policy changes. Average review times at the agency have fluctuated in recent years as a result. In addition, government funding of other government agencies that fund research and development activities is subject to the political process, which is inherently fluid and unpredictable.

Disruptions at the FDA and other agencies may also slow the time necessary for new drugs to be reviewed and/or approved by necessary government agencies, which would adversely affect our business. For example, over the last several years, including for 35 days beginning on December 22, 2018, the U.S. government has shut down several times and certain regulatory agencies, such as the FDA, have had to furlough critical FDA employees and stop critical activities. If a prolonged government shutdown occurs, it could significantly impact the ability of the FDA to timely review and process our regulatory submissions, which could have a material adverse effect on our business.

The availability of adequate third-party coverage and reimbursement for newly approved drugs is uncertain, and failure to obtain adequate coverage and reimbursement from third-party payors could impede our ability to market any future products we may develop and could limit our ability to generate revenue.

There is significant uncertainty related to the third-party payor coverage and reimbursement of newly approved drugs. The commercial success of our future products in both domestic and international markets depends on whether such third-party coverage and reimbursement is available for our future products. Governmental payors, including Medicare and Medicaid, health maintenance organizations and other third-party payors are increasingly attempting to manage their healthcare expenditures by limiting both coverage and the level of reimbursement of new drugs and, as a result, they may not cover or provide adequate reimbursement for our future products. These payors may not view our future products as cost-effective, and coverage and reimbursement may not be available to our customers or may not be sufficient to allow our future products to be marketed on a competitive basis. Third-party payors are exerting increasing influence on decisions regarding the use of, and coverage and reimbursement levels for, particular treatments. Such third-party payors, including Medicare, are challenging the prices charged for medical products and services, and many third-party payors limit or delay coverage and reimbursement for newly approved healthcare products. In particular, third-party payors may limit the covered indications. Cost-control initiatives could cause us to decrease the price we might establish for products, which could result in lower than anticipated product revenues. If the prices for our drug candidates decrease or if governmental and other third-party payors do not provide adequate coverage or reimbursement, our prospects for revenue and profitability will suffer.

Failure to obtain regulatory approvals in foreign jurisdictions will prevent us from marketing our products internationally.

We may market future products in international markets. In order to market our future products in the EEA and many other foreign jurisdictions, we must obtain separate regulatory approvals. More concretely, in the EEA, medicinal products can only be commercialized after obtaining a Marketing Authorization, or MA. There are two types of marketing authorizations:

- The Community MA, which is issued by the European Commission through the Centralized Procedure, based on the opinion of the Committee for Medicinal Products for Human Use, or CHMP, of the EMA, and which is valid throughout the entire territory of the EEA. The Centralized Procedure is mandatory for certain types of products, such as biotechnology medicinal products, orphan medicinal products, and medicines that contain a new active substance indicated for the treatment of AIDS, cancer, neurodegenerative disorders,

diabetes, auto-immune and viral diseases. The Centralized Procedure is optional for products containing a new active substance not yet authorized in the EEA, or for products that constitute a significant therapeutic, scientific or technical innovation or which are in the interest of public health in the EU.

- National MAs, which are issued by the competent authorities of the Member States of the EEA and only cover their respective territory, are available for products not falling within the mandatory scope of the Centralized Procedure. Where a product has already been authorized for marketing in a Member State of the EEA, this National MA can be recognized in another Member State through the Mutual Recognition Procedure. If the product has not received a National MA in any Member State at the time of application, it can be approved simultaneously in various Member States through the Decentralized Procedure.

Under the above described procedures, before granting the MA, the EMA or the competent authorities of the Member States of the EEA make an assessment of the risk-benefit balance of the product on the basis of scientific criteria concerning its quality, safety and efficacy.

In the EEA, upon receiving marketing authorization, new chemical entities generally receive eight years of data exclusivity and an additional two years of market exclusivity. If granted, data exclusivity prevents regulatory authorities in the EU from referencing the innovator's data to assess a generic application. During the additional two-year period of market exclusivity, a generic marketing authorization can be submitted, and the innovator's data may be referenced, but no generic product can be marketed until the expiration of the market exclusivity. However, there is no guarantee that a product will be considered by the EU's regulatory authorities to be a new chemical entity and qualify for data exclusivity.

To meet unmet medical needs of patients and in the interest of public health, the EMA may grant, subject to certain specific obligations to be reviewed annually, a CMA, on the basis of less complete data than is normally required. To be eligible for a CMA, a medicinal product must belong to at least one of these categories: (i) be aimed at treating, preventing or diagnosing seriously debilitating or life-threatening diseases; (ii) be intended for use in emergency situations; or (iii) be designated as an orphan medicine. Further, a CMA may only be granted if the EMA finds that all the following requirements are met:

- the benefit-risk balance of the product is positive;
- it is likely that the applicant will be able to provide comprehensive data;
- unmet medical needs will be fulfilled; and
- the benefit to public health of the medicinal product's immediate availability on the market outweighs the risks due to the need for further data.

In 2016, the EMA launched its Priority Medicines, or PRIME, scheme. PRIME is a voluntary scheme aimed at enhancing the EMA's support for the development of medicines that target unmet medical needs. It is based on increased interaction and early dialogue with companies developing promising medicines, to optimize their product development plans and speed up their evaluation to help them reach patients earlier. The scheme focuses on medicines that may offer a major therapeutic advantage over existing treatments, or benefit patients without treatment options. These medicines are considered priority medicines by the EMA. To be accepted for PRIME, a medicine has to show its potential to benefit patients with unmet medical needs based on early clinical data. The benefits of a PRIME designation include the appointment of a CHMP rapporteur, before submission of the marketing authorization application, early dialogue and scientific advice at key development milestones, and the potential to qualify products for accelerated review earlier in the application process. In June 2016, avacopan was granted access to PRIME for the treatment of patients with AAV. Even though we have access to PRIME for avacopan, this may not result in a materially faster development process, review or approval compared to conventional EMA procedures. Further, obtaining access to PRIME does not assure or increase the likelihood of EMA's grant of a marketing authorization.

We have had limited interactions with foreign regulatory authorities, and the approval procedures vary among countries and can involve additional clinical testing, and the time required to obtain approval may differ

from that required to obtain FDA approval. Clinical trials conducted in one country may not be accepted by regulatory authorities in other countries. Approval by the FDA does not ensure approval by regulatory authorities in other countries, and approval by one or more foreign regulatory authorities does not ensure approval by regulatory authorities in other foreign countries or by the FDA. However, a failure or delay in obtaining regulatory approval in one country may have a negative effect on the regulatory process in others. The foreign regulatory approval process may include all of the risks associated with obtaining FDA approval. We may not obtain foreign regulatory approvals on a timely basis, if at all. We may not be able to file for regulatory approvals and even if we file we may not receive necessary approvals to commercialize our products in any market.

Healthcare reform measures could hinder or prevent our drug candidates' commercial success.

In the United States, there have been and we expect there will continue to be a number of legislative and regulatory changes to the healthcare system in ways that could affect our future revenues and profitability and the future revenues and profitability of our potential customers. Federal and state lawmakers regularly propose and, at times, enact legislation that would result in significant changes to the healthcare system, some of which are intended to contain or reduce the costs of medical products and services. For example, in March 2010, the Affordable Care Act was signed into law. It contained a number of provisions, including those governing enrollment in federal healthcare programs, reimbursement changes and fraud and abuse measures, all of which impacted existing government healthcare programs and resulted in the development of new programs. The Affordable Care Act, among other things:

- imposed a non-deductible annual fee on pharmaceutical manufacturers or importers who sell "branded prescription drugs" to specified federal government programs;
- increased the minimum level of Medicaid rebates payable by manufacturers of brand-name drugs from 15.1% to 23.1%;
- required collection of rebates for drugs paid by Medicaid managed care organizations;
- required manufacturers to participate in a coverage gap discount program, under which they must agree to offer 70% point-of-sale discounts off negotiated prices of applicable brand drugs to eligible beneficiaries during their coverage gap period, as a condition for the manufacturer's outpatient drugs to be covered under Medicare Part D; and
- mandated a further shift in the burden of Medicaid payments to the states.

We expect that the current presidential administration and U.S. Congress will seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Since taking office, President Trump has continued to support the repeal of all or portions of the Affordable Care Act. In December 2017, the Tax Cuts and Jobs Act was enacted, which, among other things, removed penalties for not complying with the individual mandate to carry health insurance. On December 14, 2018, a U.S. District Court Judge in the Northern District of Texas, ruled that the individual mandate is a critical and inseverable feature of the ACA, and therefore, because it was repealed as part of the Tax Act, the remaining provisions of the ACA are invalid as well. While the Trump Administration and CMS have both stated that the ruling will have no immediate effect, it is unclear how this decision, subsequent appeals, if any, and other efforts to repeal and replace the ACA will impact the ACA and our business. There may be additional challenges and amendments to the ACA in the future. There is still uncertainty with respect to the impact President Trump's administration and the U.S. Congress may have, if any, and any changes will likely take time to unfold, and could have an impact on coverage and reimbursement for healthcare items and services covered by plans that were authorized by the Affordable Care Act. However, we cannot predict the ultimate content, timing or effect of any healthcare reform legislation or the impact of potential legislation on us.

Other legislative changes have been proposed and adopted since the Affordable Care Act was enacted. These changes include aggregate reductions to Medicare payments to providers of 2% per fiscal year, which went

into effect in April 2013 and, due to subsequent legislative amendments, will remain in effect through 2027 unless additional Congressional action is taken. In January 2013, the ATRA was enacted, which, among other things, further reduced Medicare payments to several providers, including hospitals, imaging centers and cancer treatment centers, and increased the statute of limitations period for the government to recover overpayments to providers from three to five years. Recently, there has also been heightened governmental scrutiny over the manner in which manufacturers set prices for their marketed products, which has resulted in several Congressional inquiries and proposed bills designed to, among other things, reform government program reimbursement methodologies. These new laws may result in additional reductions in Medicare and other healthcare funding, which could have a material adverse effect on our potential customers and accordingly, our financial operations.

There likely will continue to be legislative and regulatory proposals at the federal and state levels directed at containing or lowering the cost of health care. We cannot predict the initiatives that may be adopted in the future or their full impact, particularly in light of the current presidential administration and U.S. Congress. The continuing efforts of the government, insurance companies, managed care organizations and other payors of healthcare services to contain or reduce costs of health care may adversely affect:

- our ability to set a price we believe is fair for our products;
- our ability to generate revenues and achieve or maintain profitability; and
- the availability of capital.

Further, changes in regulatory requirements and guidance may occur and we may need to amend clinical trial protocols to reflect these changes. Amendments may require us to resubmit our clinical trial protocols to IRBs for reexamination, which may impact the costs, timing or successful completion of a clinical trial. In light of widely publicized events concerning the safety risk of certain drug products, regulatory authorities, members of Congress, the Governmental Accounting Office, medical professionals and the general public have raised concerns about potential drug safety issues. These events have resulted in the recall and withdrawal of drug products, revisions to drug labeling that further limit use of the drug products and establishment of risk management programs that may, for instance, restrict distribution of drug products or require safety surveillance and/or patient education. The increased attention to drug safety issues may result in a more cautious approach by the FDA to clinical trials and the drug approval process. Data from clinical trials may receive greater scrutiny with respect to safety, which may make the FDA or other regulatory authorities more likely to terminate or suspend clinical trials before completion, or require longer or additional clinical trials that may result in substantial additional expense and a delay or failure in obtaining approval or approval for a more limited indication than originally sought.

Even if we are able to commercialize one or more of our drug candidates, the drugs may become subject to unfavorable pricing regulations or third party reimbursement practices, which could harm our business.

Successful sales of our drug candidates, if approved, depend on the availability of adequate coverage and reimbursement from third-party payors. Patients who are provided medical treatment for their conditions generally rely on third-party payors to reimburse all or part of the costs associated with their treatment. Adequate coverage and reimbursement from governmental healthcare programs, such as Medicare and Medicaid in the United States, and commercial payors are critical to new drug acceptance.

Our ability to commercialize any drugs successfully also will depend in part on the extent to which coverage and reimbursement for these drugs and related treatments will be available from government health administration authorities, private health insurers and other organizations. The regulations that govern regulatory approvals, pricing and reimbursement for new therapeutic products vary widely from country to country. Some countries require approval of the sale price of a drug before it can be marketed. In many countries, the pricing review period begins after marketing or licensing approval is granted. In some non-U.S. markets, prescription

pharmaceutical pricing remains subject to continuing governmental control even after initial approval is granted. As a result, we might obtain regulatory approval for a drug in a particular country, but be subject to price regulations that delay our commercial launch of the drug and negatively impact the revenues we are able to generate from the sale of the drug in that country. Adverse pricing limitations may hinder our ability to recover our investment in one or more drug candidates, even if our drug candidates obtain regulatory approval. Government authorities and third-party payors, such as private health insurers and health maintenance organizations, decide which medications they will pay for and establish reimbursement levels. Coverage and reimbursement by a third-party payor may depend upon a number of factors, including the third-party payor's determination that use of a drug is:

- a covered benefit under its health plan;
- safe, effective and medically necessary;
- appropriate for the specific patient;
- cost-effective; and
- neither experimental nor investigational.

We cannot be sure that reimbursement will be available for any drug that we commercialize and, if coverage and reimbursement are available, what the level of reimbursement will be. Reimbursement may impact the demand for, or the price of, any drug for which we obtain regulatory approval. Obtaining reimbursement for our drugs may be difficult because of the higher prices often associated with branded drugs and drugs administered under the supervision of a physician. If reimbursement is not available or is available only at limited levels, we may not be able to successfully commercialize any drug candidate that we successfully develop.

In the United States, no uniform policy of coverage and reimbursement for drugs exists among third-party payors. As a result, obtaining coverage and reimbursement approval of a drug from a government or other third-party payor is a time-consuming and costly process that could require us to provide to each payor supporting scientific, clinical and cost-effectiveness data for the use of our drugs on a payor-by-payor basis, with no assurance that coverage and adequate reimbursement will be obtained. Even if we obtain coverage for a given drug, the resulting reimbursement payment rates might not be adequate for us to achieve or sustain profitability or may require co-payments that patients find unacceptably high. Additionally, third-party payors may not cover, or provide adequate reimbursement for, long-term follow-up evaluations required following the use of our drugs.

If we obtain approval in one or more non-U.S. jurisdictions for our drug candidates, we will be subject to rules and regulations in those jurisdictions. In some non-U.S. countries, the pricing of drugs and biologics is subject to governmental control. In these countries, pricing negotiations with governmental authorities can take considerable time after obtaining regulatory approval of a drug candidate. In addition, market acceptance and sales of our drug candidates will depend significantly on the availability of adequate coverage and reimbursement from third-party payors for our drug candidates and may be affected by existing and future health care reform measures.

If we fail to comply with healthcare regulations, we could face substantial penalties and our business, operations and financial condition could be adversely affected. Additionally, any challenge to or investigation into our practices under these laws could cause adverse publicity and be costly to respond to, and thus could harm our business.

Certain federal and state healthcare laws and regulations pertaining to fraud and abuse and patients' rights are and will be applicable to our business. We could be subject to healthcare fraud and abuse and patient privacy regulation by both the federal government and the states in which we conduct our business. The regulations that may affect our ability to operate include, without limitation:

- the federal Anti-Kickback Statute, which prohibits, among other things, any person from knowingly and willfully offering, soliciting, receiving or providing remuneration, directly or indirectly, to induce

either the referral of an individual, for an item or service or the purchasing or ordering of a good or service, for which payment may be made under federal healthcare programs such as the Medicare and Medicaid programs. A person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation. In addition, the government may assert that a claim including items or services resulting from a violation of the federal Anti-Kickback Statute constitutes a false or fraudulent claim for purposes of the False Claims Act. Violations of the federal Anti-Kickback Statute may result in civil monetary penalties up to $100,000 for each violation, plus up to three times the remuneration involved. Civil penalties for such conduct can further be assessed under the federal False Claims Act. Violations can also result in criminal penalties, including criminal fines of up to $100,000 and imprisonment of up to ten years. Similarly, violations can result in exclusion from participation in government healthcare programs, including Medicare and Medicaid;

- the federal False Claims Act, which prohibits, among other things, individuals or entities from knowingly presenting, or causing to be presented, false claims, or knowingly using false statements, to obtain payment from the federal government including the Medicare and Medicaid or other federal healthcare programs. Private individuals can bring False Claims Act "qui tam" actions, on behalf of the government and such individuals, commonly known as "whistleblowers," may share in amounts paid by the entity to the government in fines or settlement. When an entity is determined to have violated the federal civil False Claims Act, the government may impose civil fines and penalties ranging from $11,181 to $22,363 for each false claim, plus treble damages, and exclude the entity from participation in Medicare, Medicaid and other federal healthcare programs;
- federal criminal laws that prohibit executing a scheme to defraud any healthcare benefit program or making false statements relating to healthcare matters. Similar to the federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation;
- HIPAA, as amended by the Health Information Technology for Economic and Clinical Health Act, and their respective implementing regulations, which impose requirements on certain covered healthcare providers, health plans and healthcare clearinghouses as well as their business associates that perform services for them that involve individually identifiable health information, relating to the privacy, security and transmission of individually identifiable health information without appropriate authorization, including mandatory contractual terms as well as directly applicable privacy and security standards and requirements. Failure to comply with the HIPAA privacy and security standards can result in civil monetary penalties up to $57,051 per violation, not to exceed $1.71 million per calendar year for non-compliance of an identical provision, and, in certain circumstances, criminal penalties with fines up to $250,000 per violation and/or imprisonment. State attorneys general can also bring a civil action to enjoin a HIPAA violation or to obtain statutory damages on behalf of residents of his or her state;
- the federal Physician Sunshine Act, which requires certain applicable manufacturers of drugs, devices, biologics and medical supplies for which payment is available under Medicare, Medicaid or the Children's Health Insurance Program, or CHIP, to report annually to CMS information related to payments and other transfers of value to physicians, which is defined broadly to include other healthcare providers and teaching hospitals, and applicable manufacturers and group purchasing organizations, to report annually ownership and investment interests held by physicians and their immediate family members. Applicable manufacturers are required to submit annual reports to CMS. Failure to submit required information may result in civil monetary penalties of $11,278 per failure up to an aggregate of $169,170 per year (or up to an aggregate of $1.127 million per year for "knowing failures"), for all payments, transfers of value or ownership or investment interests that are not timely, accurately, and completely reported in an annual submission, and may result in liability under other federal laws or regulations; and
- state law equivalents of each of the above federal laws, such as anti-kickback and false claims laws which may apply to items or services reimbursed by any payor, including commercial insurers; state

laws that require pharmaceutical companies to comply with the industry's voluntary compliance guidelines and the applicable compliance guidance promulgated by the federal government, or otherwise restrict payments that may be made to healthcare providers and other potential referral sources; state laws that require drug manufacturers to report information related to payments and other transfers of value to physicians and other healthcare providers or marketing expenditures and pricing information; and state laws governing the privacy and security of health information in certain circumstances, many of which differ from each other in significant ways and may not have the same effect, thus complicating compliance efforts.

In addition, certain states mandate that we comply with a state code of conduct, adopt a company code of conduct under state criteria, disclose marketing payments made to physicians and other healthcare providers, and/or report compliance information to the state authorities. The shifting compliance environment and the need to build and maintain robust and expandable systems to comply in multiple jurisdictions with different compliance and reporting requirements increases the possibility that a pharmaceutical company may run afoul of one or more of the requirements.

If our operations are found to be in violation of any of the laws described above or any other governmental regulations that apply to us, we may be subject to penalties, including civil and criminal penalties, damages, fines, the exclusion from participation in U.S. federal or state health care programs and the curtailment or restructuring of our operations. Any penalties, damages, fines, curtailment or restructuring of our operations could adversely affect our ability to operate our business and our financial results. Any action against us for violation of these laws, even if we successfully defend against it, could cause us to incur significant legal expenses and divert our management's attention from the operation of our business. Moreover, achieving and sustaining compliance with applicable federal and state privacy, security and fraud laws may prove costly.

Risks Related to the Securities Markets and an Investment in Our Stock

There may not be a viable market for our common stock or the price of our common stock may be volatile, and stockholders may not be able to sell their shares at prices that are attractive to them.

There was no public market for our common stock prior to our initial public offering in February 2012, the trading volume of our common stock on the Nasdaq Global Select Market has been limited and there can be no assurance that an active and liquid trading market for our common stock will develop or be sustained. We cannot predict the extent to which investor interest in our company will lead to the development or maintenance of an active trading market on the Nasdaq Global Select Market or otherwise or how liquid that market might become. If an active public market does not develop or is not sustained, it may be difficult for stockholders to sell their shares of common stock at prices that are attractive to them, or at all. Further, an inactive market may also impair our ability to raise capital by selling shares of our common stock and may impair our ability to enter into strategic partnerships or acquire companies or drugs, drug candidates or technologies by using our shares of common stock as consideration.

Stockholders may also be unable to sell their shares of common stock at prices that are attractive to them due to fluctuations in the market price of our common stock. The market prices for securities of biotechnology and pharmaceutical companies have historically been highly volatile. Since the commencement of trading in connection with our initial public offering in February 2012, the publicly traded shares of our common stock have themselves experienced significant price and volume fluctuations. During the year ended December 31, 2018, the price per share for our common stock on the Nasdaq Global Select Market ranged from a low sale price of $5.72 to a high sale price of $15.08. This market volatility is likely to continue. These and other factors could reduce the market price of our common stock, regardless of our operating performance. In addition, the trading price of our common stock could change significantly, both over short periods of time and the longer term, due to

many factors, including, but not limited to, those described elsewhere in this "Risk Factors" section and the following:

- results from, and any delays in, clinical trial programs relating to our drug candidates, including the ongoing and planned clinical trials for avacopan, CCX140, CCX872, and other drug candidates;
- announcements of regulatory approvals or disapprovals of our drug candidates, including avacopan and CCX140, or delays in any regulatory agency review or approval processes;
- failure or discontinuation of any of our research programs;
- announcements relating to future collaborations;
- general economic conditions in the United States and abroad;
- acquisitions and sales of new products, technologies or business;
- delays in the commercialization of any of our drug candidates;
- market conditions in the pharmaceutical, biopharmaceutical and biotechnology sectors;
- the issuance of new or changed securities analysts' reports or recommendations regarding us, our competitors or our industry in general;
- actual and anticipated fluctuations in our quarterly operating results;
- disputes concerning our intellectual property or other proprietary rights;
- introduction of technological innovations or new products by us or our competitors;
- manufacturing issues related to our drug candidates for clinical trials or future products for commercialization;
- market acceptance of our future products;
- deviations in our operating results from the estimates of analysts, or other analyst comments;
- third-party payor coverage and reimbursement policies;
- new legislation in the United States relating to the sale or pricing of pharmaceuticals;
- FDA, EMA or other U.S. or foreign regulatory actions affecting us or our industry;
- product liability claims or other litigation or public concern about the safety of our drug candidates or future drugs;
- our ability to obtain necessary intellectual property licenses;
- the outcome of any future legal actions to which we are party;
- sales of our common stock by our officers, directors or significant stockholders;
- additions or departures of key personnel; and
- external factors, including natural disasters and other crises.

In addition, the stock markets in general, and the markets for pharmaceutical, biopharmaceutical and biotechnology stocks in particular, have experienced extreme volatility that have been often unrelated to the operating performance of the issuer. These broad market fluctuations may adversely affect the trading price or liquidity of our common stock. In the past, when the market price of a stock has been volatile, holders of that stock have sometimes instituted securities class action litigation against the issuer. If any of our stockholders were to bring such a lawsuit against us, we could incur substantial costs defending the lawsuit and the attention of our management would be diverted from the operation of our business.

The ownership of our common stock is highly concentrated, and these stockholders could delay or prevent a change of control.

As of December 31, 2018, our officers and directors, together with holders of 5% or more of our outstanding common stock and their respective affiliates, beneficially owned approximately 61% of our outstanding common stock. Accordingly, these stockholders, acting as a group, have significant influence over the outcome of corporate actions requiring stockholder approval, including the election of directors, any merger, consolidation or sale of all or substantially all of our assets or any other significant corporate transaction. The interests of these stockholders may not be the same as or may even conflict with the interests of our other stockholders. For example, these stockholders could delay or prevent a change of control of our company, even if such a change of control would benefit our other stockholders, which could deprive our stockholders of an opportunity to receive a premium for their common stock as part of a sale of our company or our assets and might affect the prevailing market price of our common stock. The significant concentration of stock ownership may adversely affect the trading price of our common stock due to investors' perception that conflicts of interest may exist or arise.

Future sales of our common stock or securities convertible or exchangeable for our common stock may depress our stock price.

Persons who were our stockholders prior to the sale of shares in our initial public offering continue to hold a substantial number of shares of our common stock that they are able to sell in the public market, subject in some cases to certain legal restrictions. If our stockholders or holders of our options or warrants sell, or indicate an intention to sell, substantial amounts of our common stock in the public market, the trading price of our common stock could decline. The perception in the market that these sales may occur could also cause the trading price of our common stock to decline. As of December 31, 2018, we had 50,652,238 shares of common stock outstanding. Of these shares, approximately 30,421,626 are freely tradeable, without restriction, in the public market. In addition, approximately 12,864,960 of the outstanding shares of common stock, and an additional 150,000 shares of common stock issuable upon exercise of outstanding warrants that we issued to Bio-Techne Corporation (formerly Techne Corporation), or Bio-Techne, in connection with our initial public offering, are eligible for sale in the public market, subject to volume limitations under Rule 144 under the Securities Act of 1933, as amended, or the Securities Act, with respect to shares held by directors, executive officers and other affiliates. In addition, shares of common stock that are either subject to outstanding options or reserved for future issuance under our employee benefit plans are eligible for sale in the public market to the extent permitted by the provisions of various vesting schedules and Rule 144 and Rule 701 under the Securities Act and, in any event, we have filed a registration statement permitting shares of common stock issued on exercise of options to be freely sold in the public market. In addition, as of February 28, 2019, we had the remaining capacity to sell up to an additional $55.0 million of our common stock from time to time under our Equity Distribution Agreement, or EDA. If additional shares of common stock are sold, or if it is perceived that they will be sold, in the public market, the trading price of our common stock could decline.

Certain of our directors and executive officers have established, programmed selling plans under Rule 10b5-1 of the Exchange Act, for the purpose of effecting sales of our common stock. Any sales of securities by these stockholders, or the perception that those sales may occur, including the entry into such programmed selling plans, could have a material adverse effect on the trading price of our common stock.

If we sell shares of our common stock in future financings, common stockholders may experience immediate dilution and, as a result, our stock price may decline.

We may from time to time issue additional shares of common stock at a discount from the current trading price of our common stock. As a result, our common stockholders would experience immediate dilution upon the purchase of any shares of our common stock sold at such discount. In addition, as opportunities present themselves, we may enter into financing or similar arrangements in the future, including the issuance of debt securities, preferred stock or common stock. For example, in connection with our initial public offering, in

February 2012, we issued Bio-Techne a warrant with a ten-year term to purchase up to 150,000 shares of our common stock at an exercise per share equal to 200% of the initial public offering price of a share of our common stock and such warrant, if exercised, would likely be exercised at a time when the exercise price of such warrant represented a discount to the trading price of our common stock. In addition, pursuant to our collaboration and license agreement with Vifor in May 2016 for the commercialization of avacopan, we entered into a stock purchase agreement with Vifor for the purchase of 3,333,333 unregistered shares of our common stock at a price of $7.50 per share. Similarly, in December 2018, our EDA would allow us to sell up to an additional $75.0 million of shares of our common stock from time to time. In January 2019, we sold 1,666,367 shares of our common stock pursuant to the EDA at a price per share of $12.00, for gross proceeds of $20.0 million, before deducting offering-related transaction costs and commissions. If we issue common stock or securities convertible into common stock, our common stockholders would experience additional dilution and, as a result, our stock price may decline.

Our quarterly operating results may fluctuate significantly or may fall below the expectations of investors or securities analysts, each of which may cause our stock price to fluctuate or decline.

We expect our operating results to be subject to quarterly fluctuations. Our net loss and other operating results will be affected by numerous factors, including:

- variations in the level of expenses related to our drug candidates or future development programs;
- if any of our drug candidates receives regulatory approval, the level of underlying demand for these drug candidates and wholesalers' buying patterns;
- addition or termination of clinical trials or funding support;
- our execution of any collaborative, licensing or similar arrangements, and the timing of payments we may make or receive under such arrangements, or the termination of such arrangements;
- any intellectual property infringement lawsuit in which we may become involved;
- regulatory developments affecting our drug candidates or those of our competitors; and
- our ability to secure new government contracts and allocation of our resources to or away from performing work under government contracts.

If our quarterly operating results fall below the expectations of investors or securities analysts, the price of our common stock could decline substantially. Furthermore, any quarterly fluctuations in our operating results may, in turn, cause the price of our stock to fluctuate substantially. We believe that quarterly comparisons of our financial results are not necessarily meaningful and should not be relied upon as an indication of our future performance.

We have broad discretion in the use of our cash and may not use it effectively.

Our management has broad discretion over the use of our cash. Because of the number and variability of factors that will determine our use of cash, stockholders may not agree with how we allocate or spend our cash. We may pursue collaborations or clinical trials that do not result in an increase in the market value of our common stock and that may increase our losses, or we may place our cash in investments that do not produce significant investment returns or that may lose value. Our failure to allocate and spend our cash effectively would have a material adverse effect on our financial condition and business and could cause our stock price to decline.

Provisions of our charter documents or Delaware law could delay or prevent an acquisition of our company, even if the acquisition would be beneficial to our stockholders, and could make it more difficult for our stockholders to change management.

Provisions of our amended and restated certificate of incorporation and amended and restated bylaws may discourage, delay or prevent a merger, acquisition or other change in control that stockholders may consider

favorable, including transactions in which stockholders might otherwise receive a premium for their shares. In addition, these provisions may frustrate or prevent any attempt by our stockholders to replace or remove our current management by making it more difficult to replace or remove our board of directors. These provisions include:

- a classified board of directors so that not all directors are elected at one time;
- a prohibition on stockholder action through written consent;
- a requirement that special meetings of stockholders be called only by the chairman of the board of directors, the chief executive officer, the president or by the board of directors;
- an advance notice requirement for stockholder proposals and nominations;
- the authority of our board of directors to issue preferred stock with such terms as our board of directors may determine; and
- a requirement of approval of not less than 66 2/3% of all outstanding shares of our capital stock entitled to vote to amend any bylaws by stockholder action, or to amend specific provisions of our certificate of incorporation.

In addition, Delaware law prohibits a publicly held Delaware corporation from engaging in a business combination with an interested stockholder, generally a person who, together with its affiliates, owns or within the last three years has owned 15% of our voting stock, for a period of three years after the date of the transaction in which the person became an interested stockholder, unless the business combination is approved in a prescribed manner. Accordingly, Delaware law may discourage, delay or prevent a change in control of our company.

Our employment agreements with our named executive officers may require us to pay severance benefits to any of those persons who are terminated in connection with a change of control of us, which could harm our financial condition or results.

Our named executive officers are parties to employment agreements providing for aggregate cash payments of up to approximately $5.2 million for severance and other benefits and acceleration of vesting of stock awards with an intrinsic value of $10.0 million as of December 31, 2018 in the event of a termination of employment in connection with a change of control of us. The accelerated vesting of options could result in dilution to our stockholders and harm the market price of our common stock. The payment of these severance benefits could harm our financial condition and results. In addition, these potential severance payments may discourage or prevent third parties from seeking a business combination with us.

We do not anticipate paying any cash dividends on our capital stock in the foreseeable future, therefore capital appreciation, if any, of our common stock will be our stockholders' sole source of gain for the foreseeable future.

We have never declared or paid cash dividends on our capital stock. We do not anticipate paying any cash dividends on our capital stock in the foreseeable future. In addition, our ability to pay dividends is currently restricted by the terms of our credit facility with Hercules. We currently intend to retain all available funds and any future earnings to fund the development and growth of our business. Further, any future debt financing arrangement may contain additional terms prohibiting or limiting the amount of dividends that may be declared or paid on our common stock. As a result, capital appreciation, if any, of our common stock will be our stockholders' sole source of gain for the foreseeable future.

If securities or industry analysts do not publish research, or publish inaccurate or unfavorable research, about our business, our stock price and trading volume could decline.

The trading market for our common stock depends, in part, on the research and reports that securities or industry analysts publish about us or our business. As of January 2019, we had research coverage by five

securities analysts. In the event one or more of the analysts who covers us downgrades our stock or publishes inaccurate or unfavorable research about our business, our stock price would likely decline. In addition, if our operating results fail to meet the forecast of analysts, our stock price would likely decline. If one or more of these analysts cease coverage of our company or fail to publish reports on us regularly, demand for our stock could decrease, which might cause our stock price and trading volume to decline.

The results of the United Kingdom's referendum on withdrawal from the European Union may have a negative effect on global economic conditions, financial markets and our business.

In June 2016, a majority of voters in the United Kingdom elected to withdraw from the EU in a national referendum. The referendum was advisory, and the terms of any withdrawal are subject to a negotiation period that could last at least until March 2019. Nevertheless, the referendum has created significant uncertainty about the future relationship between the United Kingdom and the EU, including with respect to the laws and regulations that will apply as the United Kingdom determines which EU laws to replace or replicate in the event of a withdrawal. The referendum has also given rise to calls for the governments of other EU member states to consider withdrawal. These developments, or the perception that any of them could occur, have had and may continue to have a material adverse effect on global economic conditions and the stability of global financial markets, and may significantly reduce global market liquidity and restrict the ability of key market participants to operate in certain financial markets. Any of these factors could depress economic activity and restrict our access to capital, which could have a material adverse effect on our business, financial condition and results of operations and reduce the price of our common stock.

Item 1B. Unresolved Staff Comments.

Not applicable.

Item 2. Properties.

Our corporate headquarters are located in Mountain View, California, where we lease 35,755 square feet of office and laboratory space. In April 2004, we entered into a ten-year lease agreement for that facility. In August 2012, we entered into an amendment to the lease agreement for the same facility to extend the term of the lease through April 2019. In April 2017, we entered into a second amendment to the lease agreement for the same facility to extend the term of the lease through April 2020.

We believe that our existing facilities are adequate for our current needs, as the facility has sufficient laboratory space to house additional scientists to be hired as we expand. When our leases expire, we may exercise our renewal options or look for additional or alternate space for our operations and we believe that suitable additional or alternative space will be available in the future on commercially reasonable terms.

Item 3. Legal Proceedings.

We are not currently a party to any legal proceedings.

Item 4. Mine Safety Disclosures.

Not Applicable.

PART II

Item 5. Market for Registrant's Common Equity, Related Stockholder Matters and Issuer Purchases of Equity Securities.

Market Information

Our common stock is traded on the Nasdaq Global Select Market under the symbol "CCXI."

Holders of Common Stock

As of February 28, 2019, there were approximately 42 holders of record of our common stock. Certain shares are held in "street" name and accordingly, the number of beneficial owners of such shares is not known or included in the foregoing number.

Dividend Policy

We have never declared or paid cash dividends on our capital stock. We intend to retain all available funds and any future earnings, if any, to fund the development and expansion of our business and we do not anticipate paying any cash dividends in the foreseeable future. Any future determination related to dividend policy will be made at the discretion of our board of directors. In addition, our ability to pay dividends is currently restricted by the terms of our credit facility with Hercules.

Equity Compensation Plan Information

The following table summarizes securities available under our equity compensation plans as of December 31, 2018:

Plan Category	Shares Issuable Upon Exercise of Outstanding Options, Warrants and Rights(2)	Weighted-Average Exercise Price of Outstanding Options, Warrants and Rights(3)	Number of Securities Available for Future Issuance(4)
Equity compensation plans approved by security holders: (1)	11,160,554	$ 8.39	2,103,067
Equity compensation plans not approved by security holders:	—	—	—
Total	11,160,554	$ 8.39	2,103,067

(1) Consists of our Amended and Restated 1997 Stock Option/Stock Issuance Plan, our Amended and Restated 2002 Equity Incentive Plan and our 2012 Equity Incentive Award Plan, our Non-Employee Director Compensation Policy and our Employee Stock Purchase Plan, or ESPP.

(2) Includes 10,720,200 shares subject to outstanding stock options and 440,354 shares subject to outstanding restricted stock units as of December 31, 2018.

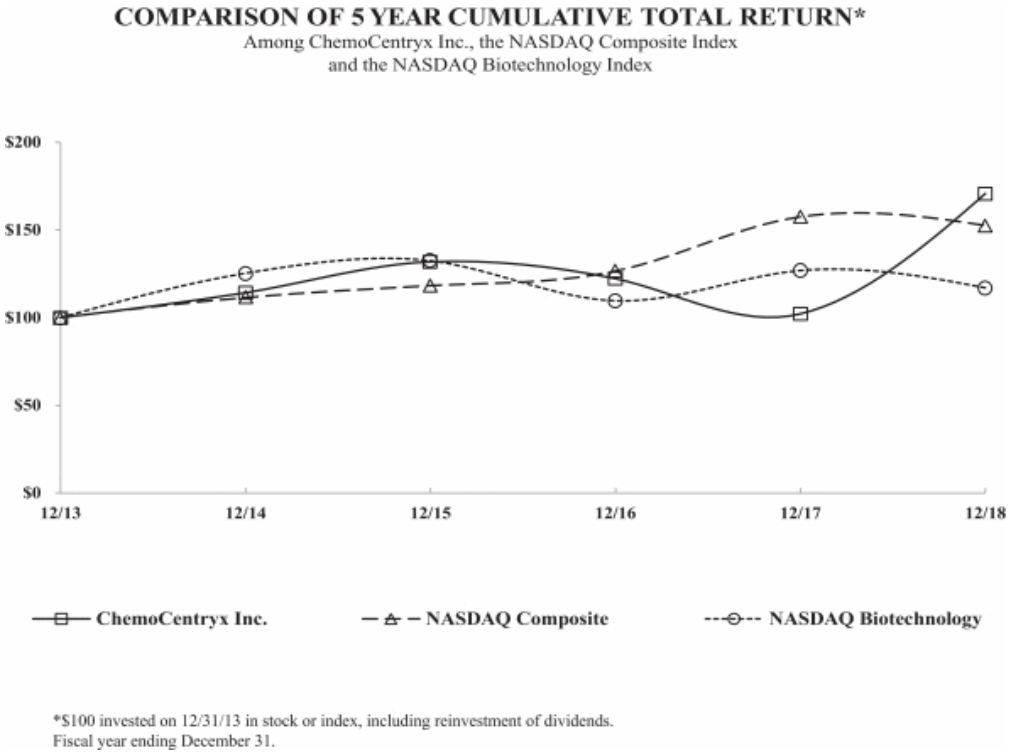
(3) Calculated exclusive of outstanding restricted stock unit awards.

(4) Of these shares, 1,708,516 shares were available for stock option awards, restricted stock units and restricted stock awards, and 394,551 were available for the ESPP, in each case as of December 31, 2018.

Performance Graph

The information contained in this Performance Graph section shall not be deemed "soliciting material" or to be "filed" with the SEC, for purposes of Section 18 of the Securities Exchange Act of 1934, as amended, or the Exchange Act, or otherwise subject to the liabilities under that Section, and shall not be deemed to be incorporated by reference into any filing of ChemoCentryx, Inc. under the Securities Act of 1933, as amended, or the Exchange Act.

The following graph shows a comparison from December 31, 2013 (the date our common stock commenced trading on the Nasdaq Global Select Market) through December 31, 2018 of cumulative total return for our common stock, the Nasdaq Composite Index and the Nasdaq Biotechnology Index. Such returns are based on historical results and are not intended to suggest future performance. Data for the Nasdaq Composite Index and the Nasdaq Biotechnology Index assume reinvestment of dividends.



	12/13	12/14	12/15	12/16	12/17	12/18
ChemoCentryx Inc.	**100.00**	**117.96**	**139.90**	**127.81**	**102.76**	**188.43**
NASDAQ Composite	**100.00**	**114.62**	**122.81**	**133.19**	**172.11**	**165.84**
NASDAQ Biotechnology	**100.00**	**131.71**	**140.56**	**112.25**	**133.67**	**121.24**

Item 6. Selected Financial Data.

The following selected financial data have been derived from our audited financial statements. The information set forth below is not necessarily indicative of future results and should be read in conjunction with "Item 7. Management's Discussion and Analysis of Financial Condition and Results of Operations" and "Item 8. Financial Statements and Supplementary Data" included elsewhere in this Annual Report on Form 10-K.

	Year Ended December 31,				
	2018	2017	2016	2015	2014
	(in thousands, except share and per share data)				
Consolidated Statements of Operations Data:					
Revenue:					
Collaboration and license revenue from related party	$ 42,875	$ 82,497	$ 11,435	$ —	$ —
Grant revenue	—	—	500	—	—
Total revenue	42,875	82,497	11,935	—	—
Operating expenses:					
Research and development	62,736	49,495	37,945	33,183	33,815
General and administrative	20,409	16,509	14,710	14,506	13,584
Total operating expenses	83,145	66,004	52,655	47,689	47,399
Income (loss) from operations	(40,270)	16,493	(40,720)	(47,689)	(47,399)
Interest income	3,528	1,370	757	384	494
Interest expense	(1,224)	(4)	—	—	(24)
Net income (loss)	$ (37,966)	$ 17,859	$ (39,963)	$ (47,305)	$ (46,929)
Net income (loss) per share, basic (1)	$ (0.76)	$ 0.37	$ (0.86)	$ (1.08)	$ (1.08)
Net income (loss) per share, diluted (1)	$ (0.76)	$ 0.36	$ (0.86)	$ (1.08)	$ (1.08)
Shares used to compute net income (loss) per share, basic	49,814,162	48,412,531	46,431,501	43,889,677	43,275,276
Shares used to compute net income (loss) per share, diluted	49,814,162	49,615,406	46,431,501	43,889,677	43,275,276

(1) See Note 2 within the notes to our audited consolidated financial statements appearing elsewhere in this Annual Report on Form 10-K for a description of the method used to compute basic and diluted net income (loss) per share.

	As of December 31,				
	2018	2017	2016	2015	2014
			(in thousands)		
Consolidated Balance Sheets Data:					
Cash, cash equivalents and investments	$ 176,984	$ 135,220	$ 123,761	$ 76,289	$ 114,620
Accounts receivable from related party(2)	2,058	51,090	30,205	—	—
Working capital	116,988	146,893	110,356	66,541	66,139
Total assets	183,310	189,328	155,872	78,155	116,981
Long-term debt, net	19,689	4,676	—	—	—
Accumulated deficit	(374,497)	(289,200)	(307,059)	(267,096)	(219,791)
Total stockholders' equity	14,738	79,267	49,889	72,507	108,606

(2) As of December 31, 2017, accounts receivable excluded the remaining $10.0 million cash commitment due from Vifor in February 2018 in connection with the Avacopan Amendment. As of December 31, 2016, accounts receivable excluded the additional $20.0 million cash commitment due from Vifor in December 2017 in connection with the CCX140 Agreement. See "Note 9. Related-Party Transactions" for a detailed discussion.

Item 7. Management's Discussion and Analysis of Financial Condition and Results of Operations.

You should read the following discussion and analysis of financial condition and results of operations together with "Item 6. Selected Financial Data" and our financial statements and related notes included elsewhere in this Annual Report on Form 10-K. This discussion and other parts of this Annual Report on Form 10-K contain forward-looking statements that involve risk and uncertainties, such as statements of our plans, objectives, expectations and intentions. Our actual results could differ materially from those discussed in these forward-looking statements. Factors that could cause or contribute to such differences include, but are not limited to, those discussed in "Item 1A. Risk Factors" of this Annual Report on Form 10-K.

Overview

ChemoCentryx is a biopharmaceutical company developing new medications targeted at inflammatory disorders, autoimmune diseases and cancer. Each of our drug candidates is designed to selectively block a specific chemoattractant receptor, leaving the rest of the immune system intact. Our drug candidates are small molecules, which are orally administered, and, if approved, could address unmet medical needs, including improved efficacy, and offer significant quality of life benefits, since patients swallow a capsule or pill instead of having to visit a clinic for an infusion or undergo an injection.

In 2016, we executed on our strategy to form an alliance with a partner that could provide upfront fees and milestone payments to support the clinical development of our two leading drug candidates, avacopan and CCX140, to registration and pay us royalties upon sales in international markets, while we develop our own commercial infrastructure to sell directly in the United States.

To help communicate the breadth of our drug discovery platform, we have segmented our pipeline into early stage and late stage drug candidates.

Late Stage Drug Candidates

We have chosen to focus initially on orphan indications, where drug candidates tend to enjoy a faster path to market and better reimbursement. Our leading drug candidates address areas of clear unmet need, where the current standard of care, or SOC, is insufficient to halt progression of the disease and/or where today's treatment options come with serious side effects, such as those which accompany the prolonged use of steroids:

Avacopan (CCX168) — Inhibition of Complement-Mediated Pathways in Orphan Diseases

Avacopan (formerly CCX168) is an orally-administered complement inhibitor targeting the C5a receptor, or C5aR, and is being developed for orphan diseases, including (i) anti-neutrophil cytoplasmic auto-antibody associated vasculitis, or AAV, a devastating autoimmune disease that damages blood vessels and can lead to kidney failure, pulmonary failure and damage to other tissues; (ii) complement 3 glomerulopathy, or C3G, a debilitating disease that can lead to kidney failure; and (iii) moderate and severe hidradenitis suppurativa, or HS, a chronic, inflammatory, debilitating skin disease characterized by recurrent, painful, nodules and abscesses, ultimately leading to the formation of draining fistulas (also known as sinus tracts) as well as scarring.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV and C3G and by the European Medicines Agency, or EMA, for the treatment of C3G and microscopic polyangiitis and granulomatosis with polyangiitis, both forms of AAV. Additionally, avacopan has been granted PRIority MEdicines, or PRIME, designation from the EMA, to expedite its clinical development, and to potentially accelerate its marketing authorization.

Following completion of two Phase II clinical trials in patients with AAV, in which avacopan was well-tolerated and provided effective steroid-free control of the disease, we launched the Phase III ADVOCATE

trial in December 2016. The FDA and the EMA concurred with the design of the study. ADVOCATE is a randomized, double-blind two-arm study which enrolled over 300 patients at over 200 sites in the United States, Canada, Europe, Australia, New Zealand and Japan. Patient enrollment of the Phase III ADVOCATE trial was completed in July 2018 and we expect to report topline data from this trial in the fourth quarter of 2019. Additionally, we launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G for which we aim to complete enrollment in 2019 and initiated a large placebo-controlled Phase IIb clinical trial, the AURORA trial, for the treatment of patients with moderate to severe HS in the fourth quarter of 2018.

CCX140 — Chronic and Orphan Kidney Diseases

CCX140, an orally-administered inhibitor of the chemokine receptor known as CCR2, has been in development for diabetic nephropathy, or DN, a form of chronic kidney disease, or CKD, and is now being developed for focal segmental glomerulosclerosis, or FSGS, a rare renal disease characterized by progressive proteinuria, excess protein in the urine, and impaired renal function. CCX140 has been granted orphan drug designation by the FDA for the treatment of FSGS.

A global Phase II clinical trial of CCX140 in patients with DN met its primary endpoint by demonstrating that CCX140 given orally once-daily added to a SOC renin-angiotensin-aldosterone system inhibitor treatment resulted in a statistically significant reduction in proteinuria, beyond that achieved with SOC alone, with the most pronounced effect shown in the patients with highest levels of proteinuria. Based on the safety and efficacy data related to reduction in proteinuria observed in the Phase II trial in DN, we launched two Phase IIb clinical trials (the LUMINA trials) of CCX140 for the treatment of primary FSGS, with and without nephrotic syndrome, for which there are currently no FDA-approved treatments.

Kidney Health Alliance with Vifor

In May 2016, we announced a partnership, which we refer to as the Avacopan Agreement, with Vifor (International) Ltd., and/or its affiliates, or collectively, Vifor, a European-based world leader specializing in kidney disease. While under this agreement we retained all rights to the United States and China, we granted Vifor exclusive commercialization rights to avacopan in Europe and certain other international markets. In December 2016, we entered into an additional agreement with Vifor, which we refer to as the CCX140 Agreement, relating to CCX140, our other late stage drug candidate. Under the CCX140 Agreement, we again retained all rights to the United States and China and we granted Vifor exclusive worldwide commercialization rights outside of the United States and China. In February 2017, we announced a further agreement with Vifor that harmonized the geographic commercialization rights underlying the agreements for both drug candidates, which we refer to as the Avacopan Amendment. In June 2018, we entered into additional agreements with Vifor to further expand Vifor's exclusive commercialization rights to include China under the Avacopan Agreement (the Avacopan Letter Agreement) and the CCX140 Agreement (CCX140 Letter Agreement).

We have secured $215 million in upfront cash and milestone payments pursuant to our agreements with Vifor and are eligible for additional substantial milestone payments. Through our alliance, we maintain the commercialization rights to avacopan and CCX140 in the United States, and also retain control of the clinical development programs for orphan renal disease. Vifor gained the exclusive commercialization rights for all other international markets, and is obligated to pay us tiered royalties, with rates ranging from ten to the mid-twenties, on potential net sales.

At a future time defined in the CCX140 Agreement, Vifor has an option to solely develop and commercialize CCX140 in more prevalent forms of CKD. Should Vifor later exercise the CKD option, we would receive co-promotion rights for CKD in the United States, and we estimate that the clinical development and registration process for CKD would end at approximately the same time as Orphan Drug exclusivity.

Early Stage Drug Candidates

While we have focused initially on kidney disease, our target-specific and selective approach designed to stop the spread of inflammatory disease-inducing cells shows promise in other disease areas. Over time we plan to bring forward drug candidates to treat a range of inflammatory and autoimmune disorders, as well as cancer, where our drug candidate CCX872 has shown promise in a Phase Ib trial for advanced pancreatic cancer. We expect that our ability to do so will grow as we increase our scale and to the extent that we start to earn revenues and royalties from the commercialization of our late stage kidney disease franchise.

Since commencing our operations in 1997, our efforts have focused on research, development and the advancement of our drug candidates into and through clinical trials. As a result, we have incurred significant losses. We have funded our operations primarily through the sale of convertible preferred and common stock, contract revenue under our collaborations, government contracts and grants and borrowings under equipment financing arrangements.

As of December 31, 2018, we had an accumulated deficit of $374.5 million. We expect to continue to incur net losses as we develop our drug candidates, expand clinical trials for our drug candidates currently in clinical development, expand our research and development activities, expand our systems and facilities, seek regulatory approvals and engage in commercialization preparation activities in anticipation of FDA approval of our drug candidates. In addition, if a product is approved for commercialization, we will need to expand our organization. Significant capital is required to launch a product and many expenses are incurred before revenues are received. We are unable to predict the extent of any future losses or when we will become profitable, if at all.

Recent Developments and Corporate Highlights

Equity Distribution Agreement with Piper Jaffray & Co.

In December 2018, we entered into an Equity Distribution Agreement, or EDA, with Piper Jaffray & Co., or Piper Jaffray, having an aggregate offering price of up to $75.0 million. Through this EDA, we may offer and sell, from time to time, through Piper Jaffray, shares of our common stock, par value $0.001 per share. In January 2019, we sold 1,666,367 shares of our common stock pursuant to the EDA at a price per share of $12.00, for gross proceeds of $20.0 million, before deducting offering-related transaction costs and commissions.

HS Phase IIb Clinical Study Initiation

In December 2018, we launched a large placebo-controlled Phase IIb clinical trial of avacopan for the treatment of patients with HS, the AURORA trial. The AURORA trial aims to enroll 390 patients with moderate to severe HS. The primary endpoint will assess avacopan against placebo at 12 weeks of treatment, using the HiSCR (hidradenitis suppurativa clinical response) scale, which has been validated by the FDA.

Critical Accounting Policies and Significant Judgments and Estimates

The preparation of our consolidated financial statements requires us to make estimates and assumptions that affect the reported amounts of assets and liabilities and the disclosure of contingent assets and liabilities at the date of our financial statements as well as the reported revenues and expenses during the reported periods. We base our estimates on historical experience and on various other factors that we believe are reasonable under the circumstances, the results of which form the basis for making judgments about the carrying value of assets and liabilities that are not apparent from other sources. Actual results may differ from these estimates under different assumptions or conditions.

While our significant accounting policies are described in the Notes to our consolidated financial statements appearing elsewhere in this Annual Report on Form 10-K, we believe that the following critical accounting policies relating to revenue recognition, clinical trial expenses and stock-based compensation are most important to understanding and evaluating our reported financial results.

Revenue Recognition

Effective January 1, 2018, we adopted Accounting Standards Codification, or ASC, Topic 606, *Revenue from Contracts with Customers,* or ASC 606, using the modified retrospective transition method. This standard applies to all contracts with customers, except for contracts that are within the scope of other standards, such as leases, insurance, collaboration arrangements and financial instruments. Under ASC 606, we recognize revenue when our customer obtains control of promised goods or services, in an amount that reflects the consideration which we expect to receive in exchange for those goods or services. To determine revenue recognition for arrangements that we determine are within the scope of ASC 606, we perform the following five steps: (i) identify the contract(s) with a customer; (ii) identify the performance obligations in the contract; (iii) determine the transaction price; (iv) allocate the transaction price to the performance obligations in the contract; and (v) recognize revenue when (or as) we satisfy a performance obligation. We only apply the five-step model to contracts when it is probable that we will collect the consideration we are entitled to in exchange for the goods or services we transfer to the customer. At contract inception, once the contract is determined to be within the scope of ASC 606, we assess the goods or services promised within each contract and determine those that are performance obligations and assess whether each promised good or service is distinct. We then recognize as revenue the amount of the transaction price that is allocated to the respective performance obligation when (or as) the performance obligation is satisfied.

We enter into corporate collaborations under which we may obtain upfront license fees, research and development funding and development and regulatory and commercial milestone payments and royalty payments. Our performance obligations under these arrangements may include licenses of intellectual property, distribution rights, research and development services, delivery of manufactured product, and/or participation on joint steering committees.

Licenses of intellectual property: If the license to our intellectual property is determined to be distinct from the other performance obligations identified in the arrangement, we recognize revenue from upfront license fees allocated to the license when the license is transferred to the customer and the customer is able to use and benefit from the license. For licenses that are bundled with other promises, we utilize judgement to assess the nature of the combined performance obligation to determine whether the combined performance obligation is satisfied over time or at a point in time and, if over time, the appropriate method of measuring proportional performance for purposes of recognizing revenue from non-refundable, up-front fees. We evaluate the measure of proportional performance each reporting period and, if necessary, adjust the measure of performance and related revenue recognition.

Milestone payments: At the inception of each arrangement that includes development, regulatory or commercial milestone payments, we evaluate whether the milestones are considered probable of being reached and estimate the amount to be included in the transaction price. ASC 606 suggests two alternatives to use when estimating the amount of variable consideration: the expected value method and the most likely amount method. Under the expected value method, an entity considers the sum of probability-weighted amounts in a range of possible consideration amounts. Under the most likely amount method, an entity considers the single most likely amount in a range of possible consideration amounts. Whichever method is used, it should be consistently applied throughout the life of the contract; however, it is not necessary for us to use the same approach for all contracts. We expect to use the most likely amount method for development and regulatory milestone payments. If it is probable that a significant revenue reversal would not occur, the associated milestone value is included in the transaction price. Milestone payments that are not within our or the licensee's control, such as regulatory approvals, are not considered probable of being achieved until those approvals are received. The transaction price is then allocated to each performance obligation on a relative stand-alone selling price basis. We recognize revenue as or when the performance obligations under the contract are satisfied. At the end of each subsequent reporting period, we re-evaluate the probability of achievement of each such milestone and any related constraint, and if necessary, adjust our estimates of the overall transaction price. Any such adjustments are recorded on a cumulative catch-up basis, which would affect revenues and earnings in the period of adjustment.

Commercial milestones and royalties: For arrangements that include sales-based royalties, including milestone payments based on the level of sales, and in which the license is deemed to be the predominant item to which the royalties relate, we recognize revenue when the related sales occur. To date, we have not recognized any royalty revenue resulting from our collaboration arrangements.

Up-front payments and fees are recorded as deferred revenue upon receipt or when due, and may require deferral of revenue recognition to a future period until we perform our obligations under these arrangements. Amounts payable to us are recorded as accounts receivable when our right to consideration is unconditional.

Clinical Trial Accruals and Related Expenses

We accrue and recognize expenses for clinical trial activities performed by third parties, including clinical research organizations, or CROs, and clinical investigators, based upon estimates made as of the reporting date of the work completed over the life of the individual trial in accordance with agreements established with CROs and clinical trial sites. Some CROs invoice us on a monthly basis, while others invoice upon milestones achieved and the expense is recorded as services are rendered. We determine the estimates of clinical activities incurred at the end of each reporting period through discussion with internal personnel and outside service providers as to the progress or stage of completion of trials or services, as of the end of each reporting period, pursuant to contracts with numerous clinical trial centers and CROs and the agreed upon fee to be paid for such services. The significant factors considered in estimating accruals include the number of patients enrolled and the percentage of work completed to date. Costs of setting up clinical trial sites for participation in the trials that are paid for in advance are expensed over the estimated set-up period. While the set-up periods vary from one arrangement to another, such set-up periods generally take from two to six months. Such set-up activities include clinical site identification, local ethics committee submissions, regulatory submissions, clinical investigator kick-off meetings and pre-study site visits. Clinical trial site costs related to patient enrollments are accrued as patients are entered into the trial.

To date, we have not experienced significant changes in our estimates of clinical trial accruals after a reporting period. However, due to the nature of estimates, we cannot assure you that we will not make changes to our estimates in the future as we become aware of additional information about the status or conduct of our clinical trials.

Stock-Based Compensation

Stock-based compensation cost is measured at the grant date, based on the fair value of the award, and is recognized as an expense over the employee's requisite service period on a straight line basis. The fair value of the stock options is estimated using the Black-Scholes valuation model. We recorded non-cash stock-based compensation expense of $10.8 million, $8.7 million and $8.5 million for the years ended December 31, 2018, 2017 and 2016, respectively. At December 31, 2018 and 2017, we had $15.0 million and $8.2 million, respectively, of total unrecognized stock-based compensation expense, net of estimated forfeitures, related to employee stock options that will be recognized over a weighted-average period of 2.5 years for both years. We expect to continue to grant stock options in the future, and to the extent that we do, our actual stock-based compensation expense recognized in future periods will likely increase. Determining an estimate of the fair value of equity awards using the Black-Scholes valuation model requires that use of subjective assumptions related to expected stock price volatility, term, risk-free interest rate and dividend yield.

Results of Operations

Revenue

We have not generated any revenue from product sales. For the years ended December 31, 2018 and 2017, our revenues were derived from collaboration and license revenue related to the Avacopan Agreement and

CCX140 Agreement, in each case, as amended, and the related letter agreements. For the year ended December 31, 2016, our revenues were derived from the Avacopan Agreement, as well as grant revenue from the FDA Orphan Products Development grant to support the clinical development of avacopan for the treatment of patients with AAV.

Total revenues were as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Collaboration and license revenue from related party	$ 42,875	$82,497	$11,435
Grant revenue	—	—	500
Total revenue	$ 42,875	$82,497	$11,935
Dollar increase (decrease)	$(39,622)	$70,562	
Percentage increase (decrease)	-48%	591%	

On January 1, 2018, we adopted ASC 606 under the modified retrospective transition method and recognized the cumulative effect of initially applying the new revenue standard of $47.3 million as an adjustment to the opening balance of accumulated deficit and an increase in deferred revenue. Revenue recognized prior to January 1, 2018 has not been restated and continues to be reported under the accounting standards in effect for those periods.

We use a cost-based input method to measure proportional performance and to calculate the corresponding amount of revenue to recognize in 2018. In applying the cost-based input method of revenue recognition, we measure actual costs incurred relative to budgeted costs to fulfill the combined performance obligation. These costs consist primarily of third-party contract costs. Revenue is recognized based on actual costs incurred as a percentage of total budgeted costs as we complete our performance obligations.

Before the adoption of ASC 606, we recognized upfront fees straight-line under ASC 605 over the estimated performance period and recognized milestones when earned under the milestone method of accounting. In 2017, revenue recognized represents amortization of the upfront license fee commitments from Vifor pursuant to the Avacopan Agreement and CCX140 Agreement, in each case, as amended. The increase in revenue from 2016 to 2017 was due to: (i) recognition of a milestone payment related to the Avacopan Agreement; (ii) amortization of the upfront license fee commitments from Vifor pursuant to the Avacopan Agreement, Avacopan Amendment and CCX140 Agreement, as well as (iii) collaboration revenue for development services under the CCX140 Agreement in 2017. These increases were partially offset by a decrease in grant revenue from the FDA to support the clinical development of avacopan for the treatment of patients with AAV.

The revenue in 2016 was due to: (i) amortization of the upfront payment from Vifor pursuant to the Avacopan Agreement over the service period, which began in May 2016 and (ii) grant revenue from the FDA to support the clinical development of avacopan for the treatment of patients with AAV.

Research and development expenses

Research and development expenses represent costs incurred to conduct basic research, the discovery and development of novel small molecule therapeutics, development of our suite of proprietary drug discovery technologies, preclinical studies and clinical trials of our drug candidates. We recognize all research and development expenses as they are incurred. These expenses consist primarily of salaries and related benefits, including stock-based compensation, third-party contract costs relating to research, formulation, manufacturing, preclinical study and clinical trial activities, laboratory consumables, and allocated facility costs. Total research and development expenses, as compared to the prior years, were as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Research and development expenses	$62,736	$49,495	$37,945
Dollar increase	$13,241	$11,550	
Percentage increase	27%	30%	

The increase in research and development expenses from 2017 to 2018 was primarily due to the advancement of the avacopan ADVOCATE Phase III pivotal trial which completed enrollment in July 2018, initiation and patient enrollment of the avacopan Phase II clinical trial in patients with C3G and start-up, initiation and patient enrollment of the avacopan Phase IIb clinical trial in patients with HS and the CCX140 Phase II clinical trials in patients with FSGS.

The increase in research and development expenses from 2016 to 2017 was primarily due to the initiation and patient enrollment of the avacopan Phase III ADVOCATE trial in patients with AAV and start-up expenses related to the Phase II clinical trials of FSGS and C3G. These increases were partially offset by lower Phase II clinical development expenses primarily due to the completion of avacopan CLEAR and CLASSIC clinical trials for the treatment of AAV in 2016 and lower Phase I development expense due to the completion of enrollment in the clinical trial for CCX872 in patients with advanced pancreatic cancer in 2016.

The following table summarizes our research and development expenses by project (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Phase I	$ 1,168	$ 785	$ 5,959
Phase II	13,895	8,854	10,866
Phase III	32,876	26,198	8,019
Research and drug discovery	14,797	13,658	13,101
Total R&D	$62,736	$49,495	$37,945

We track development expenses that are directly attributable to our clinical development candidates by phase of clinical development. Such development expenses include third-party contract costs relating to formulation, manufacturing, preclinical studies and clinical trial activities. We allocate research and development salaries, benefits or indirect costs to our development candidates and we have included such costs in research and development expenses. All remaining research and development expenses are reflected in “Research and drug discovery” which represents early stage drug discovery programs. Such expenses include allocated employee salaries and related benefits, stock-based compensation, consulting and contracted services to supplement our in-house laboratory activities, laboratory consumables and allocated facility costs associated with these earlier stage programs.

At any given time, we typically have several active early stage research and drug discovery projects. Our internal resources, employees and infrastructure are not directly tied to any individual research or drug discovery project and are typically deployed across multiple projects. As such, we do not maintain information regarding

these costs incurred for our early stage research and drug discovery programs on a project specific basis. We expect our research and development expenses to increase as we advance our development programs further and increase the number and size of our clinical trials. The process of conducting preclinical studies and clinical trials necessary to obtain regulatory approval is costly and time consuming. We or our partners may never succeed in achieving marketing approval for any of our drug candidates. The probability of success for each drug candidate may be affected by numerous factors, including preclinical data, clinical data, competition, manufacturing capability and commercial viability. Our strategy includes entering into additional partnerships with third parties for the development and commercialization of some of our independent drug candidates.

The successful development of our drug candidates is highly uncertain and may not result in approved products. Completion dates and completion costs can vary significantly for each drug candidate and are difficult to predict for each product. Given the uncertainty associated with clinical trial enrollments and the risks inherent in the development process, we are unable to determine the duration and completion costs of the current or future clinical trials of our drug candidates or if, or to what extent, we will generate revenues from the commercialization and sale of any of our drug candidates. We anticipate we will make determinations as to which programs to pursue and how much funding to direct to each program on an ongoing basis in response to the scientific and clinical success of each drug candidate, as well as ongoing assessment as to each drug candidate's commercial potential. We will need to raise additional capital or may seek additional strategic alliances in the future in order to complete the development and commercialization of our drug candidates, including avacopan, CCX140 and CCX872.

General and administrative expenses

Total general and administrative expenses were as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
General and administrative expenses	$20,409	$16,509	$14,710
Dollar increase	$ 3,900	$ 1,799	
Percentage increase	24%	12%	

General and administrative expenses consist primarily of salaries and related benefits, including stock-based compensation and travel expenses, in executive, finance, business and corporate development and other administrative functions. Other general and administrative expenses include allocated facility-related costs not otherwise included in research and development expenses, legal costs of pursuing patent protection of our intellectual property, and professional fees for auditing, tax, and legal services.

The increase from 2017 to 2018 was primarily due to higher employee-related expenses, including those associated with our commercialization planning efforts, and higher professional fees. The increase from 2016 to 2017 was primarily due to higher intellectual property related expenses and accounting related fees associated with preparing to meet the requirements pursuant to the Sarbanes-Oxley Act of 2002 partially offset by lower travel expenses.

We anticipate that our general and administrative expenses will increase substantially in the future primarily because of increased pre-commercial activities and personnel costs to support the potential launch of avacopan for the treatment of AAV in the United States.

Other income, net

Other income, net primarily consists of interest income earned on our marketable securities. Total other income, net as compared to prior years was as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Interest income	$ 3,528	$1,370	$757
Interest expense	(1,224)	(4)	—
Total other income, net	$ 2,304	$1,366	$757
Dollar increase	$ 938	$ 609	
Percentage increase	69%	80%	

The increase in total other income, net from 2017 to 2018 was primarily due to increased interest income resulting from higher cash and investment balances and a higher return on the investment portfolio in 2018, partially offset by interest expense related to the loan and security agreement, or Credit Facility, with Hercules Capital, Inc., or Hercules.

The increase in total other income, net from 2016 to 2017 was primarily due to higher cash and investment balances in 2017 due to the upfront payments totaling $145.0 million from Vifor in connection with the Avacopan Agreement, Avacopan Amendment and CCX140 Agreement.

We expect interest expense to increase in the future as a result of higher debt balance in 2019, additional amounts borrowed under the Credit Facility, if any, or if interest rates continue to rise.

Liquidity and Capital Resources

As of December 31, 2018, we had approximately $177.0 million in cash, cash equivalents and investments. The following table shows a summary of our cash flows for each of the three years ended December 31, 2018, 2017 and 2016 (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Cash provided by (used in)			
Operating activities	$ 16,436	$ 4,878	$ 39,145
Investing activities	$(53,068)	$15,602	$(48,764)
Financing activities	$ 24,700	$ 7,516	$ 8,820

Operating activities. Net cash provided by operating activities was $16.4 million for the year ended December 31, 2018, compared to $4.9 million provided by operating activities for the same period in 2017. This change was primarily due to changes in working capital items, which was partially offset by a higher net loss. For the year ended December 31, 2018, net cash provided by operating activities included the receipt of a $50.0 million milestone payment in connection with the Avacopan Agreement, a $10.0 million upfront commitment under the Avacopan Amendment, a $10.0 million of aggregate payments under the June 2018 Avacopan Letter Agreement and the CCX140 Letter Agreement and an $11.5 million payment for CCX140 development funding by Vifor. For the year ended December 31, 2017, net cash provided by operating activities included the receipt of $60.0 million from collaboration agreements, of which $50.0 million was the upfront payment under the CCX140 Agreement and $10.0 million for the first installment of the upfront commitment under the Avacopan Amendment. In 2016, net cash provided by operating activities included $78.0 million in connection with the Avacopan Agreement.

Investing activities. Net cash provided by or used in investing activities for periods presented primarily relate to the purchase, sale and maturity of investments used to fund the day-to-day needs of our business. The

use of cash in investing activities in all periods presented represents the investment of funds received under the Avacopan Agreement and CCX140 Agreement, in each case, as amended, and the related letter agreements.

Financing activities. Net cash provided by financing activities was $24.7 million for the year ended December 31, 2018, compared to $7.5 million for the same period in 2017. For the years ended December 31, 2018 and 2017, net cash provided by financing activities included $15.0 million and $5.0 million received under the Credit Facility, respectively. Net cash provided by financing activities was $8.8 million for the year ended December 31, 2016, which was primarily due to $7.0 million in net proceeds from the issuance of 3,333,333 shares of our common stock in connection with the Avacopan Agreement. Net cash provided by financing activities for the years presented included proceeds from the exercise of stock options and stock purchases from contributions to our 2012 Employee Stock Purchase Plan and cash used for tendered ChemoCentryx, Inc. common stock to satisfy employee tax withholding requirements upon vesting of restricted stock units.

In December 2017, we entered into a Credit Facility with Hercules, under which we may borrow up to $50.0 million in three tranches, subject to certain terms and conditions. As of December 31, 2018, we had borrowed $20.0 million under the Credit Facility, with $25.0 million subject to Hercules' approval through June 15, 2019. We intend to use the net proceeds from the Credit Facility for general corporate purposes, which may include the repayment of debt and working capital. We were in compliance with all loan covenants as of December 31, 2018. See "Note 7. Long-term Debt" in the Notes to Consolidated Financial Statements included in Item 8 of this Annual Report on Form 10-K for additional information regarding our borrowings.

As of December 31, 2018, we had approximately $177.0 million in cash, cash equivalents and investments. Additionally, in the first quarter of 2019, we sold 1,666,367 shares of our common stock pursuant to our "at-the market" equity offering program for net proceeds of $19.4 million. We believe that our available cash, cash equivalents and investments will be sufficient to fund our anticipated level of operations for at least 12 months following our financial statement issuance date, March 11, 2019. However, our forecast of the period of time through which our financial resources will be adequate to support our operations is a forward-looking statement that involves risks and uncertainties, and actual results could vary materially.

Our future capital requirements are difficult to forecast and will depend on many factors, including:

- the terms and timing of any other collaborative, licensing and other arrangements that we may establish;
- the initiation, progress, timing and completion of preclinical studies and clinical trials for our drug candidates and potential drug candidates;
- the number and characteristics of drug candidates that we pursue;
- the progress, costs and results of our clinical trials;
- the outcome, timing and cost of regulatory approvals;
- delays that may be caused by changing regulatory approvals;
- the cost and timing of hiring new employees to support continued growth;
- the costs involved in filing and prosecuting patent applications and enforcing and defending patent claims;
- the cost and timing of procuring clinical and commercial supplies of our drug candidates;
- the cost and timing of establishing sales, marketing and distribution capabilities; and
- the extent to which we acquire or invest in businesses, products or technologies.

Contractual Obligations and Commitments

The following is a summary of our long-term contractual cash obligations as of December 31, 2018 (in thousands):

	Payments Due by Period				
	Total	Less than One Year	1-3 Years	3-5 Years	More than 5 Years
Long-term debt (1)	$20,000	$ —	$ 6,363	$13,637	$ —
Aggregate interest obligation (2)	7,140	1,766	3,456	1,918	—
Operating lease (3)	1,816	1,316	500	—	—
Total contractual obligations	$28,956	$ 3,082	$10,319	$15,555	$ —

(1) These amounts represent the future principal payments, excluding the end of the term charge, of the Credit Facility we entered into with Hercules. See "Note 7. Long-term Debt" in the Notes to Consolidated Financial Statements included in Item 8 of this Annual Report on Form 10-K for additional information.

(2) These amounts represent the estimated interest for our outstanding debt obligations that are payable in cash and the end of term charge, excluding non-cash amortization of debt discount. See "Note 7. Long-term Debt" in the Notes to Consolidated Financial Statements included in Item 8 of this Annual Report on Form 10-K for additional information.

(3) We lease our facility in Mountain View, California. The lease expires in 2020.

We enter into contracts in the normal course of business with CROs for clinical trials and clinical supply manufacturing and with vendors for preclinical research studies, research supplies and other services and products for operating purposes. These contracts generally provide for termination on notice, and therefore are cancelable contracts and not included in the table of contractual obligations and commitments.

Off-Balance Sheet Arrangements

We do not have any off-balance sheet arrangements (as defined by applicable SEC regulations) that are reasonably likely to have a current or future material effect on our financial condition, results of operations, liquidity, capital expenditures or capital resources, except warrants and stock options.

Recent Accounting Pronouncements

See "Note 2. Summary of Significant Accounting Policies" in the Notes to Consolidated Financial Statements of this Annual Report on Form 10-K for a full description of recently issued accounting pronouncements, including the respective expected dates of adoption and effects on our consolidated financial position and results of operations.

Item 7A. Quantitative and Qualitative Disclosures About Market Risk.

The primary objectives of our investment activities are to ensure liquidity and to preserve principal while at the same time maximizing the income we receive from our marketable securities without significantly increasing risk. Some of the securities that we invest in may have market risk. This means that a change in prevailing interest rates may cause the principal amount of the marketable securities to fluctuate. To minimize the risk in the future, we intend to maintain our portfolio of cash equivalents and short-term investments in a variety of securities, including commercial paper, money market funds, government and non-government debt securities and corporate obligations. Because of the short-term maturities of our cash equivalents and marketable securities, we do not believe that an increase or decrease in interest rates would have any significant impact on the realized value of our marketable securities.

Advances under the Credit Facility will bear an interest rate equal to the greater of (i) 8.05% plus the prime rate as reported from time to time in The Wall Street Journal minus 4.75%, and (ii) 8.05%. We are affected by

market risk exposure primarily through the effect of changes in interest rates on amounts payable under the Credit Facility. At December 31, 2018, borrowings under the Credit Facility totaled $20.0 million with an interest rate of 8.80%. We are obligated to make interest-only payments through July 1, 2021, at which point we will then be obligated to repay the principal balance and interest on the advances in equal monthly installments after the interest-only period and continuing through December 1, 2022. If the amount outstanding under the Credit Facility remained at this level for an entire year and the interest rate increased by 1%, our annual interest expense would increase by an additional $200,000. See "Note 7. Long-term Debt" in the Notes to Consolidated Financial Statements included in Item 8 of this Annual Report on Form 10-K for additional information regarding our borrowings.

Item 8. Financial Statements and Supplementary Data.

Our consolidated financial statements and the reports of our independent registered public accounting firm are included in this Annual Report on Form 10-K on pages F-1 through F-33 and are incorporated herein by reference.

Item 9. Changes in and Disagreements with Accountants on Accounting and Financial Disclosure.

None.

Item 9A. Controls and Procedures.

Conclusions Regarding the Effectiveness of Disclosure Controls and Procedures

As of December 31, 2018, management, with the participation of our Disclosure Committee, performed an evaluation of the effectiveness of the design and operation of our disclosure controls and procedures as defined in Rules 13a-15(e) and 15d-15(e) of the Exchange Act. Our disclosure controls and procedures are designed to ensure that information required to be disclosed in the reports we file or submit under the Exchange Act is recorded, processed, summarized and reported within the time periods specified in the Securities and Exchange Commission's rules and forms, and that such information is accumulated and communicated to our management, including the Chief Executive Officer and the Chief Financial and Administrative Officer, to allow timely decisions regarding required disclosures.

Any controls and procedures, no matter how well designed and operated, can provide only reasonable assurance of achieving the desired control objective. Based on this evaluation, our Chief Executive Officer and Chief Financial and Administrative Officer concluded that, as of December 31, 2018, the design and operation of our disclosure controls and procedures were effective.

Management's Annual Report on Internal Control Over Financial Reporting

Our management is responsible for establishing and maintaining adequate internal control over financial reporting as such term is defined in Rules 13a-15(f) and 15d-15(f) of the Exchange Act. Our internal control over financial reporting is a process designed to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with U.S. generally accepted accounting principles, or GAAP. Our internal control over financial reporting includes those policies and procedures that: (i) pertain to the maintenance of records that in reasonable detail accurately and fairly reflect the transactions and dispositions of our assets, (ii) provide reasonable assurance that transactions are recorded as necessary to permit preparation of financial statements in accordance with GAAP, and that our receipts and expenditures are being made only in accordance with authorizations of our management and directors, and (iii) provide reasonable assurance regarding prevention or timely detection of unauthorized acquisition, use or disposition of our assets that could have a material effect on our financial statements.

Because of its inherent limitations, internal control over financial reporting may not prevent or detect misstatements. Also, projections of any evaluation of effectiveness to future periods are subject to the risk that

controls may become inadequate because of changes in conditions, or that the degree of compliance with the policies or procedures may deteriorate.

Under the supervision and with the participation of our management, including our Chief Executive Officer and Chief Financial and Administrative Officer, we conducted an evaluation of the effectiveness of our internal control over financial reporting as of December 31, 2018 based on the criteria established in Internal Control - Integrated Framework issued by the Committee of Sponsoring Organizations of the Treadway Commission, or COSO (the 2013 Framework). Based on our evaluation under the criteria set forth in Internal Control - Integrated Framework issued by the COSO, our management concluded our internal control over financial reporting was effective as of December 31, 2018.

Our independent registered public accounting firm, Ernst & Young LLP, has audited our Consolidated Financial Statements included in Item 8 of this Annual Report on Form 10-K and has issued a report on our internal control over financial reporting as of December 31, 2018, which appears below.

Changes in Internal Control Over Financial Reporting

There has been no change in our internal control over financial reporting during the quarter ended December 31, 2018, that has materially affected, or is reasonably likely to materially affect, our internal control over financial reporting.

Item 9B. Other Information.

None.

PART III

Item 10. Directors, Executive Officers and Corporate Governance.

Information required by this item will be contained in our Definitive Proxy Statement to be filed with the Securities and Exchange Commission in connection with our 2019 Annual Meeting of Stockholders, or the Definitive Proxy Statement, which is expected to be filed not later than 120 days after the end of our fiscal year ended December 31, 2018, under the headings "Election of Directors," "Corporate Governance," "Our Executive Officers," and "Section 16(a) Beneficial Ownership Reporting Compliance," and is incorporated herein by reference.

We have adopted a Code of Business Conduct and Ethics that applies to our officers, directors and employees which is available on our website at www.chemocentryx.com. The Code of Business Conduct and Ethics contains general guidelines for conducting the business of our company consistent with the highest standards of business ethics, and is intended to qualify as a "code of ethics" within the meaning of Section 406 of the Sarbanes-Oxley Act of 2002 and Item 406 of Regulation S-K. In addition, we intend to promptly disclose (1) the nature of any amendment to our Code of Business Conduct and Ethics that applies to our principal executive officer, principal financial officer, principal accounting officer or controller or persons performing similar functions and (2) the nature of any waiver, including an implicit waiver, from a provision of our code of ethics that is granted to one of these specified officers, the name of such person who is granted the waiver and the date of the waiver on our website in the future.

Item 11. Executive Compensation.

Information required by this item will be contained in our Definitive Proxy Statement under the heading "Executive Compensation and Other Information," and is incorporated herein by reference.

Item 12. Security Ownership of Certain Beneficial Owners and Management and Related Stockholder Matters.

The information under the heading "Equity Compensation Plan Information" in Part II, Item 5, "Market for Registrant's Common Equity, Related Stockholder Matters and Issuer Purchases of Equity Securities" is incorporated herein by reference. Additional information required by this item will be contained in our Definitive Proxy Statement under the heading "Security Ownership of Certain Beneficial Owners and Management" and is incorporated herein by reference.

Item 13. Certain Relationships and Related Transactions, and Director Independence.

Information required by this item will be contained in our Definitive Proxy Statement under the headings "Certain Relationships and Related Party Transactions," "Board Independence" and "Committees of the Board of Directors" and is incorporated herein by reference.

Item 14. Principal Accounting Fees and Services.

Information required by this item will be contained in our Definitive Proxy Statement under the heading "Independent Registered Public Accountants' Fees," and is incorporated herein by reference.

PART IV

Item 15. Exhibits, Financial Statement Schedules

(a) Documents filed as part of this Annual Report on Form 10-K:

1. Financial Statements.

The following consolidated financial statements of ChemoCentryx, Inc., together with the reports thereon of Ernst & Young LLP, an independent registered public accounting firm, are included in this Annual Report on Form 10-K:

2. Financial Statement Schedules.

All schedules are omitted because they are not applicable or the required information is shown in the financial statements or notes thereto.

3. Exhibits.

A list of exhibits is set forth on the Exhibit Index immediately preceding the signature page of this Annual Report on Form 10-K, and is incorporated herein by reference.

Item 16. Form 10-K Summary

None.

ChemoCentryx, Inc.

Consolidated Financial Statements
As of December 31, 2018 and 2017
and for each of the three years in the period ended December 31, 2018

Contents

Report of Independent Registered Public Accounting Firm

The Stockholders and Board of Directors of ChemoCentryx, Inc.

Opinion on the Financial Statements

We have audited the accompanying consolidated balance sheets of ChemoCentryx, Inc. (the Company) as of December 31, 2018 and 2017, the related consolidated statements of operations, comprehensive income (loss), stockholders' equity and cash flows for each of the three years in the period ended December 31, 2018, and the related notes (collectively referred to as the "consolidated financial statements"). In our opinion, the consolidated financial statements present fairly, in all material respects, the financial position of the Company at December 31, 2018 and 2017, and the results of its operations and its cash flows for each of the three years in the period ended December 31, 2018, in conformity with U.S. generally accepted accounting principles.

We also have audited, in accordance with the standards of the Public Company Accounting Oversight Board (United States) (PCAOB), the Company's internal control over financial reporting as of December 31, 2018, based on criteria established in Internal Control - Integrated Framework issued by the Committee of Sponsoring Organizations of the Treadway Commission (2013 Framework) and our report dated March 11, 2019 expressed an unqualified opinion thereon.

Adoption of New Accounting Standard

As discussed in Note 2 to the consolidated financial statements, the Company changed its method of accounting for the recognition of revenue in 2018.

Basis for Opinion

These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on the Company's financial statements based on our audits. We are a public accounting firm registered with the PCAOB and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audits in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement, whether due to error or fraud. Our audits included performing procedures to assess the risks of material misstatement of the financial statements, whether due to error or fraud, and performing procedures that respond to those risks. Such procedures included examining, on a test basis, evidence regarding the amounts and disclosures in the financial statements. Our audits also included evaluating the accounting principles used and significant estimates made by management, as well as evaluating the overall presentation of the financial statements. We believe that our audits provide a reasonable basis for our opinion.

/s/ Ernst & Young LLP

We have served as the Company's auditor since 2000.

Redwood City, California
March 11, 2019

Report of Independent Registered Public Accounting Firm

The Stockholders and Board of Directors of ChemoCentryx, Inc.

Opinion on Internal Control over Financial Reporting

We have audited ChemoCentryx, Inc.'s internal control over financial reporting as of December 31, 2018, based on criteria established in Internal Control - Integrated Framework issued by the Committee of Sponsoring Organizations of the Treadway Commission (2013 framework) (the COSO criteria). In our opinion, ChemoCentryx, Inc. (the Company) maintained, in all material respects, effective internal control over financial reporting as of December 31, 2018, based on the COSO criteria.

We also have audited, in accordance with the standards of the Public Company Accounting Oversight Board (United States) (PCAOB), the consolidated balance sheets of the Company as of December 31, 2018 and 2017, and the related consolidated statements of operations, comprehensive income (loss), stockholders' equity and cash flows for each of the three years in the period ended December 31, 2018, and the related notes and our report dated March 11, 2019 expressed an unqualified opinion thereon.

Basis for Opinion

The Company's management is responsible for maintaining effective internal control over financial reporting and for its assessment of the effectiveness of internal control over financial reporting included in the accompanying Management's Annual Report on Internal Control over Financial Reporting. Our responsibility is to express an opinion on the Company's internal control over financial reporting based on our audit. We are a public accounting firm registered with the PCAOB and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audit in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether effective internal control over financial reporting was maintained in all material respects. Our audit included obtaining an understanding of internal control over financial reporting, assessing the risk that a material weakness exists, testing and evaluating the design and operating effectiveness of internal control based on the assessed risk, and performing such other procedures as we considered necessary in the circumstances. We believe that our audit provides a reasonable basis for our opinion.

Definition and Limitations of Internal Control Over Financial Reporting

A company's internal control over financial reporting is a process designed to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles. A company's internal control over financial reporting includes those policies and procedures that (1) pertain to the maintenance of records that, in reasonable detail, accurately and fairly reflect the transactions and dispositions of the assets of the company; (2) provide reasonable assurance that transactions are recorded as necessary to permit preparation of financial statements in accordance with generally accepted accounting principles, and that receipts and expenditures of the company are being made only in accordance with authorizations of management and directors of the company; and (3) provide reasonable assurance regarding prevention or timely detection of unauthorized acquisition, use, or disposition of the company's assets that could have a material effect on the financial statements.

Because of its inherent limitations, internal control over financial reporting may not prevent or detect misstatements. Also, projections of any evaluation of effectiveness to future periods are subject to the risk that controls may become inadequate because of changes in conditions, or that the degree of compliance with the policies or procedures may deteriorate.

/s/ Ernst & Young LLP

Redwood City, California
March 11, 2019

CHEMOCENTRYX, INC.

Consolidated Balance Sheets
(In thousands, except share and par value data)

	December 31,	
	2018	2017
Assets		
Current assets:		
Cash and cash equivalents	$ 28,088	$ 40,020
Short-term investments	148,896	87,271
Accounts receivable from related party	2,058	51,090
Prepaid expenses and other current assets	2,342	1,449
Total current assets	181,384	179,830
Property and equipment, net	1,536	1,210
Long-term investments	—	7,929
Other assets	390	359
Total assets	$ 183,310	$ 189,328
Liabilities and Stockholders' Equity		
Current liabilities:		
Accounts payable	$ 966	$ 1,400
Accrued liabilities	12,969	8,575
Deferred revenue from related party	50,461	22,962
Total current liabilities	64,396	32,937
Long-term debt, net	19,689	4,676
Noncurrent deferred revenue from related party	84,100	72,197
Other non-current liabilities	387	251
Total liabilities	168,572	110,061
Commitments (Note 8)		
Stockholders' equity:		
Preferred stock, $0.001 par value, 10,000,000 shares authorized; no shares issued and outstanding	—	—
Common stock, $0.001 par value, 200,000,000 shares authorized; 50,652,238 and 48,837,060 shares issued and outstanding at December 31, 2018 and 2017, respectively	51	49
Additional paid-in capital	389,398	368,553
Note receivable	(16)	(16)
Accumulated other comprehensive loss	(198)	(119)
Accumulated deficit	(374,497)	(289,200)
Total stockholders' equity	14,738	79,267
Total liabilities and stockholders' equity	$ 183,310	$ 189,328

See accompanying notes.

CHEMOCENTRYX, INC.

Consolidated Statements of Operations
(In thousands, except per share data)

	Year Ended December 31,		
	2018	2017	2016
Revenue:			
Collaboration and license revenue from related party	$ 42,875	$82,497	$ 11,435
Grant revenue	—	—	500
Total revenue	42,875	82,497	11,935
Operating expenses:			
Research and development	62,736	49,495	37,945
General and administrative	20,409	16,509	14,710
Total operating expenses	83,145	66,004	52,655
Income (loss) from operations	(40,270)	16,493	(40,720)
Other income (expense):			
Interest income	3,528	1,370	757
Interest expense	(1,224)	(4)	—
Total other income, net	2,304	1,366	757
Net income (loss)	$(37,966)	$17,859	$(39,963)
Net income (loss) per common share			
Basic	$ (0.76)	$ 0.37	$ (0.86)
Diluted	$ (0.76)	$ 0.36	$ (0.86)
Shares used to compute net income (loss) per common share			
Basic	49,814	48,413	46,432
Diluted	49,814	49,615	46,432

See accompanying notes.

CHEMOCENTRYX, INC.

Consolidated Statements of Comprehensive Income (Loss)
(In thousands)

	Year Ended December 31,		
	2018	2017	2016
Net income (loss)	$(37,966)	$17,859	$(39,963)
Unrealized loss on available-for-sale securities	(79)	(69)	(10)
Comprehensive income (loss)	$(38,045)	$17,790	$(39,973)

See accompanying notes.

CHEMOCENTRYX, INC.

Consolidated Statements of Stockholders' Equity
(In thousands, except share data)

	Common Stock		Additional Paid-In Capital	Note Receivable	Accumulated Other Comprehensive Loss	Accumulated Deficit	Total Stockholders' Equity
	Shares	Amount					
Balance as of December 31, 2015	44,185,506	$ 44	$ 339,615	$ (16)	$ (40)	$ (267,096)	$ 72,507
Net loss	—	—	—	—	—	(39,963)	(39,963)
Unrealized gain / (loss) on investments	—	—	—	—	(10)	—	(10)
Issuance of common stock pursuant to collaboration and licensing agreement	3,333,333	3	6,997	—	—	—	7,000
Issuance of common stock under equity incentive and employee stock purchase plans	539,081	1	1,819	—	—	—	1,820
Employee stock-based compensation	—	—	8,222	—	—	—	8,222
Compensation expense related to options granted to consultants	—	—	313	—	—	—	313
Balance as of December 31, 2016	48,057,920	48	356,966	(16)	(50)	(307,059)	49,889
Net income	—	—	—	—	—	17,859	17,859
Unrealized gain / (loss) on investments	—	—	—	—	(69)	—	(69)
Issuance of common stock under equity incentive and employee stock purchase plans	838,107	1	3,268	—	—	—	3,269
Repurchased shares upon vesting of restricted stock units for tax withholdings	(58,967)	—	(429)	—	—	—	(429)
Employee stock-based compensation	—	—	8,119	—	—	—	8,119
Compensation expense related to options granted to consultants	—	—	629	—	—	—	629
Balance as of December 31, 2017	48,837,060	49	368,553	(16)	(119)	(289,200)	79,267
Net loss	—	—	—	—	—	(37,966)	(37,966)
Adoption of accounting standards (Note 2)	—	—	—	—	—	(47,331)	(47,331)
Unrealized gain / (loss) on investments	—	—	—	—	(79)	—	(79)
Issuance of common stock under equity incentive and employee stock purchase plans	1,912,703	2	10,690	—	—	—	10,692
Repurchased shares upon vesting of restricted stock units for tax withholdings	(97,525)	—	(678)	—	—	—	(678)
Employee stock-based compensation	—	—	9,971	—	—	—	9,971
Compensation expense related to options granted to consultants	—	—	862	—	—	—	862
Balance as of December 31, 2018	50,652,238	$ 51	$ 389,398	$ (16)	$ (198)	$ (374,497)	$ 14,738

See accompanying notes.

CHEMOCENTRYX, INC.

Consolidated Statements of Cash Flows
(In thousands)

	Year Ended December 31,		
	2018	2017	2016
Operating activities			
Net income (loss)	$ (37,966)	$ 17,859	$ (39,963)
Adjustments to reconcile net income (loss) to net cash provided by operating activities:			
Depreciation of property and equipment	512	418	348
Stock-based compensation	10,833	8,748	8,535
Noncash interest (income) expense, net	(1,071)	143	179
Changes in assets and liabilities:			
Accounts receivable due from related party	49,032	(20,885)	(30,205)
Prepaids and other current assets	(668)	(727)	35
Other assets	(31)	(80)	(119)
Accounts payable	(434)	729	(4)
Other liabilities	4,158	80	3,773
Deferred revenue from related party	(7,929)	(1,407)	96,566
Net cash provided by operating activities	16,436	4,878	39,145
Investing activities			
Purchases of property and equipment, net	(838)	(723)	(304)
Purchases of investments	(192,480)	(133,845)	(136,234)
Maturities of investments	140,250	150,170	87,774
Net cash provided by (used in) investing activities	(53,068)	15,602	(48,764)
Financing activities			
Proceeds from issuance of common stock	—	—	7,000
Proceeds from exercise of stock options and employee stock purchase plan	10,467	3,269	1,820
Employees' tax withheld and paid for restricted stock units	(678)	(429)	—
Borrowings under credit facility agreement, net of issuance costs	14,911	4,676	—
Net cash provided by financing activities	24,700	7,516	8,820
Net increase (decrease) in cash and cash equivalents	(11,932)	27,996	(799)
Cash and cash equivalents at beginning of period	40,020	12,024	12,823
Cash and cash equivalents at end of period	$ 28,088	$ 40,020	$ 12,024
Supplemental disclosures of cash flow information			
Cash paid for interest	$ 748	$ —	$ —

See accompanying notes.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements

December 31, 2018

1. Description of Business

ChemoCentryx, Inc. (the Company) commenced operations in 1997. The Company is a clinical-stage biopharmaceutical company focused on developing new medications targeted at inflammatory disorders, autoimmune diseases and cancer. The Company's principal operations are in the United States and it operates in one segment.

2. Summary of Significant Accounting Policies

Consolidation

The consolidated financial statements include the Company's accounts and those of its wholly owned subsidiaries, ChemoCentryx Ireland Limited and ChemoCentryx Limited. The operations of ChemoCentryx Ireland Limited and ChemoCentryx Limited have been immaterial to date. All intercompany amounts have been eliminated in consolidation.

Use of Estimates

The preparation of financial statements in conformity with accounting principles generally accepted in the United States (GAAP) requires management to make estimates and assumptions that affect the amounts reported in the consolidated financial statements and accompanying notes. Actual results could differ from these estimates.

Basis of Presentation

The financial statements are prepared in conformity with GAAP. The Company has made estimates and assumptions that affect the amounts reported in the financial statements and accompanying notes. Actual results could differ from those estimates.

Cash Equivalents and Investments

The Company considers all highly liquid investments with an original maturity at the date of purchase of three months or less to be cash equivalents. The Company limits its concentration of risk by diversifying its investments among a variety of issuers. All investments are classified as available for sale and are recorded at fair value based on quoted prices in active markets or based upon other observable inputs, with unrealized gains and losses excluded from earnings and reported in other comprehensive income (loss). Purchase premiums and discounts are recognized in interest income using the interest method over the terms of the securities. Realized gains and losses and unrealized declines in fair value that are deemed to be other than temporary are reflected in the statement of operations. The cost of securities sold is based on the specific-identification method.

Fair Value of Financial Instruments

The carrying amounts of certain of the Company's financial instruments, including cash and cash equivalents, short-term investments, accounts receivable and accounts payable, approximate their fair value due to their short maturities.

Fair value is considered to be the price at which an asset could be exchanged or a liability transferred (an exit price) in an orderly transaction between knowledgeable, willing parties in the principal or most advantageous

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

market for the asset or liability. Where available, fair value is based on or derived from observable market prices or other observable inputs. Where observable prices or inputs are not available, valuation models are applied. The valuation techniques involve management estimation and judgment, the degree of which is dependent on the price transparency for the instruments or market and the instruments' complexity.

Concentration of Credit Risk

The Company invests in a variety of financial instruments and, by its policy, limits the amount of credit exposure with any one issuer, industry or geographic area.

For the years ended December 31, 2018, 2017 and 2016, 100%, 100% and 96%, respectively, of the Company's total revenue was derived from the Company's collaboration with Vifor (International) Ltd., and/or its affiliates, or collectively, Vifor. Accounts receivable are typically unsecured and are concentrated in the pharmaceutical industry and government sector. Accordingly, the Company may be exposed to credit risk generally associated with pharmaceutical companies and government funded entities. The Company has not historically experienced any significant losses due to concentration of credit risk.

Accounts receivable consists of the following (in thousands):

	December 31,	
	2018	2017
Vifor (1)	$2,058	$51,090

(1) As of December 31, 2017, accounts receivable excluded the $10.0 million cash commitment received from Vifor in February 2018 in connection with the agreement that harmonized the geographic commercialization rights underlying the agreements for both avacopan and CCX140 drug candidates (the Avacopan Amendment). See "Note 9. Related-Party Transactions" for a detailed discussion.

Property and Equipment

Property and equipment are stated at cost less accumulated depreciation. Depreciation is calculated using the straight-line method over the estimated useful lives of the assets, which range from five to seven years. Tenant improvements are depreciated over the lesser of the estimated useful life or the remaining life of the lease at the time the asset is placed into service.

Impairment of Long-Lived Assets

The Company reviews long-lived assets, including property and equipment, for impairment whenever events or changes in business circumstances indicate that the carrying amount of the assets may not be fully recoverable. An impairment loss would be recognized when estimated undiscounted future cash flows expected to result from the use of an asset are less than its carrying amount. The impairment loss would be based on the excess of the carrying value of the impaired asset over its respective fair value. To date, the Company has not recorded any impairment losses.

Revenue Recognition

Effective January 1, 2018, the Company adopted Accounting Standards Codification (ASC) Topic 606, *Revenue from Contracts with Customers* (ASC 606) using the modified retrospective transition method. This standard applies to all contracts with customers, except for contracts that are within the scope of other

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

standards, such as leases, insurance, collaboration arrangements and financial instruments. Under ASC 606, the Company recognizes revenue when its customer obtains control of promised goods or services, in an amount that reflects the consideration which the Company expects to receive in exchange for those goods or services. To determine revenue recognition for arrangements that the Company determines are within the scope of ASC 606, the Company performs the following five steps: (i) identify the contract(s) with a customer; (ii) identify the performance obligations in the contract; (iii) determine the transaction price; (iv) allocate the transaction price to the performance obligations in the contract; and (v) recognize revenue when (or as) the Company satisfies a performance obligation. The Company only applies the five-step model to contracts when it is probable that Company will collect the consideration it is entitled to in exchange for the goods or services it transfers to the customer. At contract inception, once the contract is determined to be within the scope of ASC 606, the Company assesses the goods or services promised within each contract and determines those that are performance obligations and assesses whether each promised good or service is distinct. The Company then recognizes as revenue the amount of the transaction price that is allocated to the respective performance obligation when (or as) the performance obligation is satisfied.

The Company enters into corporate collaborations under which it may obtain upfront license fees, research and development funding and development and regulatory and commercial milestone payments and royalty payments. The Company's performance obligations under these arrangements may include licenses of intellectual property, distribution rights, research and development services, delivery of manufactured product, and/or participation on joint steering committees.

Licenses of intellectual property: If the license to the Company's intellectual property is determined to be distinct from the other performance obligations identified in the arrangement, the Company recognizes revenue from upfront license fees allocated to the license when the license is transferred to the customer and the customer is able to use and benefit from the license. For licenses that are bundled with other promises, the Company utilizes judgement to assess the nature of the combined performance obligation to determine whether the combined performance obligation is satisfied over time or at a point in time and, if over time, the appropriate method of measuring proportional performance for purposes of recognizing revenue from non-refundable, up-front fees. The Company evaluates the measure of proportional performance each reporting period and, if necessary, adjusts the measure of performance and related revenue recognition.

Milestone payments: At the inception of each arrangement that includes development, regulatory or commercial milestone payments, the Company evaluates whether the milestones are considered probable of being reached and estimates the amount to be included in the transaction price. There are two alternatives to use when estimating the amount of variable consideration: the expected value method and the most likely amount method. Under the expected value method, an entity considers the sum of probability-weighted amounts in a range of possible consideration amounts. Under the most likely amount method, an entity considers the single most likely amount in a range of possible consideration amounts. Whichever method is used, it should be consistently applied throughout the life of the contract; however, it is not necessary for the Company to use the same approach for all contracts. The Company expects to use the most likely amount method for development and regulatory milestone payments. If it is probable that a significant revenue reversal would not occur, the associated milestone value is included in the transaction price. Milestone payments that are not within the Company's or the licensee's control, such as regulatory approvals, are not considered probable of being achieved until those approvals are received. The transaction price is then allocated to each performance obligation on a relative stand-alone selling price basis. The Company recognizes revenue as or when the performance obligations under the contract are satisfied. At the end of each subsequent reporting period, the Company re-evaluates the

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

probability of achievement of each such milestone and any related constraint, and if necessary, adjusts its estimates of the overall transaction price. Any such adjustments are recorded on a cumulative catch-up basis, which would affect revenues and earnings in the period of adjustment.

Commercial milestones and royalties: For arrangements that include sales-based royalties, including milestone payments based on the level of sales, and in which the license is deemed to be the predominant item to which the royalties relate, the Company recognizes revenue when the related sales occur. To date, the Company has not recognized any royalty revenue resulting from its collaboration arrangements.

Up-front payments and fees are recorded as deferred revenue upon receipt or when due, and may require deferral of revenue recognition to a future period until the Company performs its obligations under these arrangements. Amounts payable to the Company are recorded as accounts receivable when the Company's right to consideration is unconditional.

Upon adoption of ASC 606 under the modified retrospective transition method, the Company recognized the cumulative effect of initially applying the new revenue standard of $47.3 million as an adjustment to the opening balance of accumulated deficit and an increase in deferred revenue. The comparative information has not been restated and continues to be reported under the accounting standards in effect for those periods. Before the adoption of ASC 606, the Company recognized upfront fees straight-line under ASC 605 over the estimated performance period and recognized milestones when earned under the milestone method of accounting. See "Note 2. Summary of Significant Accounting Policies – Revenue Recognition" in the Company's Annual Report on Form 10-K for the fiscal year ended December 31, 2017, filed with the SEC on March 12, 2018 for a detailed discussion.

The impact of adoption on the Company's consolidated statement of operations and balance sheet was as follows (in thousands):

	For the Year Ended December 31, 2018		
	As Reported	Balances Without Adoption of ASC606	Effect of Change
Statement of Operations			
Collaboration and license revenue from related party	$ 42,875	$ 28,083	$14,792
Loss from operations	(40,270)	(55,062)	14,792
Net loss	(37,966)	(52,758)	14,792

	December 31, 2018		
	As Reported	Balances Without Adoption of ASC606	Effect of Change
Balance Sheet			
Liabilities:			
Deferred revenue from related party	$ 50,461	$ 19,939	$ 30,522
Noncurrent deferred revenuefrom related party	84,100	82,083	2,017
Stockholders' equity:			
Accumulated deficit	(374,497)	(341,958)	(32,539)

Revenue from government and private agency grants are recognized as the related research and development expenses are incurred and to the extent that funding is approved.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

Research and Development Expenses

All research and development expenses are recognized as incurred. Research and development expenses include, but are not limited to, salaries and related benefits, including stock-based compensation, third-party contract costs relating to research, formulation, manufacturing, preclinical study and clinical trial activities, laboratory consumables and allocated facility costs.

Clinical Trial Accruals

Clinical trial costs are a component of research and development expenses. The Company accrues and expenses clinical trial activities performed by third parties based upon estimates of the percentage of work completed over the life of the individual study in accordance with agreements established with clinical research organizations and clinical trial sites. The Company determines the estimates through discussions with internal clinical personnel and external service providers as to the progress or stage of completion of trials or services and the agreed-upon fee to be paid for such services.

Nonrefundable advance payments for goods and services that will be used or rendered in future research and development activities, are deferred and recognized as expense in the period that the related goods are delivered or services are performed.

Income Taxes

The Company uses the liability method for income taxes, whereby deferred tax assets and liabilities are determined based on the differences between the financial reporting and tax reporting bases of assets and liabilities and are measured using enacted tax rates and laws that are expected to be in effect when the differences are expected to reverse. Valuation allowances are provided when the expected realization for the deferred tax assets does not meet the more-likely-than-not criteria.

The Company accounts for uncertain tax positions in the financial statements when it is not more likely than not that the position will be sustained upon examination by the tax authorities. Such tax positions must initially and subsequently be measured at the largest amount of tax benefit that has a greater than 50% likelihood of being realized upon ultimate settlement with the tax authority assuming full knowledge of the position and relevant facts. The Company's policy is to recognize any interest and penalties related to unrecognized tax benefits in income tax expense.

Comprehensive Income (loss)

Comprehensive income (loss) comprises net income (loss) and other comprehensive income (loss). For the periods presented, other comprehensive loss consists of unrealized losses on the Company's available-for-sale securities. For the years ended December 31, 2018, 2017 and 2016, there were no sales of investments, and therefore there were no reclassifications.

Stock-Based Compensation

The Company accounts for employee stock-based compensation using a fair-value-based method, which measures stock-based compensation cost at the grant date based on the fair value of the award, and recognizes as an expense over the award's vesting periods on a straight-line basis. Because stock compensation expense is

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

based on awards ultimately expected to vest, it has been reduced by an estimate for future forfeitures. Forfeitures are estimated at the time of grant and revised, if necessary, in subsequent periods if actual forfeitures differ from those estimates.

The Company accounts for stock-based compensation arrangements with nonemployees using a fair-value approach. For stock options granted to nonemployees, the fair value of the stock options is estimated using the Black-Scholes valuation model. This model utilizes the estimated fair value of common stock and requires that, at the date of grant, assumptions are made with respect to the remaining contractual term of the option, the volatility of the fair value of its common stock, the risk-free interest rates and the expected dividend yields of its common stock. The measurement of nonemployee stock-based compensation is subject to periodic adjustment as the underlying equity instruments vest.

The Company accounts for restricted stock compensation arrangements with nonemployee directors using a fair-value approach. For restricted stock units (RSUs) and restricted stock awards (RSAs) granted to nonemployee directors, the fair value of a RSU or RSA is valued at the closing price of the Company's common stock on the date of the grant. The Company recognizes stock-based compensation expense associated with these RSUs and RSAs over the requisite service period, with no adjustment in future periods based on the Company's actual stock price over the vesting period.

Net Income (Loss) Per Share

Basic net income (loss) per common share is computed by dividing net income (loss) attributable to common stockholders by the weighted-average number of common shares outstanding during the period, without consideration for common stock equivalents.

Diluted net income (loss) per share is computed by dividing net income (loss) attributable to common stockholders by the sum of the weighted-average number of common shares outstanding and dilutive common stock equivalent shares outstanding for the period. The Company's potentially dilutive common stock equivalent shares, which include incremental common shares issuable upon (i) the exercise of outstanding stock options and warrants, (ii) vesting of RSUs and RSAs, and (iii) the purchase from contributions to the 2012 Employee Stock Purchase Plan (the ESPP) (calculated based on the treasury stock method), are only included in the calculation of diluted net income (loss) per share when their effect is dilutive.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

The following table is a reconciliation of the numerator and denominator used in the calculation of basic and diluted net income (loss) per share:

	Year Ended December 31,		
	2018	2017	2016
	(in thousands, except per share data)		
Numerator:			
Net income (loss)	$(37,966)	$17,859	$(39,963)
Denominator:			
Weighted average shares outstanding - basic	49,814	48,413	46,432
Dilutive stock options, RSUs and RSAs	—	1,202	—
Weighted average shares outstanding - diluted	49,814	49,615	46,432
Net income (loss) per common share			
Basic	$ (0.76)	$ 0.37	$ (0.86)
Diluted	$ (0.76)	$ 0.36	$ (0.86)

The following potentially dilutive securities were excluded from the calculation of diluted net income (loss) per share due to their anti-dilutive effect:

	Year Ended December 31,		
	2018	2017	2016
Options to purchase common stock, including purchases from contributions to ESPP	10,731,164	6,320,038	9,358,389
Restricted stock units	440,354	—	440,344
Restricted stock awards	27,278	—	31,306
Warrants to purchase common stock	150,000	150,000	150,000
	11,348,796	6,470,038	9,980,039

Recent Accounting Pronouncements

In February 2016, the FASB issued ASU No. 2016-02, Leases (Topic 842). The new standard requires all lessees recognize the assets and liabilities that arise from leases on the balance sheet and disclose qualitative and quantitative information about its leasing arrangements. On January 1, 2019, the Company adopted this new standard using the modified retrospective approach. The Company anticipates the impact of adoption will be an increase in the right of use asset of approximately $1.3 million, an increase in lease liability of approximately $1.7 million and a decrease of deferred rent of approximately $0.4 million on its balance sheets. The actual, final quantitative effect of the adoption of Topic 842 will be determined in the first quarter of 2019.

On December 22, 2017, the Tax Cuts and Jobs Act (the Act) was enacted into law. ASC 740, Income Taxes, requires companies to recognize the effect of the tax law changes in the period of enactment. Shortly after the enactment of the Act, the SEC staff issued Staff Accounting Bulletin No. 118 (SAB 118) to address the application of U.S. GAAP in situations when a registrant does not have the necessary information available, prepared, or analyzed (including computations) in reasonable detail to complete the accounting for certain income tax effects of the Act. The Company has adjusted its deferred tax assets and liabilities based on the reduction of the U.S. federal corporate tax rate from 34% to 21% and assessed the realizability of its deferred tax

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

assets based on its current understanding of the provisions of the new law. During 2018, the Company finalized its computation of the impact of the Act. See "Note 13. Income Taxes" for a detailed discussion.

In June 2018, the Financial Accounting Standard Board issued ASU No. 2018-07, Compensation – Stock Compensation (Topic 718). The new standard simplifies the accounting for share-based payments to nonemployees by aligning it with the accounting for share-based payments to employees, with certain exceptions. The new standard was effective for the Company on January 1, 2019. The Company does not expect the adoption of this accounting guidance to have a material impact on the consolidated financial statements.

The Company has reviewed other recent accounting pronouncements and concluded they are either not applicable to the business or that no material effect is expected on the consolidated financial statements as a result of future adoption.

In August 2018, the SEC adopted amendments to certain disclosure requirements in Securities Act Release No. 33-10532, Disclosure Update and Simplification. These amendments eliminate, modify, or integrate into other SEC requirements certain disclosure rules. Among the amendments is the requirement to present an analysis of changes in stockholders' equity in the interim financial statements included in quarterly reports on Form 10-Q. The analysis, which can be presented as a footnote or separate statement, is required for the current and comparative quarter and year-to-date interim periods. The amendments are effective for the Company in its interim financial statements for the quarter ended March 31, 2019. The Company does not anticipate that the adoption of these SEC amendments will have a material effect on the Company's financial position, results of operations, cash flows or stockholders' equity.

3. Cash Equivalents and Investments

The amortized cost and fair value of cash equivalents and investments at December 31, 2018 and 2017 were as follows (in thousands):

	December 31, 2018			
	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
Money market fund	$ 22,073	$ —	$ —	$ 22,073
U.S. treasury securities	23,013	—	(13)	23,000
Commercial paper	45,683	—	—	45,683
Asset-backed securities	29,127	—	(34)	29,093
Corporate debt securities	55,228	—	(151)	55,077
Total available-for-sale securities	$175,124	$ —	$ (198)	$174,926
Classified as:				
Cash equivalents				$ 26,030
Short-term investments				148,896
Long-term investments				—
Total available-for-sale securities				$174,926

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

3. Cash Equivalents and Investments (continued)

	December 31, 2017			
	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Fair Value
Money market fund	$ 29,848	$ —	$ —	$ 29,848
U.S. treasury securities	29,005	—	(52)	28,953
Commercial paper	46,184	—	—	46,184
Corporate debt securities	27,095	—	(67)	27,028
Total available-for-sale securities	$132,132	$ 0	$ (119)	$132,013
Classified as:				
Cash equivalents				$ 36,813
Short-term investments				87,271
Long-term investments				7,929
Total available-for-sale securities				$132,013

Cash equivalents in the tables above exclude cash of $2.1 million and $3.2 million as of December 31, 2018 and 2017, respectively. All available-for-sale securities held as of December 31, 2018 had contractual maturities of less than two years. There have been no significant realized gains or losses on available-for-sale securities for the periods presented. No significant available-for-sale securities held as of December 31, 2018 have been in a continuous unrealized loss position for more than 12 months. As of December 31, 2018, unrealized losses on available-for-sale investments are not attributed to credit risk and are considered to be temporary. The Company believes that it is more-likely-than-not that investments in an unrealized loss position will be held until maturity or the recovery of the cost basis of the investment. The Company believes it has no other-than-temporary impairments on its securities because it does not intend to sell these securities and it believes it is not more likely than not that it will be required to sell these securities before the recovery of their amortized cost basis. To date, the Company has not recorded any impairment charges on marketable securities related to other-than-temporary declines in market value.

4. Fair Value Measurements

The Company determines the fair value of financial assets and liabilities using three levels of inputs as follows:

Level 1—Inputs which include quoted prices in active markets for identical assets and liabilities.

Level 2—Inputs other than Level 1 that are observable, either directly or indirectly, such as quoted prices for similar assets or liabilities; quoted prices in markets that are not active; or other inputs that are observable or can be corroborated by observable market data for substantially the full term of the assets or liabilities.

Level 3—Unobservable inputs that are supported by little or no market activity and that are significant to the fair value of the assets or liabilities.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

4. Fair Value Measurements (continued)

The Company's financial assets subject to fair value measurements on a recurring basis and the level of inputs used in such measurements were as follows as of December 31, 2018 and 2017 (in thousands):

	December 31, 2018			
	Level 1	Level 2	Level 3	Total
Description				
Money market fund	$ 22,073	$ —	$ —	$ 22,073
U.S. treasury securities	—	23,000	—	23,000
Commercial paper	—	45,683	—	45,683
Asset-backed securities	—	29,093	—	29,093
Corporate debt securities	—	55,077	—	55,077
Total assets	$ 22,073	$ 152,853	$ —	$ 174,926

	December 31, 2017			
	Level 1	Level 2	Level 3	Total
Description				
Money market fund	$ 29,848	$ —	$ —	$ 29,848
U.S. treasury securities	—	28,953	—	28,953
Commercial paper	—	46,184	—	46,184
Corporate debt securities	—	27,028	—	27,028
Total assets	$ 29,848	$ 102,165	$ —	$ 132,013

During the year ended December 31, 2018 there were no transfers between Level 1 and Level 2 financial assets. When the Company uses observable market prices for identical securities that are traded in less active markets, the Company classifies its marketable debt instruments as Level 2. When observable market prices for identical securities are not available, the Company prices its marketable debt instruments using non-binding market consensus prices that are corroborated with observable market data; quoted market prices for similar instruments; or pricing models, such as a discounted cash flow model, with all significant inputs derived from or corroborated with observable market data. Non-binding market consensus prices are based on the proprietary valuation models of pricing providers or brokers. These valuation models incorporate a number of inputs, including non-binding and binding broker quotes; observable market prices for identical or similar securities; and the internal assumptions of pricing providers or brokers that use observable market inputs and, to a lesser degree, unobservable market inputs. The Company corroborates non-binding market consensus prices with observable market data using statistical models when observable market data exists. The discounted cash flow model uses observable market inputs, such as LIBOR-based yield curves, currency spot and forward rates, and credit ratings.

Other Fair Value Measurements

The carrying amount and estimated fair value of financial instruments not recorded at fair value at December 31, 2018 and 2017 were as follows (in thousands):

	December 31, 2018		December 31, 2017	
	Carrying Amount	Estimated Fair Value	Carrying Amount	Estimated Fair Value
Long-term debt, net (1)	$ 19,689	$ 19,847	$ 4,676	$ 4,812

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

4. Fair Value Measurements (continued)

(1) Carrying amounts of long-term debt were net of unamortized debt discounts of $311,000 and $324,000 as of December 31, 2018 and 2017, respectively.

The fair value of the Company's long-term debt is estimated using the net present value of future debt payments, discounted at an interest rate that is consistent with market interest rates, which is a Level 2 input.

5. Property and Equipment

Property and equipment consist of the following (in thousands):

	December 31,	
	2018	2017
Lab equipment	$ 6,263	$ 5,897
Computer equipment and software	1,809	1,688
Furniture and fixtures	554	551
Tenant improvements	1,030	893
	9,656	9,029
Less: accumulated depreciation	(8,120)	(7,819)
	$ 1,536	$ 1,210

6. Accrued Liabilities

Accrued liabilities consist of the following (in thousands):

	December 31,	
	2018	2017
Research and development related	$ 8,466	$4,962
Compensation related	2,767	2,345
Consulting and professional services	811	1,012
Other	925	256
	$12,969	$8,575

7. Long-term Debt

On December 28, 2017 (the Closing Date), the Company entered into a Loan and Security Agreement with Hercules Capital Inc. (Hercules) which the Company amended in December 2018, pursuant to which term loans in an aggregate principal amount of up to $50.0 million (the Credit Facility) are available to the Company in three tranches, subject to certain terms and conditions. As of December 31, 2018, the Company had borrowed $20.0 million under the Credit Facility with up to $25.0 million available for future borrowing, subject to Hercules' approval through June 15, 2019.

Advances under the Credit Facility will bear an interest rate equal to the greater of either (i) 8.05% plus the prime rate as reported from time to time in The Wall Street Journal minus 4.75%, and (ii) 8.05%. At December 31, 2018, interest rate on the outstanding borrowings under the Credit Facility was 8.80%. For advances under the first and second tranches, the Company will make interest-only payments through July 1, 2021, and will then

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

7. Long-term Debt (continued)

repay the principal balance and interest on the advances in equal monthly installments after the interest-only period and continuing through December 1, 2022. For advances made under the third tranche, the Company will make interest-only payments for the first 30 months, and will then repay the principal balance and interest on the advances in equal monthly installments after the interest-only period with each advance repaid 48 months after it is drawn.

The Company may prepay advances under the Credit Facility, in whole or in part, at any time, subject to a prepayment charge equal to: (a) 1.5% of the amount so prepaid, if such prepayment occurs during the second year following the Closing Date; and (b) 1.0% of the amount so prepaid, if such prepayment occurs after the second year following the Closing Date. The Credit Facility is secured by substantially all of the Company's assets, excluding intellectual property.

In addition, Hercules has the right to participate, in an amount up to $2.0 million in any subsequent equity financing broadly marketed to multiple investors in an amount greater than $20.0 million. The Credit Facility also includes customary affirmative restrictions on the payment of dividends and negative covenants, and events of default, the occurrence and continuance of which provide Hercules with the right to demand immediate repayment of all principal and unpaid interest under the Credit Facility, and to exercise remedies against the Company and the collateral securing the Credit Facility. The Company was in compliance with all loan covenants for all periods presented.

The Company will pay an end-of-term charge for each tranche which will occur on the earliest of (i) the applicable tranche maturity date; (ii) the date that the Company prepays all of the outstanding principal under each tranche in full, or (iii) the date the loan payments are accelerated due to an event of default. For the first tranche, the end of term charge is $0.9 million. In the case of the second and third tranches, the charge is 6.25% of the aggregate amount of the advances applicable to such tranche.

In addition, the Company pays a commitment charge of 1% of the advances made under the Credit Facility, with a minimum charge of $162,500 paid on the Closing Date. Also, the Company reimbursed Hercules for costs incurred related to the Credit Facility. These charges were recorded as discounts to the carrying value of the loan and are amortized over the term of the loan using the effective interest method.

As of December 31, 2018, the Company had outstanding borrowings under the Credit Facility of $19.7 million, net of discounts of $0.3 million. Future minimum principal payments, which exclude the end of term charge, related to the Credit Facility as of December 31, 2018 are as follows (in thousands):

	Amounts
Year ending December 31:	
2019	$ —
2020	—
2021	6,363
2022	13,637
Total minimum payments	20,000
Less: amount representing debt discount	(311)
Present value of remaining debt payments	19,689
Less: current portion	—
Noncurrent portion	$19,689

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

8. Commitments

Operating Leases

In May 2004, the Company entered into a noncancelable operating lease for its current office and primary research facility located in Mountain View, California. The Company received a discounted lease rate during the first year of the agreement. In August 2012, the Company entered into an amendment to the lease agreement for the same facility to extend the term through April 2019. In April 2017, the Company entered into a second amendment to the lease agreement for the same facility to extend the term of the lease through April 2020. The total rent obligation is being expensed ratably over the term of the agreement, as amended. Rental expenses for the years ended December 31, 2018, 2017 and 2016 were $1.4 million, $1.3 million and $1.2 million, respectively.

Future minimum lease payments under all noncancelable operating leases as of December 31, 2018, are as follows (in thousands):

	Amounts
Year ending December 31:	
2019	$1,316
2020	500
Total minimum lease payments	$1,816

9. Related-Party Transactions

Bio-Techne

Bio-Techne Corporation, formerly Techne Corporation, one of the Company's principal stockholders, held 6,385,056 shares of the Company's common stock as of December 31, 2018. In connection with the Company's initial public offering (IPO) in February 2012, Bio-Techne received a warrant with a ten-year term to purchase 150,000 shares of the Company's common stock at an exercise price per share equal to $20.00 per share, or 200% of the IPO price of its common stock, which was outstanding as of December 31, 2018 and 2017.

For the years ended December 31, 2018, 2017 and 2016, the Company paid Bio-Techne $70,000, $96,000 and $114,000, respectively, for research materials. As of December 31, 2018, there was no accounts payable balance due to Bio-Techne. As of December 31, 2017, the Company had an accounts payable balance due to Bio-Techne for the purchase of research materials of $6,000.

Vifor

In October 2018, Vifor acquired 7,343,492 shares of the Company's common stock from Glaxo Group Limited, bringing their aggregate holdings of the Company's common stock to 10,676,825 as of December 31, 2018.

The Company has collaboration agreements with Vifor: the Avacopan Agreements and CCX140 Agreement. See "Note 2. Summary of Significant Accounting Policies – Concentration of Credit Risk" for additional information on accounts receivable balance due from Vifor.

Avacopan Agreements

In May 2016, the Company entered into an exclusive collaboration and license agreement with Vifor pursuant to which the Company granted Vifor exclusive rights to commercialize avacopan in Europe and certain

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Related-Party Transactions (continued)

other markets (the Avacopan Agreement). Avacopan is the Company's lead drug candidate for the treatment of patients with anti-neutrophil cytoplasmic auto-antibody associated vasculitis (AAV) and other rare diseases. The Avacopan Agreement also provided Vifor with an exclusive option to negotiate during 2016 a worldwide license agreement for one of the Company's other drug candidates, CCX140, an orally-administered inhibitor of the chemokine receptor known as CCR2. In connection with the Avacopan Agreement, the Company received a non-refundable upfront payment of $85.0 million, comprising $60.0 million in cash and $25.0 million in the form of an equity investment to purchase 3,333,333 shares of the Company's common stock at a price of $7.50 per share.

In February 2017, Vifor and the Company expanded the Vifor territories under the Avacopan Agreement to include all markets outside the United States and China (the Avacopan Amendment). In connection with this February 2017 arrangement, the Company received a $20.0 million upfront payment for the expanded rights. In June 2018, Vifor and the Company further expanded the Vifor territories under the Avacopan Agreement to provide Vifor with exclusive commercialization rights in China (the Avacopan Letter Agreement). The Company retains control of ongoing and future development of avacopan (other than country-specific development in the licensed territories) and all commercialization rights to avacopan in the United States. In consideration for the Avacopan Letter Agreement, the Company received a $5.0 million payment for the expanded rights.

Upon achievement of certain regulatory and commercial milestones with avacopan, the Company will receive additional payments of up to $460.0 million under the Avacopan Agreement. In addition, the Company will receive royalties, with rates ranging from the low teens to the mid-twenties, on future potential net sales of avacopan by Vifor in the licensed territories. In December 2017, the Company achieved a $50.0 million regulatory milestone when the European Medicines Agency (EMA) validated the Company's Conditional marketing authorisation (CMA) application for avacopan for the treatment of AAV.

The Company identified the following material promises under the Avacopan Agreement, the Avacopan Amendment, and the Avacopan Letter Amendment: (1) the license related to avacopan; (2) the development and regulatory services for the submission of the marketing authorisation application (MAA); and (3) an exclusive option to negotiate a worldwide license agreement for CCX140, which expired in 2016. The Company considered that the license has standalone functionality and is capable of being distinct. However, the Company determined that the license is not distinct from the development and regulatory services within the context of the agreement because Vifor is dependent on the Company to execute the development and regulatory activities in order for Vifor to benefit from the license. As such, the license is combined with the development and regulatory services into a single performance obligation. The exclusive option related to CCX140 is a separate performance obligation and the Company determined that its transaction price is not material. As such, the transaction price under this arrangement is allocated to the license and the development and regulatory services.

As of December 31, 2018, the transaction price of $153.0 million consists of the following:

- $78.0 million upfront payment under the May 2016 Avacopan Agreement. Of the total $85.0 million upfront payment received under the May 2016 Avacopan Agreement, $7.0 million was allocated to the issuance of 3,333,333 shares of the Company's common stock valued at $2.10 per share, the closing stock price on the effective date of the agreement, May 9, 2016. The remaining $78.0 million was allocated to the transaction price under this arrangement;
- $20.0 million upfront payment under the February 2017 Avacopan Amendment;

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Related-Party Transactions (continued)

- $50.0 million regulatory milestone payment achieved upon the validation of the Company's CMA application by the EMA, for avacopan for the treatment of AAV in December 2017; and
- $5.0 million payment under the Avacopan Letter Agreement.

The Company will re-evaluate the transaction price in each reporting period and as uncertain events are resolved or other changes in circumstances occur.

The Company determined that the combined performance obligation will be performed over the duration of the contract, which began on the effective date of May 9, 2016 and ends upon completion of development and regulatory services. The Company uses a cost-based input method to measure proportional performance and to calculate the corresponding amount of revenue to recognize. The Company believes this is the best measure of progress because other measures do not reflect how the Company transfers its performance obligation to Vifor. In applying the cost-based input method of revenue recognition, the Company measures actual costs incurred relative to budgeted costs to fulfill the combined performance obligation. These costs consist primarily of third-party contract costs. Revenue is recognized based on actual costs incurred as a percentage of total budgeted costs as the Company completes its performance obligations.

For the year ended December 31, 2018, the Company recognized $37.1 million of collaboration and license revenue under the Avacopan Agreement, the Avacopan Amendment and the Avacopan Letter Agreement. Prior to the adoption of ASC 606 on January 1, 2018, the Company accounted for its performance obligations under the Avacopan Agreement and Avacopan Amendment as one combined unit of accounting with the upfront fees being recognized over the estimated period of performance. See "Note 10. Collaboration and License Agreements – Avacopan Agreements" in the Company's Annual Report on Form 10-K for the fiscal year ended December 31, 2017, filed with the SEC on March 12, 2018, for further discussion. For the years ended December 31, 2017 and 2016, the Company recognized $72.5 million and $11.4 million of collaboration and license revenue, respectively, under the Avacopan Agreement and Avacopan Amendment under ASC 605.

CCX140 Agreement

In December 2016, the Company entered into a second collaboration and license agreement with Vifor pursuant to which the Company granted Vifor exclusive rights to commercialize CCX140 (the CCX140 Agreement) in markets outside the United States and China. CCX140 is an orally-administered inhibitor of the chemokine receptor known as CCR2. The Company retains marketing rights in the United States and China, while Vifor has commercialization rights in the rest of the world. Pursuant to the CCX140 Agreement, the Company is responsible for the clinical development of CCX140 in rare renal diseases and is reimbursed for Vifor's equal share of such development cost. Vifor retains an option to solely develop and commercialize CCX140 in more prevalent forms of chronic kidney disease (CKD). Should Vifor later exercise the CKD option, the Company would receive co-promotion rights for CKD in the United States. Under the terms of the CCX140 Agreement, the Company received a non-refundable upfront payment of $50.0 million in 2017.

In June 2018, the Company and Vifor entered into a letter agreement to expand Vifor's rights to include the right to exclusively commercialize CCX140 in China (the CCX140 Letter Agreement). In connection with the CCX140 Letter Agreement, the Company received a non-refundable payment of $5.0 million. The Company and Vifor also entered into an amendment to the CCX140 Agreement (the CCX140 Amendment) to clarify the timing of certain payments with respect to development funding of the CCX140 program by Vifor, and the Company received a non-refundable payment of $11.5 million. The Company retains control of ongoing and future development of CCX140 (other than country-specific development in the licensed territories), and all commercialization rights to CCX140 in the United States.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Related-Party Transactions (continued)

Upon achievement of certain regulatory and commercial milestones with CCX140, the Company will receive additional payments of up to $625.0 million under the CCX140 Agreement. In addition, the Company will receive tiered royalties, with rates ranging from ten to the mid-twenties, on net sales of CCX140 in the licensed territories.

The Company identified the following material promises under the CCX140 Agreement, the CCX140 Amendment, and CCX140 Letter Agreement: (1) the license related to CCX140; and (2) the development and regulatory services for the submission of the MAA. The Company considered that the license has standalone functionality and is capable of being distinct. However, the Company determined that the license is not distinct from the development and regulatory services within the context of the agreement because Vifor is dependent on the Company to execute the development and regulatory activities in order for Vifor to benefit from the license. As such, the license is combined with the development and regulatory services into a single performance obligation.

As of December 31, 2018, the transaction price of $113.5 million consists of the following:

- $50.0 million upfront payment under the CCX140 Agreement;
- $58.5 million of CCX140 development funding by Vifor; and
- $5.0 million upfront payment under the CCX140 Letter Agreement.

The Company will re-evaluate the transaction price in each reporting period and as uncertain events are resolved or other changes in circumstances occur.

The Company determined that the combined performance obligation will be performed over the duration of the contract, which began on the effective date of December 22, 2016 and ends upon completion of development and regulatory services. The Company uses a cost-based input method to measure proportional performance and to calculate the corresponding amount of revenue to recognize. The Company believes this is the best measure of progress because other measures do not reflect how the Company transfers its performance obligation to Vifor. In applying the cost-based input method of revenue recognition, the Company measures actual costs incurred relative to budgeted costs to fulfill the combined performance obligation. These costs consist primarily of third-party contract costs. Revenue is recognized based on actual costs incurred as a percentage of total budgeted costs as the Company completes its performance obligations. For the year ended December 31, 2018, the Company recognized $5.8 million of collaboration and license revenue under the CCX140 Agreement, the CCX140 Amendment and the CCX140 Letter Agreement.

Prior to the adoption of ASC 606 on January 1, 2018, the Company accounted for its performance obligations under the CCX140 Agreement as one combined unit of accounting with the upfront fee of $50.0 million being recognized over the estimated period of performance. See “Note 10. Collaboration and License Agreements – CCX140 Agreement” in the Company’s Annual Report on Form 10-K for the fiscal year ended December 31, 2017, filed with the SEC on March 12, 2018, for further discussion. For the year ended December 31, 2017, the Company recognized $10.0 million of collaboration and license revenue under the CCX140 Agreement under ASC 605.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Related-Party Transactions (continued)

The following table presents the contract assets and liabilities for all of the Company's revenue contracts as of the following dates (in thousands):

	December 31, 2018	December 31, 2017
Contract asset:		
Accounts receivable	$ 2,058	$ 51,090
Contract liability:		
Deferred revenue (1)	(134,561)	(95,159)

(1) Upon adoption of ASC 606 under the modified retrospective transition method, the Company recognized the cumulative effect of initially applying the new revenue standard of $47.3 million as an adjustment to the opening balance of accumulated deficit and an increase in deferred revenue. See "Note 2. Summary of Significant Accounting Policies – Revenue Recognition" for a detailed discussion.

During the year ended December 31, 2018, the Company recognized the following revenue as a result of changes in the contract asset and the contract liability balances (in thousands):

	Year Ended December 31, 2018
Revenue recognized in the period from:	
Amount included in contract liability at the beginning of the period	$ 39,815
Performance obligations satisfied (or partially satisfied) in previous periods	$ (3,357)

10. Government Grant

In April 2016, the Company was awarded an Orphan Products Development grant by the U.S. Food and Drug Administration to support the clinical development of avacopan in the amount of $500,000, which was fully recognized during the year ended December 31, 2016. The term of the grant expired in May 2017.

11. Stockholders' Equity

Equity Incentive Plans

In May 2002, the stockholders approved the Amended and Restated 1997 Stock Option/Stock Issuance Plan (the 1997 Plan) and in September 2002, the stockholders approved the 2002 Equity Incentive Plan (the 2002 Plan). In February 2012, the stockholders approved the 2012 Equity Incentive Award Plan (the 2012 Plan). As of December 31, 2018, a total of 13,440,000 shares of the Company's common stock were reserved for issuance under the 2012 Plan. In addition, the number of shares available for issuance under the 2012 Plan will be annually increased by an amount equal to the lesser of: 2,000,000 shares; 4% of the outstanding shares of the Company's common stock as of the last day of the Company's immediately preceding fiscal year; or an amount determined by the Company's Board of Directors. In November 2018, the Board of Directors approved an increase to the number of shares reserved forissuance under the 2012 Plan by 2,000,000 shares effective January 1, 2019. Collectively, the 1997 Plan, the 2002 Plan and the 2012 Plan are known as the Stock Plans.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Stockholders' Equity (continued)

Restricted Stock

Restricted Stock Awards (RSAs) and Restricted Stock Units (RSUs) are independent of stock option grants and are not transferrable, and are subject to forfeiture if recipients terminate their service to the Company prior to the release of the vesting restrictions. RSUs granted to employees generally vest over a period of three years. RSUs and RSAs granted to its nonemployee directors vest over a one-year period, or over a three-year period in the case of an initial grant pursuant to the Company's Non-Employee Director Compensation Policy (Directors Plan). In the case of a change in control, RSUs and RSAs granted to nonemployee directors will vest in full. RSUs and RSAs are valued at the closing price of the Company's common stock on the date of grant. During the years ended December 31, 2017 and 2016, the weighted-average grant date fair value for restricted stock granted was $6.72 and $4.48, respectively. The total fair value of restricted stock vested during the years ended December 31, 2018, 2017 and 2016 was $2.4 million, $1.7 million and $0.2 million, respectively.

The activity for restricted stock is summarized as follows:

	Shares	Weighted-Average Grant-Date Fair Value
Balance at December 31, 2017	508,444	$ 5.79
Granted	228,860	11.32
Vested	(269,672)	5.83
Canceled	—	—
Unvested at December 31, 2018	467,632	$ 8.47

As of December 31, 2018, there was $2.0 million of unrecognized compensation expense associated with unvested restricted stock, which is expected to be recognized over a weighted-average period of 1.3 years.

Stock Options

Under the Stock Plans, incentive stock options may be granted by the Board of Directors to employees at exercise prices of not less than 100% of the fair value at the date of grant. Nonstatutory options may be granted by the Board of Directors to employees, officers, and directors of the Company or consultants at exercise prices of not less than 85% of the fair value of the common stock on the date of grant. The fair value at the date of grant is determined by the Board of Directors. Under the Stock Plans, options may be granted with different vesting terms from time to time, but not to exceed 10 years from the date of grant. Outstanding options generally vest over four years, with 25% of the total grant vesting on the first anniversary of the option grant date and 1/36th of the remaining grant vesting each month thereafter.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Stockholders' Equity (continued)

The following table summarizes stock option activity and related information under the Company's Stock Plans:

	Available for Grant	Shares	Weighted Average Exercise Price	Weighted Average Remaining Contractual Term (in years)	Aggregate Intrinsic Value
Balance at December 31, 2017	2,028,880	10,203,571	$ 7.68		
Shares authorized	1,940,000	—			
Granted (1)	(2,565,372)	2,336,512	9.97		
Exercised (2)	97,525	(1,612,400)	6.26		
Forfeited and expired	207,483	(207,483)	7.92		
Outstanding at December 31, 2018	1,708,516	10,720,200	$ 8.39	6.34	$33,230,822
Vested and expected to vest, net of estimated forfeiture at December 31, 2018		10,462,992	$ 8.38	6.27	$32,631,253
Exercisable at December 31, 2018		6,977,786	$ 8.40	5.05	$23,063,000

(1) The difference between shares granted in the number of shares available for grant and outstanding options represents the RSUs and RSAs granted for the period.
(2) Shares presented as available for grant represents shares repurchased for tax withholding upon vesting of RSUs.

The aggregate intrinsic value represents the value of the Company's closing stock price on the last trading day of the period in excess of the weighted-average exercise price multiplied by the number of options outstanding or exercisable. Total intrinsic value of options exercised was $9.8 million, $1.3 million and $0.7 million during 2018, 2017 and 2016, respectively. As of December 31, 2018, there was $15.0 million of unrecognized compensation expense, net of estimated forfeitures, associated with outstanding stock options, which is expected to be recognized over an estimated weighted-average period of 2.5 years.

As of December 31, 2018, stock options outstanding were as follows:

	Options Outstanding	
Exercise Price Range	Shares	Weighted-Average Contractual Life
$2.10 - $3.57	1,258,034	7.18
$4.50 - $6.08	1,073,633	4.71
$6.19 - $6.30	1,092,556	5.78
$6.50 - $6.60	77,225	8.31
$6.62	1,099,056	8.11
$6.90 - $7.28	1,083,118	5.17
$7.42 - $8.19	1,272,523	6.41
$8.22 - $10.82	536,534	6.09
$10.86	1,119,200	9.18
$10.91 - $15.90	2,108,321	5.05
	10,720,200	6.34

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Stockholders' Equity (continued)

Employee Stock Purchase Plan

In February 2012, the stockholders approved the ESPP. As of December 31, 2018, a total of 1,100,000 shares of the Company's common stock were reserved for issuance under the ESPP. In addition, the number of shares available for issuance under the ESPP may be annually increased on the first day of each fiscal year during the term of the ESPP, beginning with the 2012 fiscal year, by an amount equal to the lesser of: 300,000 shares; 1% of outstanding shares of the Company's common stock; or an amount determined by the Company's Board of Directors. The ESPP provides for an aggregate limit of 3,000,000 shares of common stock that may be issued under the ESPP during the term of the ESPP. In November 2018, the Board of Directors approved an increase to the number of shares reserved for issuance under the ESPP by 300,000 shares effective January 1, 2019.

The Company issued 88,784, 93,221 and 157,893 shares under the ESPP in 2018, 2017 and 2016, respectively. As of December 31, 2018, 394,551 shares were available for issuance under the ESPP. As of December 31, 2018, there was $0.1 million of unrecognized compensation expense, net of estimated forfeitures, associated with the ESPP, which is expected to be recognized over an estimated weighted-average period of 0.4 years.

Stock Awards Granted to Employees

Employee stock-based compensation expense recognized is calculated based on awards ultimately expected to vest and reduced for estimated forfeitures. Forfeitures are estimated at the time of grant and revised, if necessary, in subsequent periods if actual forfeitures differ from those estimates.

Total employee stock-based compensation expense recognized associated with restricted stock, stock options, and the ESPP, was as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Research and development	$3,632	$3,154	$3,245
General and administrative	6,339	4,965	4,977
Total	$9,971	$8,119	$8,222

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Stockholders' Equity (continued)

Valuation Assumptions

Fair value of options granted under the Stock Plans and purchases under the Company's ESPP were estimated at grant or purchase dates using a Black-Scholes option valuation model. The Black-Scholes valuation model requires that assumptions are made with respect to various factors, including the expected volatility of the fair value of the Company's common stock. The Company has based its expected volatility on the average historical volatilities of public entities having similar characteristics including: industry, stage of life cycle, size, and financial leverage. The weighted-average expected term of options was calculated using the simplified method as prescribed by accounting guidance for stock-based compensation. This decision was based on the lack of relevant historical data due to the Company's limited historical experience. The fair values of the employee stock options granted under the Company's Stock Plans and the option component of the shares purchased under the ESPP during 2018, 2017 and 2016 were estimated at the date of grant using the Black-Scholes option-pricing model with the following assumptions:

	Employee Stock Options			Employee Stock Purchase Plan		
	2018	2017	2016	2018	2017	2016
Dividend yield	0%	0%	0%	0%	0%	0%
Volatility	67.8%	68.3%	65.6%	73.8%	52.9%	99.7%
Weighted-average expected life (in years)	6.0	6.0	6.0	0.5	0.5	0.5
Risk-free interest rate	2.66%	2.04%	1.58%	2.33%	1.22%	0.47%
Weighted-average grant date fair value	$6.22	$4.30	$2.43	$3.73	$2.16	$2.29

Stock Options Granted to Nonemployees

During 2018, 2017 and 2016, the Company granted to consultants options to purchase 28,534, 239,266 and 15,000 shares of common stock, respectively. The stock-based compensation expense related to nonemployees will fluctuate as the fair value of the Company's common stock fluctuates. In connection with grants of stock options to nonemployees, the Company recorded stock-based compensation expense as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Research and development	$862	$629	$313
General and administrative	—	—	—
Total	$862	$629	$313

Valuation Assumptions

Stock-based compensation expense associated with stock options granted to nonemployees is recognized as the stock options vest. The estimated fair values of the stock options granted are calculated at each reporting date using the Black-Scholes option-pricing model, with the following assumptions:

	Year Ended December 31,		
	2018	2017	2016
Dividend yield	0%	0%	0%
Volatility	67-68%	69-70%	65-68%
Weighted-average expected life (in years)	5.7-9.9	5.5-10.0	6.1-9.9
Risk-free interest rate	2.7-3.0%	1.9-2.5%	1.3-2.4%

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Stockholders' Equity (continued)

Equity Distribution Agreement

In December 2018, the Company entered into an Equity Distribution Agreement (EDA), pursuant to which the Company may offer and sell, from time to time, shares of our common stock, par value $0.001 per share, having an aggregate offering price of up to $75.0 million.

12. 401(k) Plan

In October 1997, the Company established the ChemoCentryx 401(k) Plan and Trust (the 401(k) Plan). Employees may contribute, up to the percentage limit imposed by the Internal Revenue Code of 1986, as amended, an amount of their salary each calendar year until termination of their employment with the Company. The Company may elect to make matching contributions, as per the Plan; however, no matching contributions were made in the years ended December 31, 2018, 2017 and 2016.

13. Income Taxes

The Company's loss before tax is only attributable to U.S. operations. The components of the income tax (benefit) expense are as follows (in thousands):

	Year Ended December 31,		
	2018	2017	2016
Current (benefit from) provision for income taxes:			
Federal	$ —	$ —	$ —
State	—	—	—
Total current (benefit from) provision for income taxes	—	—	—
Deferred (benefit from) provision for income taxes:	—	—	—
Federal	—	—	—
State	—	—	—
Total deferred tax (benefit from) provision for income taxes	—	—	—
(Benefit from) provision for income taxes	$ —	$ —	$ —

A reconciliation of the federal statutory income tax rate to the Company's effective income tax rate is as follows:

	Year Ended December 31,		
	2018	2017	2016
Federal statutory income tax rate	(21.0%)	34.0%	(34.0%)
State income taxes, net of federal benefit	—	—	(5.8)
Permanent items	1.6	5.5	2.1
Excess tax benefit for stock-based compensation	(2.8)	(2.0)	—
Research and development credits	(3.5)	(7.2)	(2.8)
Change in valuation allowance	24.5	(224.1)	38.1
Change in tax rate	—	193.8	—
Other	1.2	—	2.4
(Benefit from) provisions for income taxes	— %	— %	— %

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

13. Income Taxes (continued)

The tax effects of temporary differences and carryforwards that give rise to significant portions of the deferred tax assets consist of the following (in thousands):

	December 31,	
	2018	2017
Deferred tax assets:		
Net operating loss carryforwards	$ 62,405	$ 55,569
Research and development credit	11,794	10,470
Amortization of deferred stock compensation - non-qualified	6,569	6,385
Reserves and accruals	1,140	785
Deferred revenue	20,991	10,297
Depreciation and amortization	—	231
Gross deferred tax assets	102,899	83,737
Less: valuation allowance	(102,891)	(83,737)
Total deferred tax assets	8	—
Deferred tax liabilities:		
Depreciation and amortization	(8)	—
Total deferred tax liabilities	(8)	—
Net deferred tax assets	$ —	$ —

On December 22, 2017, the Act was enacted into law. The Act contains several key provisions including the reduction of the corporate income tax rate to 21%, effective January 1, 2018, as well as a variety of other changes including the limitation of the tax deductibility of interest expense, acceleration of expensing of certain business assets and reductions in the amount of executive pay that could qualify as a tax deduction. Additionally, the Act introduced a territorial-style system for taxing foreign source income of domestic multinational corporations. As a result of the Act and the federal rate reduction the federal deferred tax assets were reduced by $36.0 million with an offset to the Company's valuation allowance which is reflected in the financial statements for the tax year ended December 31, 2017.

ASC 740, Income Taxes, requires companies to recognize the effect of the tax law changes in the period of enactment. Shortly after the enactment of the Act, the SEC staff issued "SAB 118" which allows the Company to record provisional amounts during a measurement period not to extend beyond one year from the enactment date. The analysis and accounting for the tax effects of the Act were completed in the fourth quarter ended December 31, 2018, with no additional impact to the Company's deferred tax assets.

The Company concluded that it was more likely than not that its deferred tax assets would not be realized. Accordingly, the total deferred tax assets were fully offset by a valuation allowance. The Company's valuation allowance increased by approximately $19.2 million in 2018 and decreased by approximately $40.0 million in 2017.

At December 31, 2018, the Company had federal and state net operating loss carryforwards of approximately $225.4 million and $246.8 million, respectively. The federal net operating loss carryforwards will begin to expire in 2031. Due to tax reform, federal net operating loss carryforwards generated post-2017 no longer have an expiration date. The state net operating loss carryforwards begin to expire in 2019.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

13. Income Taxes (continued)

The Company has federal and state research and development credit carryforwards of $12.7 million and $6.9 million, respectively. The federal research and development credits will begin to expire in 2019 if not utilized. California research and development credits can be carried forward indefinitely.

Utilization of the net operating loss and credit carryforwards may be subject to annual limitation due to historical or future ownership percentage change rules provided by the Internal Revenue Code of 1986, and similar state provisions. The annual limitation may result in the expiration of certain net operating loss and credit carryforwards before their utilization.

A reconciliation of the Company's unrecognized tax benefits for the years ended December 31, 2018, 2017 and 2016, is as follows (in thousands):

	Unrecognized Income Tax Benefits
Balance as of December 31, 2016	$ 5,430
Additions for current tax positions	603
Additions for prior tax positions	2,753
Balance as of December 31, 2017	$ 8,786
Additions for current tax positions	928
Balance as of December 31, 2018	$ 9,714

As of December 31, 2018 and 2017, the Company had approximately $9.7 million and $8.8 million, respectively, of unrecognized tax benefits, none of which would currently affect the Company's effective tax rate if recognized due to the Company's deferred tax assets being fully offset by a valuation allowance. In 2018, unrecognized tax benefits increased due to uncertainty associated with the Company's research and development credits. The Company is not aware of any items that will significantly increase or decrease its unrecognized tax benefits in the next 12 months.

For U.S. federal and California income tax purposes, the statute of limitations remains open for the years beginning 2015 and 2014, respectively, except for the carryforward of net operating losses and research and development credits generated in prior years.

If applicable, the Company would classify interest and penalties related to uncertain tax positions in income tax expense. Through December 31, 2018, there has been no interest expense or penalties related to unrecognized tax benefits.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

14. Selected Quarterly Financial Data (unaudited)

Selected quarterly results from operations for the years ended December 31, 2018 and 2017 are as follows (in thousands except per share amounts):

	2018 Quarter Ended			
	March 31	June 30	September 30	December 31
Revenue	$ 9,546	$15,022	$ 8,975	$ 9,332
Net loss	$(9,417)	$ (6,874)	$ (10,890)	$ (10,785)
Basic and diluted net loss per share	$ (0.19)	$ (0.14)	$ (0.22)	$ (0.21)

	2017 Quarter Ended			
	March 31	June 30	September 30	December 31
Revenue	$ 8,230	$ 8,937	$ 9,029	$ 56,301
Net income (loss)	$(5,996)	$ (9,240)	$ (6,560)	$ 39,655
Basic net income (loss) per share	$ (0.12)	$ (0.19)	$ (0.13)	$ 0.81
Diluted net income (loss) per share	$ (0.12)	$ (0.19)	$ (0.13)	$ 0.80

The four quarters of net earnings per share may not add to the total year because of differences in the weighted-average numbers of shares outstanding during the quarters and the year.

15. Subsequent Event

In January 2019, the Company sold 1,666,367 shares of its common stock pursuant to its EDA at a price per share of $12.00, for net proceeds of $19.4 million.

EXHIBIT INDEX

Exhibit Number	Description
3.1(1)	Amended and Restated Certificate of Incorporation.
3.2(1)	Amended and Restated Bylaws.
4.1(2)	Form of Common Stock Certificate.
4.2(3)	Form of Common Stock Warrant.
4.3(3)	Form of Series B Preferred Stock Warrant.
10.1#(1)	Amended and Restated 1997 Stock Option/Stock Issuance Plan and form of agreement thereunder.
10.2#(1)	Amended and Restated 2002 Equity Incentive Plan and forms of agreements thereunder.
10.3#(1)	2012 Equity Incentive Award Plan and form of agreement thereunder.
10.4#(1)	2012 Employee Stock Purchase Plan.
10.5#(1)	2012 Cash Incentive Plan.
10.6#(1)	Form of Indemnification Agreement.
10.7#(4)	Non-Employee Director Compensation Policy.
10.8#(14)	Amended and Restated Non-Employee Director Compensation Policy.
10.9#(5)	Form of Restricted Stock Unit Award Grant Notice and Restricted Stock Unit Award Agreement under the 2012 Equity Incentive Award Plan.
10.10#(6)	Form of Restricted Stock Award Grant Notice and Restricted Stock Award Agreement under the 2012 Equity Incentive Award Plan.
10.11#(3)	Amended and Restated Employment Agreement, effective as of January 1, 2008, by and between the Registrant and Thomas J. Schall, Ph.D.
10.12#(3)	Amended and Restated Employment Agreement, effective as of January 1, 2008, by and between the Registrant and Markus J. Cappel, Ph.D.
10.13#(3)	Amended and Restated Employment Agreement, effective as of January 1, 2008, by and between the Registrant and Susan M. Kanaya.
10.14#	Employment Agreement, effective as of May 2, 2016, by and between the Registrant and Rajinder Singh.
10.15(3)	Standard Industrial/Commercial Multi-Tenant Lease, dated April 20, 2004, by and between Portola Land Company and the Registrant.
10.16(7)	First Amendment to Standard Industrial/Commercial Multi-Tenant Lease, dated August 16, 2012, by and between Portola Land Company and the Registrant.
10.17(10)	Second Amendment to Lease, dated April 13, 2017, by and between Google Inc. and the Registrant.
10.18†(8)	Product Development and Commercialization Agreement, effective as of August 22, 2006, by and between the Registrant and Glaxo Group Limited.
10.19†(3)	Amendment No. 1 to Product Development and Commercialization Agreement, effective as of September 30, 2007, by and between the Registrant and Glaxo Group Limited.
10.20†(3)	Amendment No. 2 to Product Development and Commercialization Agreement, effective as of October 6, 2008, by and between the Registrant and Glaxo Group Limited.
10.21†(3)	Amendment No. 3 to Product Development and Commercialization Agreement, effective as of August 22, 2009, by and between the Registrant and Glaxo Group Limited.

Exhibit Number	Description
10.22†(3)	Amendment No. 4 to Product Development and Commercialization Agreement, effective as of February 26, 2010, by and between the Registrant and Glaxo Group Limited.
10.23†(3)	Amendment No. 5 to Product Development and Commercialization Agreement, effective as of November 15, 2010, by and between the Registrant and Glaxo Group Limited.
10.24†(9)	Collaboration and License Agreement, dated as of May 9, 2016, by and between the Registrant and Vifor (International) Ltd.
10.25(9)	Stock Purchase Agreement, dated as of May 9, 2016, by and between the Registrant and Vifor (International) Ltd.
10.26(6)	Collaboration and License Agreement, dated as of December 22, 2016, by and between the Registrant and Vifor (International) Ltd.
10.27†(4)	Letter Agreement dated as of February 13, 2017 between the Registrant and Vifor (International) Ltd.
10.28†(10)	Amendment to Collaboration and License Agreement, effective as of May 22, 2017 between the Registrant and Vifor Fresenius Medical Care Renal Pharma Ltd.
10.29(11)	Loan and Security Agreement, dated as of December 28, 2017, by and between the Registrant and Hercules Capital, Inc.
10.30(12)	Letter Agreement dated as of June 6, 2018 between the Registrant and Vifor (International) Ltd. Regarding Grant of Rights to CCX168 in China.
10.31(12)	Letter Agreement dated as of June 6, 2018 between the Registrant and Vifor (International) Ltd. Regarding Grant of Rights to CCX140 in China.
10.32†(12)	Amendment to Collaboration and License Agreement, effective as of June 6, 2018 between the Registrant and Vifor Fresenius Medical Care Renal Pharma Ltd.
10.33(13)	Equity Distribution Agreement, dated as of December 4, 2018, by and between the Registrant and Piper Jaffray & Co.
10.34	Amendment No. 1 to Loan and Security Agreement, dated as of December 13, 2018, by and between the Registrant and Hercules Capital, Inc.
21.1	Subsidiaries of the Registrant.
23.1	Consent of independent registered public accounting firm.
31.1	Certification of Chief Executive Officer pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
31.2	Certification of Chief Financial Officer pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
32.1	Certification of Chief Executive Officer pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
32.2	Certification of Chief Financial Officer pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
101.INS	XBRL Instance Document.
101.SCH	XBRL Taxonomy Extension Schema Document.
101.CAL	XBRL Taxonomy Extension Calculation Linkbase Document.
101.DEF	XBRL Taxonomy Extension Definition Linkbase Document.
101.LAB	XBRL Taxonomy Extension Label Linkbase Document.
101.PRE	XBRL Taxonomy Extension Presentation Linkbase document.

(1) Filed with Amendment No. 3 to the Registrant's Registration Statement on Form S-1 on January 23, 2012 (Registration No. 333-177332), and incorporated herein by reference.
(2) Filed with Amendment No. 4 to the Registrant's Registration Statement on Form S-1 on February 6, 2012 (Registration No. 333-177332), and incorporated herein by reference.
(3) Filed with the Registrant's Registration Statement on Form S-1 on October 14, 2011 (Registration No. 333-177332), and incorporated herein by reference.
(4) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended March 31, 2017, filed with the SEC on May 10, 2017, and incorporated herein by reference.
(5) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended June 30, 2014, filed with the SEC on August 8, 2014, and incorporated herein by reference.
(6) Filed with the Registrant's Annual Report on Form 10-K for the year ended December 31, 2016, filed with the SEC on March 14, 2017, and incorporated herein by reference.
(7) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended September 30, 2012, filed with the SEC on November 13, 2012, and incorporated herein by reference.
(8) Filed with Amendment No. 2 to Registrant's Registration Statement on Form S-1 on January 6, 2012 (Registration No. 333-177332), and incorporated herein by reference.
(9) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended June 30, 2016, filed with the SEC on August 9, 2016, and incorporated herein by reference.
(10) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended June 30, 2017, filed with the SEC on August 8, 2017, and incorporated herein by reference.
(11) Filed with the Registrant's Current Report on Form 8-K filed on January 4, 2018, and incorporated herein by reference.
(12) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended June 30, 2018, filed with the SEC on August 9, 2018, and incorporated herein by reference.
(13) Filed with the Registrant's Current Report on Form 8-K filed on December 4, 2018, and incorporated herein by reference.
(14) Filed with the Registrant's Quarterly Report on Form 10-Q for the quarterly period ended March 31, 2018, filed with the SEC on May 9, 2018, and incorporated herein by reference.
# Indicates management contract or compensatory plan.
† Confidential treatment has been granted for portions of this exhibit. These portions have been omitted and filed separately with the SEC.

SIGNATURES

Pursuant to the requirements of Section 13 or 15(d) of the Securities Exchange Act of 1934, as amended, the Registrant has duly caused this Annual Report on Form 10-K to be signed on its behalf by the undersigned, thereunto duly authorized.

CHEMOCENTRYX, INC.

Date: March 11, 2019

By: /s/ Thomas J. Schall, Ph.D.
Thomas J. Schall, Ph.D.
President and Chief Executive Officer

Pursuant to the requirements of the Securities Exchange Act of 1934, as amended, this Annual Report on Form 10-K has been signed by the following persons on behalf of the Registrant and in the capacities and on the dates indicated.

Signature	Title	Date
/s/ Thomas J. Schall, Ph.D. Thomas J. Schall, Ph.D.	President, Chief Executive Officer and Director (Principal Executive Officer)	March 11, 2019
/s/ Susan M. Kanaya Susan M. Kanaya	Executive Vice President, Chief Financial and Administrative Officer and Secretary (Principal Financial Officer)	March 11, 2019
/s/ Pui San Kwan Pui San Kwan	Vice President, Finance (Principal Accounting Officer)	March 11, 2019
/s/ Thomas A. Edwards Thomas A. Edwards	Director	March 11, 2019
/s/ Joseph M. Feczko, M.D. Joseph M. Feczko, M.D.	Director	March 11, 2019
/s/ Roger C. Lucas, Ph.D. Roger C. Lucas, Ph.D.	Director	March 11, 2019
/s/ Henry A. McKinnell, Jr., Ph.D. Henry A. McKinnell, Jr., Ph.D.	Director	March 11, 2019
/s/ Geoffrey M. Parker Geoffrey M. Parker	Director	March 11, 2019
/s/ James L. Tyree James L. Tyree	Director	March 11, 2019

Exhibit 10.34

AMENDMENT NO. 1
TO
LOAN AND SECURITY AGREEMENT

THIS AMENDMENT NO. 1 TO LOAN AND SECURITY AGREEMENT (the "First Amendment") is dated as of December 13, 2018 (the "First Amendment Date") and is entered into by and among CHEMOCENTRYX, INC., a Delaware corporation, the several banks and other financial institutions or entities from time to time parties hereto (collectively, referred to as "Lender") and HERCULES CAPITAL, INC., a Maryland corporation, in its capacity as administrative agent for itself and the Lender (in such capacity, the "Agent"). Capitalized terms used herein without definition shall have the same meanings given them in the Loan Agreement (as defined below).

RECITALS

A. Borrower, Agent and Lender have entered into that certain Loan and Security Agreement dated as of December 28, 2017 (as may be amended, restated, or otherwise modified, the "Loan Agreement"), pursuant to which Lender has agreed to extend and make available to Borrower certain advances of money.

B. Borrower has requested and Agent and Lender have agreed to modify certain provisions of the Loan Agreement, subject to the terms and conditions set forth herein.

C. Borrower, Agent and Lender have agreed to amend the Loan Agreement upon the terms and conditions more fully set forth herein.

AGREEMENT

NOW, THEREFORE, in consideration of the foregoing Recitals and intending to be legally bound, the parties hereto agree as follows:

1. AMENDMENTS.

1.1 Definitions. The following definitions in Section 1.1 of the Loan Agreement are hereby amended and restated in their entirety as follows:

"Interest Only Extension Conditions" shall mean satisfaction of each of the following events: (a) no Event of Default shall have occurred and be continuing; and (b) on or before December 15, 2018, Borrower shall have requested Tranche 2 Term Loan Advances in the amount of at least $5,000,000.

"Tranche 1 Amortization Date" means January 1, 2020; provided however, if the Interest Only Extension Conditions are satisfied, then July 1, 2021.

"Tranche 1 Maturity Date" means December 1, 2021; provided however, if the Interest Only Extension Conditions are satisfied, then December 1, 2022.

"Tranche 2 Amortization Date" means July 1, 2021.

"Tranche 2 Maturity Date" means December 1, 2022.

"Tranche 3 Amortization Date" means the first day of the 31st month after the Advance under the Tranche 3 Term Loan is drawn.

1.2 **Exhibits and Schedules**. Schedule 1.1 (Commitments) of the Loan Agreement is hereby amended and restated in its entirety by the revised form of Schedule 1.1 attached to this First Amendment and incorporated herein.

2. **BORROWER'S REPRESENTATIONS AND WARRANTIES**. Borrower represents and warrants that:

2.1 Immediately upon giving effect to this First Amendment (i) the representations and warranties contained in the Loan Documents are true, accurate and complete in all material respects as of the date hereof (except to the extent such representations and warranties relate to an earlier date, in which case they are true and correct as of such date in all material respects), and (ii) no Event of Default has occurred and is continuing with respect to which Borrower has not been notified in writing by Agent.

2.2 Borrower has the corporate power and authority to execute and deliver this First Amendment and to perform its obligations under the Loan Agreement, as amended by this First Amendment.

2.3 The certificate of incorporation, bylaws and other organizational documents of Borrower delivered to Agent and Lender on the Closing Date remain true, accurate and complete and have not been amended, supplemented or restated and are and continue to be in full force and effect.

2.4 The execution and delivery by Borrower of this First Amendment and the performance by Borrower of its obligations under the Loan Agreement, as amended by this First Amendment, have been duly authorized by all necessary corporate action on the part of Borrower.

2.5 This First Amendment has been duly executed and delivered by Borrower and is the binding obligation of Borrower, enforceable against it in accordance with its terms, except as such enforceability may be limited by bankruptcy, insolvency, reorganization, liquidation, moratorium or other similar laws of general application and equitable principles relating to or affecting creditors' rights; and

2.6 As of the date hereof, it has no defenses against the obligations to pay any amounts under the Secured Obligations. Borrower acknowledges that Agent and Lender have acted in good faith and have conducted in a commercially reasonable manner their relationships with Borrower in connection with this First Amendment and in connection with the Loan Documents.

Borrower understands and acknowledges that Agent and Lender are entering into this First Amendment in reliance upon, and in partial consideration for, the above representations and warranties, and agrees that such reliance is reasonable and appropriate.

3. **LIMITATION**. The amendments set forth in this First Amendment shall be limited precisely as written and shall not be deemed (a) to be a waiver or modification of any other term or condition of the Loan Agreement or of any other instrument or agreement referred to therein or to prejudice any right or remedy which Agent or Lender may now have or may have in the future under or in connection with the Loan Agreement (as amended hereby) or any instrument or agreement referred to therein; or (b) to be a consent to any future amendment or modification or waiver to any instrument or agreement the execution and delivery of which is consented to hereby, or to any waiver of any of the provisions thereof. Except as expressly amended hereby, the Loan Agreement shall continue in full force and effect.

4. EFFECTIVENESS. This First Amendment shall become effective upon the satisfaction of all the following conditions:

4.1 Amendments. Borrower, Agent and Lender shall have duly executed and delivered this First Amendment to Agent.

4.2 Payment of Agent and Lender Expenses. Borrower shall have paid (a) an amendment fee of $37,500.00, which fee is due to Lender on or prior to the First Amendment Date and shall be deemed fully earned on such date, and (b) to the extent invoiced on or before the First Amendment Date, all of Agent's and Lender's fees and expenses (including all reasonable attorneys' fees and reasonable expenses) then due and payable by Borrower pursuant to the Loan Agreement.

5. COUNTERPARTS. This First Amendment may be signed in any number of counterparts, and by different parties hereto in separate counterparts, with the same effect as if the signatures to each such counterpart were upon a single instrument. All counterparts shall be deemed an original of this First Amendment. This First Amendment may be executed by facsimile, portable document format (.pdf) or similar technology signature, and such signature shall constitute an original for all purposes.

6. INCORPORATION BY REFERENCE. The provisions of Section 11 of the Loan Agreement shall be deemed incorporated herein by reference, *mutatis mutandis*.

7. LOAN DOCUMENTS. This First Amendment shall constitute a Loan Document.

[*Signatures on following page*]

IN WITNESS WHEREOF, the parties have duly authorized and caused this First Amendment to be executed as of the date first written above.

BORROWER:

CHEMOCENTRYX, INC.

Signature: /s/ Thomas J. Schall
Print Name: Thomas J. Schall
Title: President and Chief Executive Officer

Accepted in Palo Alto, California:

AGENT:

HERCULES CAPITAL, INC.

Signature: /s/ Jennifer Choe
Name Jennifer Choe
Title Assistant General Counsel

LENDER:

HERCULES TECHNOLOGY III, L.P., a
Delaware limited partnership

By: Hercules Technology SBIC Management, LLC
Its: General Partner

By: Hercules Capital, Inc.,
Its: Manager

Signature: /s/ Jennifer Choe
Name Jennifer Choe
Title Assistant General Counsel

HERCULES CAPITAL, INC.

Signature: /s/ Jennifer Choe
Name Jennifer Choe
Title Assistant General Counsel

SCHEDULE 1.1
(Revised)

TERM COMMITMENTS

LENDER	TRANCHE 1 TERM LOAN	TRANCHE 2 TERM LOAN	TRANCHE 3 TERM LOAN	AGGREGATE TERM COMMITMENT
HERCULES TECHNOLOGY III, L.P.	$15,000,000	$ 7,000,000	$ 0	$ 22,000,000
HERCULES CAPITAL, INC.	$ 0	$ 3,000,000	$25,000,000	$ 28,000,000
TOTAL COMMITMENTS	$15,000,000	$10,000,000	$25,000,000	$ 50,000,000

Exhibit 21.1

SUBSIDIARIES OF THE REGISTRANT

The following is a list of subsidiaries of the Registrant as of December 31, 2018.

Company	Jurisdiction of Incorporation
ChemoCentryx Limited	United Kingdom
ChemoCentryx Ireland Limited	Ireland

Exhibit 23.1

CONSENT OF INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM

We consent to the incorporation by reference in the Registration Statements (Forms S-3 No. 333-187387 and No. 333-210731) of ChemoCentryx, Inc. and the Registration Statement (Form S-8 No. 333-179507) pertaining to the ChemoCentryx, Inc. 2012 Equity Incentive Award Plan, the ChemoCentryx, Inc. Amended and Restated 2002 Equity Incentive Plan, the ChemoCentryx, Inc. Amended and Restated 1997 Stock Option/Stock Issuance Plan, and the ChemoCentryx, Inc. 2012 Employee Stock Purchase Plan of our reports dated March 11, 2019, with respect to the consolidated financial statements of ChemoCentryx, Inc. and the effectiveness of internal control over financial reporting of ChemoCentryx, Inc. included in this Annual Report (Form 10-K) of ChemoCentryx, Inc. for the year ended December 31, 2018.

/s/ ERNST & YOUNG LLP

Redwood City, California
March 11, 2019

Exhibit 31.1

CERTIFICATION OF CHIEF EXECUTIVE OFFICER
PURSUANT TO SECTION 302 OF THE SARBANES-OXLEY ACT OF 2002

I, Thomas J. Schall, Ph.D., certify that:

1. I have reviewed this Annual Report on Form 10-K of ChemoCentryx, Inc. for the fiscal year ended December 31, 2018;

2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;

3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;

4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:

a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;

b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;

c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and

d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and

5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):

a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and

b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

/s/ Thomas J. Schall, Ph.D.
Thomas J. Schall, Ph.D.
Chief Executive Officer

Date: March 11, 2019

Exhibit 31.2

CERTIFICATION OF CHIEF FINANCIAL OFFICER
PURSUANT TO SECTION 302 OF THE SARBANES-OXLEY ACT OF 2002

I, Susan M. Kanaya, certify that:

1. I have reviewed this Annual Report on Form 10-K of ChemoCentryx, Inc. for the fiscal year ended December 31, 2018;

2. Based on my knowledge, this report does not contain any untrue statement of a material fact or omit to state a material fact necessary to make the statements made, in light of the circumstances under which such statements were made, not misleading with respect to the period covered by this report;

3. Based on my knowledge, the financial statements, and other financial information included in this report, fairly present in all material respects the financial condition, results of operations and cash flows of the registrant as of, and for, the periods presented in this report;

4. The registrant's other certifying officer(s) and I are responsible for establishing and maintaining disclosure controls and procedures (as defined in Exchange Act Rules 13a-15(e) and 15d-15(e)) and internal control over financial reporting (as defined in Exchange Act Rules 13a-15(f) and 15d-15(f)) for the registrant and have:

a) Designed such disclosure controls and procedures, or caused such disclosure controls and procedures to be designed under our supervision, to ensure that material information relating to the registrant, including its consolidated subsidiaries, is made known to us by others within those entities, particularly during the period in which this report is being prepared;

b) Designed such internal control over financial reporting, or caused such internal control over financial reporting to be designed under our supervision, to provide reasonable assurance regarding the reliability of financial reporting and the preparation of financial statements for external purposes in accordance with generally accepted accounting principles;

c) Evaluated the effectiveness of the registrant's disclosure controls and procedures and presented in this report our conclusions about the effectiveness of the disclosure controls and procedures, as of the end of the period covered by this report based on such evaluation; and

d) Disclosed in this report any change in the registrant's internal control over financial reporting that occurred during the registrant's most recent fiscal quarter (the registrant's fourth fiscal quarter in the case of an annual report) that has materially affected, or is reasonably likely to materially affect, the registrant's internal control over financial reporting; and

5. The registrant's other certifying officer(s) and I have disclosed, based on our most recent evaluation of internal control over financial reporting, to the registrant's auditors and the audit committee of the registrant's board of directors (or persons performing the equivalent functions):

a) All significant deficiencies and material weaknesses in the design or operation of internal control over financial reporting which are reasonably likely to adversely affect the registrant's ability to record, process, summarize and report financial information; and

b) Any fraud, whether or not material, that involves management or other employees who have a significant role in the registrant's internal control over financial reporting.

/s/ Susan M. Kanaya
Susan M. Kanaya
Chief Financial and Administrative Officer

Date: March 11, 2019

Exhibit 32.1

CERTIFICATION
Pursuant to Section 906 of the Sarbanes-Oxley Act of 2002
(Subsections (a) and (b) of Section 1350, Chapter 63 of Title 18, United States Code)

In connection with the Annual Report on Form 10-K of ChemoCentryx, Inc. (the "Company") for the period ended December 31, 2018, as filed with the Securities and Exchange Commission on the date hereof (the "Report"), I, Thomas J. Schall, Ph.D., as Chief Executive Officer of the Company, certify, pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002, that to my knowledge:

1. The Report fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934; and

2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

Date: March 11, 2019

/s/ Thomas J. Schall, Ph.D.
Thomas J. Schall, Ph.D.
Chief Executive Officer

The foregoing certification is being furnished solely to accompany the Report pursuant to 18 U.S.C. § 1350, and is not being filed for purposes of Section 18 of the Securities Exchange Act of 1934, as amended, and is not to be incorporated by reference into any filing of the Company, whether made before or after the date hereof, regardless of any general incorporation language in such filing. A signed original of this written statement required by Section 906 has been provided to the Company and will be retained by the Company and furnished to the Securities and Exchange Commission or its staff upon request.

Exhibit 32.2

CERTIFICATION
Pursuant to Section 906 of the Sarbanes-Oxley Act of 2002
(Subsections (a) and (b) of Section 1350, Chapter 63 of Title 18, United States Code)

In connection with the Annual Report on Form 10-K of ChemoCentryx, Inc. (the "Company") for the period ended December 31, 2018, as filed with the Securities and Exchange Commission on the date hereof (the "Report"), I, Susan M. Kanaya, as Chief Financial and Administrative Officer of the Company, certify, pursuant to 18 U.S.C. Section 1350, as adopted pursuant to Section 906 of the Sarbanes-Oxley Act of 2002, that to my knowledge:

1. The Report fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934; and

2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

Date: March 11, 2019

/s/ Susan M. Kanaya
Susan M. Kanaya
Chief Financial and Administrative Officer

The foregoing certification is being furnished solely to accompany the Report pursuant to 18 U.S.C. § 1350, and is not being filed for purposes of Section 18 of the Securities Exchange Act of 1934, as amended, and is not to be incorporated by reference into any filing of the Company, whether made before or after the date hereof, regardless of any general incorporation language in such filing. A signed original of this written statement required by Section 906 has been provided to the Company and will be retained by the Company and furnished to the Securities and Exchange Commission or its staff upon request.