

2017



GREAT SCIENCE
GREAT MEDICINE

CHEMOCENTRYX

The strength of an organization relates to the depth of its foundation.

At ChemoCentryx (CCXI) our foundation runs deep, built upon years of experience in an industry that rewards only those that innovate and persevere. We are executing on our long-term plan, creating value. Our organization is strong and resilient, every year we aim to transform our goal of new medicines for patients and excellent returns for investors into a self-sustaining, practical reality. I am proud to say that the accomplishments of 2017 were major steps toward this reality. In this letter I will briefly review these achievements and discuss how they position us for future growth.

2017 was a watershed year for CCXI.

In achieving both near term tactics and long-range plan, 2017 was an excellent year. Five key accomplishments exemplify this:

- 1) The ADVOCATE Phase III pivotal trial (of our lead drug avacopan) proceeded at an unprecedented pace.
- 2) We launched clinical trials in two other indications: C3 Glomerulopathy, C3G, (an additional indication for avacopan); and also in Focal Segmental Glomerulosclerosis, with CCX140, our second kidney asset.
- 3) We submitted an application for a Conditional Marketing Authorization (CMA) in Europe for avacopan in ANCA vasculitis, which was validated by the European Medicines Agency (EMA).
- 4) We made advances in our pancreatic cancer program with CCX872, where our clinical trial showed very promising 18-month overall survival data.
- 5) We bolstered our balance sheet considerably. 2017 was a year in which we took in more money than we spent, despite all our activity and progress on multiple clinical trials during the year.

Any one of these five achievements would make for a good year. Together, they represent a considerable leap forward in our plan to become a fully integrated biopharmaceutical company.

Advanced CCXI programs focus on kidney disease, an immense opportunity for new medicines, where the need in orphan renal patients is particularly acute.

The two most clinically advanced drug candidates at CCXI are avacopan and CCX140. Avacopan blocks the receptor for the complement fragment C5a (C5aR), a known driver of destruction in autoimmune diseases, and CCX140 blocks the chemokine receptor known as CCR2, which is implicated in chronic inflammation and increasingly in kidney dysfunction. Both avacopan and CCX140 represent innovative precision medicine approaches: they selectively target chemoattractant receptors that specifically drive inflammatory and autoimmune cells responsible for tissue destruction in the diseases to be treated. Unlike current standard therapies (where the idea has been to limit autoimmune disease by generally suppressing the immune system), avacopan and CCX140 do not cause systemic immunosuppression.

We are actively pursuing both avacopan and CCX140 in kidney disease, particularly orphan kidney disease. We believe that the clinical needs and the commercial opportunities for new therapies in kidney disease are immense. Consider the following astonishing fact: US taxpayers spend about as much per year just on end stage renal disease as we do on all other basic medical research and medical science combined. (ESRD costs ~8% of total Medicare budget; this is bigger than the entire annual budget for the National Institutes of Health.)

That fact alone should say something about need and opportunity.

Orphan diseases are particularly tractable for a company of our size and scope, and the unmet needs for patients with orphan diseases of the kidney are profound. Take, for example, anti-neutrophil cytoplasmic autoantibody (or ANCA) vasculitis. ANCA vasculitis occurs when inflammatory cells in the blood (the neutrophils) become activated by auto-antibodies and the body's complement system. In particular, the C5a complement fragment, binds to its receptor (the

C5aR) on the neutrophils, and these complement-activated neutrophils attack vasculature with chronic inflammation (vasculitis). This ultimately destroys the blood vessels, leading to organ failure particularly of the kidney.

ANCA vasculitis is currently treated with drugs that are designed to suppress the immune system (such as cyclophosphamide or rituximab) combined with the long-term use of high doses of corticosteroids (such as prednisone). Steroids have been shown to cause serious adverse events, including death. ANCA vasculitis remains a serious, potentially life-threatening disease, with at least an 11% first-year mortality rate. Astonishingly, current treatments contribute to most of this mortality (mostly due to serious infections and other complications of steroids). And, despite all the risks of current therapy, the standard regimen can also be slow to act in relieving signs and symptoms of ANCA vasculitis, and does not provide a durable benefit. The standard of care regimen is a source of much of the *total burden of disease* in ANCA, and the inadequacies of current care are a large part of the unmet need.

We have a new approach with avacopan.

Precision targeting leads to precision medicine: avacopan represents a differentiated approach to complement-related diseases.

Avacopan is the first late-stage drug candidate to block only the C5a receptor in the complement system. By blocking the C5aR, we leave the rest of the complement cascade and the immune system functioning normally. Avacopan's action selectively stifles the activation of the harmful neutrophils that cause the ANCA disease, avoiding major limitations of other complement system-targeting therapies that are in development.

In short, avacopan possesses a highly specific mechanism of action. It is the first targeted therapy developed for ANCA vasculitis. We and others believe avacopan could make the current, decades-old treatment regimen in ANCA vasculitis irrelevant, marking a paradigm shift in the treatment of ANCA. With avacopan, data suggests that patients can get better, get better faster, stay healthy, and avoid the physical and psychological side effects that come with the previous century's standard of care, enjoying a markedly better quality of life.

Avacopan is also fundamentally different from other drug candidates in development in what might be called the 'complement cascade intervention space':

- Avacopan is an orally administered inhibitor of C5aR, not an injectable or infusible protein, such as an antibody;
- This is important because antibody therapies may become immunogenic, losing their effects over time;
- Also, by specifically targeting C5aR (known to drive inflammatory destruction), we are not blocking other, beneficial effects of C5a occurring through other pathways.

In 2017 we achieved key business milestones and raised awareness of ANCA Vasculitis patients' needs.

Late in 2017 the EMA accepted for review our conditional registration dossier, based on Phase II clinical data, in support of a CMA for avacopan for the treatment of patients with ANCA vasculitis. The application and its validation revealed the maturity of our avacopan program for ANCA vasculitis (e.g., in manufacturing, toxicology, and other exacting standards required for a product license), and triggered a \$50 million milestone payment from our commercial partner Vifor Pharma.

The validation of our CMA application by the EMA was a pivotal business milestone for us. It reinforced our belief that status quo of standard care is simply not good enough for ANCA patients. Innovative therapies, urgently needed, deserve accelerated assessment. The CMA validation heightened the awareness of the plight of ANCA vasculitis patients. For all of CCXI, I am proud to say to the ANCA community: your cause is our cause; we are determined to succeed.

We are executing on our 'pipeline in a drug' strategy for avacopan.

C5aR inhibition is relevant to several very high value / high need clinical conditions. We intend to maximize the value of avacopan by pursuing these conditions in parallel.

First in ANCA vasculitis – the Phase III ADVOCATE pivotal trial had excellent momentum through 2017. We opened 200 sites in 20 countries around the world for

ADVOCATE and we are on track to complete enrollment (300 patients) by the middle of 2018. Such progress is remarkable because orphan diseases are by their very nature difficult to enroll in randomized controlled trials which involve more than a handful of patients. This is one of the historical hurdles to advancing innovative medicines in orphan diseases. But the facts of the ADVOCATE trial are impressive: it is the largest clinical trial of its type ever for ANCA vasculitis, and on pace for the fastest recruitment rate in a randomized controlled clinical trial in this important disease. The enthusiastic welcome that physicians and patients alike are giving the ADVOCATE trial may also be an indication of the pent-up need that modern drugs like avacopan exemplify: potentially changing the treatment paradigm in ANCA and other complement-related diseases.

Although the ADVOCATE Phase III trial is a major focus for us, it is just the beginning. We are also moving ahead with clinical trials which are designed ultimately to support registration in kidney diseases for which there are currently no FDA approved treatments.

Specifically, we are also enrolling a clinical trial of avacopan in C3G, a rare disorder that affects younger patients, requiring dialysis and, too often, kidney transplant. There is simply no effective treatment today: immuno-suppressive drugs are minimally beneficial and even kidney transplants frequently fail. We and other experts believe we have designed the most stringent, controlled trial yet for C3G.

Other very high value indications also exist for avacopan outside of renal disease. One such indication is the debilitating and deforming skin disease, hidradenitis suppurativa (HS). HS is widely regarded as only marginally controlled by the lone existing approved therapy, an anti-TNF antibody. Yet even with a less than adequate benefit, that current therapy commands revenues approximating \$1 billion per year in HS. Recent clinical evidence suggests complement C5a drives HS, and we intend to launch our clinical development program for the use of avacopan in patients with HS soon.

CCX140, a second kidney disease asset, bolsters our renal therapy franchise.

While avacopan continues to advance in late-stage clinical trials, we are launching trials of the second drug candidate in our kidney franchise, CCX140, in the orphan kidney indication Focal Segmental Glomerulosclerosis, FSGS. Like C3G, there is simply no approved treatment for FSGS.

We have significant clinical experience with CCX140. Previously, we successfully completed a one-year Phase II trial in diabetic nephropathy (DN) patients. In DN, CCX140 treatment significantly and durably lowered proteinuria (a sign of improved kidney function) while being very well tolerated over its long dosing period.

That finding may augur well for CCX140, since proteinuria lowering may constitute the actual registration endpoint in certain FSGS patient populations. One of our CCX140 trials involves sub-nephrotic, primary FSGS patients and the other is in primary FSGS patients with nephrotic syndrome. In nephrotic patients, we believe a significant decrease in proteinuria may constitute an accelerated approval endpoint. We intend to make significant progress in the CCX140 FSGS program in 2018.

CCXI owns entirely its kidney assets in the US, and outside the US we intend to commercialize with our partner Vifor Pharma.

Especially as avacopan undergoes its CMA review, it is important to note that we have a plan to bring our novel renal therapeutics to market. CCXI retains 100% of the rights for both avacopan and CCX140 in the US and China. We are developing our US commercial plan, having hired senior executive talent with deep expertise in marketing of orphan products. For sales abroad, our very experienced commercial partner in renal care, Vifor Pharma, has rights to commercialize avacopan and CCX140 abroad, with ChemoCentryx receiving significant royalties on sales from those territories. A large part of the global commercial strategy is in place and upon approval we intend to be ready to create value with our kidney products around the world.

New opportunities in our broad pipeline portfolio further enhance the CCXI value proposition.

It is important to know that the CCXI core technology, i.e., modifying the trafficking of cells, is therapeutically relevant to cancer as well. For example, in pancreatic cancer, we reported promising data from a clinical trial of our second CCR2 inhibitor, CCX872, in advanced/metastatic pancreatic cancer. The data showed overall survival (in all patients randomized) of 29% at 18 months

with CCX872 and FOLFIRINOX combination therapy. This was a marked advance over previously published overall survival rates of 18.6% at 18 months using FOLFIRINOX treatment alone in pancreatic cancer.

The findings demonstrated that improved patient survival could result from selectively blocking the immune suppressing cells that CCR2 maintains in the tumor environment. This new approach - which aims to mobilize the body's own potential for a powerful anti-tumor immune response - is particularly important because other therapies such as checkpoint inhibitors do not seem to work well alone in pancreatic cancer. CCX872 potentially could make the checkpoint inhibitors more effective. Our plan is to advance CCX872 in combination with other therapies.

Capital efficiency: a CCXI tradition.

While I am delighted to report excellent progress in 2017 from our clinical trials, I am equally delighted with our ability to secure non-dilutive financing that allows CCXI to energetically advance its pipeline. We did very well in 2017 in bringing revenues from partnership sources, continuing to use our capital in a highly efficient way.

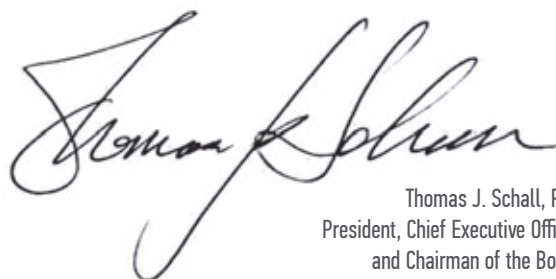
In fact, CCXI was cash flow positive for 2017. We brought in \$20M from Vifor for expanded rights to market Avacopan (ex US) in territories including Japan, and \$50M for the avacopan CMA validation. Additionally, we negotiated a \$50 million growth capital facility that we entered into at the end of 2017. Importantly, we believe that we now have sufficient funds available to advance avacopan well through topline data from our Phase III ADVOCATE trial and into additional registration filings in the U.S. and the E.U. A history of capital efficiency has become one of our hallmarks; one which has been well appreciated by our investor base. Our pledge is to continue to invest responsibly in high value programs to create value.

In 2018 we will continue to execute assertively on our plan.

Foundations are only good for what can be built upon them. The impressive accomplishments of 2017 provide the base for building more value in 2018. This year we look forward to completing enrollment of ADVOCATE, working with the EMA as they review of our CMA application, and advancing avacopan and CCX140 in registration-supporting trials in additional renal diseases. Also, we are particularly excited to develop avacopan in hidradenitis suppurativa. We have a healthy balance sheet to fund our clinical activities and to take us toward potential registration filings in the U.S. and E.U.

In the evolution of CCXI, we enter into a pivotal and exciting period. It is a time that we expect will lead ultimately to the commercialization of our innovative therapeutics and becoming a fully integrated biopharmaceutical company. As we move closer to commercialization we've added to the executive ranks seasoned veterans in commercial strategy, clinical development, quality, and other disciplines required for a mature enterprise.

Citizens of our CCXI community - the exceedingly talented clinicians, life scientists, medicinal chemists, financial and regulatory experts, and other highly dedicated professionals -- have a deep and utter commitment to create new medicines for patients who for too long have been given too little and who have endured too much. We at CCXI pledge to work unstintingly to create value for these patients, for our community, and for our shareholders. Optimism abounds, glistening and effulgent among our many prospects. Our message is simple: the future of our enterprise has never been brighter.



Thomas J. Schall, PhD
President, Chief Executive Officer
and Chairman of the Board

CCXI DISCOVERY PLATFORM HAS CREATED A BROAD PIPELINE OF UNIQUE SMALL MOLECULE THERAPEUTICS

THERAPEUTIC AREA	DRUG / TARGET	INDICATION	Preclinical	Phase I	Phase II	Phase III
Complement Inhibition in Orphan Diseases	Avacopan (formerly CCX168)/ C5aR	ANCA-ASSOCIATED VASCULITIS				
		C3 GLOMERULOPATHY				
		HIDRADENITIS SUPPURATIVA				
		ATYPICAL HEMOLYTIC UREMIC SYNDROME				
Chronic and Orphan Kidney Diseases	CCX140/ CCR2	CKD, INCLUDING ORPHAN FOCAL SEGMENTAL GLOMERULOSCLEROSIS				
		DIABETIC NEPHROPATHY, SUCCESSFULLY COMPLETED PHASE II				
Other Inflammatory and Autoimmune Diseases	CCX507/CCR9	IBD: ULCERATIVE COLITIS				
	CCX587/CCR6	TH17 DRIVEN DISEASE, e.g. PUSTULAR PSORIASIS				
Immuno-Oncology	CCX872/ CCR2	ADVANCED PANCREATIC CANCER				
	CCR1, CXCR2, Other CKR	OTHER ONCOLOGY INDICATIONS				

**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, 2017**

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
(State or Other Jurisdiction of
Incorporation or Organization)

94-3254365
(I.R.S. Employer
Identification No.)

850 Maude Avenue
Mountain View, California
(Address of Principal Executive Offices)

94043
(Zip Code)

(650) 210-2900

(Registrant's Telephone Number, Including Area Code)

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

<u>Title of Each Class</u>	<u>Name of Each Exchange on Which Registered</u>
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 and posted on its corporate Web site, if any, every Interactive Data File required to be submitted and posted 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 and post 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. (Check one):

Large accelerated filer Accelerated filer

Non-accelerated filer (Do not check if a smaller reporting company) 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 \$197.3 million, based on the closing price of the registrant's common stock on the Nasdaq Global Select Market of \$9.36 per share.

The number of outstanding shares of the registrant's common stock, par value \$0.001 per share, as of February 28, 2018 was 48,920,715.

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 2018 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, 2017.

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CHEMOCENTRYX, INC.
FORM 10-K — ANNUAL REPORT
For the Fiscal Year Ended December 31, 2017

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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 subsidiary 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 selectively blocks a specific chemoattractant receptor, leaving the rest of the immune system intact. Our drug candidates are small molecules, which are orally administered, offering 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 commitments and milestones to support the clinical development of our leading two 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 kidney disease, particularly 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) — Complement Inhibition 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; (ii) complement 3 glomerulopathy, or C3G, a debilitating disease that can lead to kidney failure; and (iii) atypical hemolytic uremic syndrome, or aHUS, a rare, life threatening disease.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV, C3G and aHUS 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 enrolling approximately 300 patients at approximately 200 sites in the United States, Canada, Europe, Australia, and New Zealand. We expect to complete patient enrollment of the Phase III ADVOCATE trial in mid-2018. We recently launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G and plan to initiate clinical studies for the treatment of patients with hidradenitis suppurativa, or HS, in 2018. Meanwhile, we are actively dosing aHUS patients under compassionate use protocols as we explore appropriate doses and dosing regimen for that indication.

In December 2017, our Conditional Marketing Authorization, or CMA, application for avacopan in the treatment of patients with AAV was accepted for review by the EMA's Committee for Medicinal Products for Human Use, or CHMP. Under the terms of our kidney health alliance with Vifor (International) Ltd., this

validation of the avacopan CMA application by the EMA resulted in a \$50.0 million milestone (see “Kidney Health Alliance with Vifor”).

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.

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 highest proteinuric patients. Based on the safety and efficacy data related to reduction in proteinuria observed in the Phase II trial in DN, we launched our clinical development program of CCX140 for the treatment of patients with primary FSGS, 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 commercial 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 this second agreement, we again retained all rights to the United States and China and we granted Vifor worldwide 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.

We have secured \$205 million in upfront cash payments and commitments and milestones pursuant to our agreements with Vifor and are eligible for additional substantial milestone payments. Through our alliance, we maintain the commercial rights to avacopan and CCX140 in the United States and China, and also retain control of the clinical development programs for orphan renal disease. Vifor gained the commercial 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 contract, 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 the science has led us to focus 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 other 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 start to earn revenues and royalties from the commercialization of our late stage kidney disease franchise.

Our Drug Candidate Pipeline

CCXI Late Stage: Kidney Disease Franchise

THERAPEUTIC AREA	DRUG/TARGET	INDICATION	PRECLINICAL	PHASE I	PHASE II	PHASE III
Complement Inhibition in Orphan Diseases	Avacopan (formerly CCX168)/ C5aR	ANCA-ASSOCIATED VASCULITIS				
		C3 GLOMERULOPATHY				
		HIDRADENITIS SUPPURATIVA				
		ATYPICAL HEMOLYTIC UREMIC SYNDROME				
Chronic and Other Orphan Kidney Diseases	CCX140/ CCR2	CKD, INCLUDING ORPHAN FOCAL SEGMENTAL GLOMERULOSCLEROSIS				
		DIABETIC NEPHROPATHY, SUCCESSFULLY COMPLETED PHASE II				

CCXI Early Stage Drug Candidates

THERAPEUTIC AREA	DRUG/TARGET	INDICATION	PRECLINICAL	PHASE I	PHASE II	PHASE III
Other Inflammatory and Autoimmune Diseases	CCX507/ CCR9 CCX587/ CCR6	IBD: ULCERATIVE COLITIS				
		TH17 DRIVEN DISEASE, E.G. PUSTULAR PSORIASIS				
Immuno-Oncology	CCX872/ CCR2 CCR1 CXCR2, Other CKR	ADVANCED PANCREATIC CANCER				
		OTHER ONCOLOGY INDICATIONS				

Late Stage Drug Candidates

Avacopan (Formerly CCX168) — Complement Inhibition 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, C3G and aHUS. 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. We recently also launched a clinical trial to study avacopan for the treatment of patients with C3G and plan to initiate clinical studies for the treatment of patients with HS in 2018. Meanwhile, we are actively dosing aHUS patients under compassionate use protocols as we explore appropriate doses and dosing regimen for that indication. All of these activities are designed to potentially support registration of avacopan in these indications. 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. We retain all rights to commercialize avacopan in the United States and China and also retain control of the clinical development programs for orphan renal diseases.

AAV, C3G and aHUS are all orphan autoimmune diseases that are characterized by inflammation that often affects the kidneys, among other major organs.

ANCA-Associated Vasculitis (AAV)

AAV is an orphan, severe, and often fatal autoimmune disease that is caused by autoantibodies called anti-neutrophil cytoplasmic autoantibodies and is characterized 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, and more than 75,000 people in Europe, with at least 7,500 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 glucocorticoid (steroid) administration. Following initial treatment, up to 30% of patients relapse within six to 18 months, and approximately 50% of all patients will 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 disease 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. Activation of the complement pathway occurs with 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 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 four Phase I clinical trials with avacopan in a total of 102 healthy subjects. 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 cyclophosphamide, or CYC, or rituximab, 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 eGRF 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 to enroll approximately 300 patients with newly diagnosed or relapsing AAV at approximately 200 sites in the United States, Canada, Europe, Australia, and New Zealand. 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 will also receive a standard background

immunosuppressant, either CYC 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. We expect to complete patient enrollment of the Phase III ADVOCATE trial in mid-2018 and believe, if successful, could form the basis of avacopan's registration for the treatment of AAV in Europe and in the United States.

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 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 and is currently under review by the EMA's CHMP.

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 about 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 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 invariably 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 recently launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G. The clinical trial will include approximately 44 patients with C3G, including both C3 Glomerulonephritis, or C3GN, 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 will start study drug treatment on Day 1. Patients will take 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.

Avacopan “Special Needs” Protocol in the United Kingdom

One patient with treatment refractory C3GN, one of the subtypes of C3G, was treated successfully with 30mg avacopan twice daily for approximately 18 months under a “Special Needs” program in the United Kingdom. This patient, previous to receiving avacopan, had progressive decline in renal function despite treatment with immunosuppressants, rituximab, and glucocorticoids, as well as a kidney transplant.

A biopsy of the patient’s kidney that was done before avacopan treatment showed severe inflammation. After two months on treatment with avacopan, a repeat biopsy showed the inflammation had decreased, and by six months a biopsy appeared close to normal. Before treatment, the patient’s kidney function was declining rapidly; during treatment the kidney function stabilized. The patient’s quality of life improved markedly, and he was able to attend college and participate in social activities.

Based on the results in this and the Phase II study in AAV, we launched our clinical development program in C3G.

Hidradenitis Suppurativa (HS)

Hidradenitis suppurativa, or 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 disease 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 plan to initiate clinical studies of avacopan in HS by the end of 2018.

Atypical Hemolytic Uremic Syndrome (aHUS)

aHUS is a genetic, chronic, rare disease that is caused by the formation of blood clots within small blood vessels, or thrombosis, throughout the body. aHUS affects both adults and children and can progressively damage vital organs, including the kidneys, but also other organs such as the brain, heart, lungs, gastrointestinal tract, and pancreas. These clots can cause serious medical problems if they restrict or block blood flow.

As a result of clot formation in small blood vessels, people with aHUS experience kidney damage and acute kidney failure that lead to ESRD in about half of all cases. These life-threatening complications prevent the kidneys from filtering plasma and eliminating waste products from the body effectively.

Role of C5a and C5aR in aHUS

aHUS often results from a combination of environmental and genetic factors. The genes associated with aHUS provide instructions for making proteins involved in regulating the complement system. In aHUS, the regulatory proteins that prevent uncontrolled activation of the complement system are defective due to gene mutations. The resulting uncontrolled activation of the complement system, including uncontrolled production of the anaphylatoxin C5a, results in damage to the vasculature and organs such as the kidneys.

The fact that C5a and its receptor C5aR play a role in the pathogenesis of aHUS is supported by studies in mice. Mice deficient in complement factor H, or CFH, develop proliferative glomerulonephritis, which is improved in mice where both the CFH and C5aR genes are deleted. Mice lacking C5aR were significantly protected from functional renal disease as assessed by blood urea nitrogen levels. This is relevant as loss-of-function CFH mutations are relatively common in humans with aHUS. In addition, C5a can prime neutrophils and enhance neutrophil activation. C5a, acting on C5aR, is a potent neutrophil chemoattractant and agonist, which triggers neutrophil aggregation. Further, C5a activates endothelial cells, promoting retraction and increased permeability.

Avacopan, as a potent and specific inhibitor of C5aR, may therefore be effective in the treatment of patients with aHUS. Compared to intravenously administered eculizumab, avacopan is a convenient, orally-administered treatment. Avacopan blocks the effect of C5a without compromising the formation of the C5b-9 MAC, which is important in fighting *Neisseria* infections. Since avacopan is a small molecule, manufacturing cost is anticipated to be lower than protein-based drugs such as eculizumab. As a small molecule, avacopan has a shorter plasma half-life (terminal half-life is approximately 70 hours) than eculizumab (272 hours, according to eculizumab prescribing information). Therefore, in the event of an undesirable adverse event requiring discontinuation of treatment, plasma clearance would be faster with avacopan.

Limitations of Current Therapies

Current aHUS treatment has limited efficacy or is very expensive, and as a result, is not a practical option for many patients with aHUS.

Plasma exchange or infusion has decreased mortality from 50% to 25% in patients with aHUS. In patients with CFH mutations, leading to de-regulation of complement activation, plasma exchange or infusion resulted in partial or complete remission in approximately 60% of patients. Plasma exchange with immunosuppressive therapy such as steroids and azathioprine or mycophenolate mofetil and RTX resulted in long-term dialysis-free survival in 60% to 70% of patients. Patients may become non-responsive to plasma exchange or infusion. It is also debatable whether renal transplantation is appropriate for patients with aHUS with ESRD, as the disease recurs in approximately 50% of patients after transplantation, and graft failure occurs in 80% to 90% with recurrent disease.

Eculizumab has been approved by the FDA for treatment of patients with aHUS to inhibit complement-mediated thrombotic microangiopathy. Eculizumab treatment improves the disease activity based on improvement in platelet count, lactate dehydrogenase, hemoglobin, and serum creatinine levels, and need for plasma exchange, infusion, or dialysis. Eculizumab is an anti-C5 antibody, designed to block the conversion of C5 to C5a and C5b. Eculizumab needs to be administered by frequent intravenous infusion, is associated with an increased risk of *Neisseria* infections, and currently can cost approximately \$500,000 per year in the United States.

Clinical Development in aHUS

Patients with aHUS have increased activity of the complement system, leading to increased conversion of complement component C5 to two active components: C5a and C5b. C5a binds to its receptor, C5aR, leading to pro-inflammatory activation of vascular endothelial cells and neutrophils. *Ex vivo* studies demonstrate that in the presence of increased C5a, neutrophils bind to vascular endothelial, followed by accumulation of platelets and formation of microscopic thrombi (clots) and damage to the vessel. This microscopic thromboangiopathy is characteristic of aHUS flares, and leads to damage to tissue and organ systems.

Eculizumab, a monoclonal antibody that binds to C5 and prevents conversion of C5 to C5a and C5b, prevents formation of microscopic thrombi *ex vivo*, and is effective and indicated for treatment of aHUS. To test the hypothesis that avacopan should also be tested in aHUS, *ex vivo* studies were performed at an expert center by external scientists. Specifically, vascular endothelial cells were exposed to blood from patients with aHUS and examined for microscopic thrombi. Then the patients with aHUS were treated with avacopan, and the study was repeated. The results demonstrated that avacopan reduced the number and size of thrombi. The positive effect of avacopan was similar to that observed with eculizumab, or with soluble C5aR1, which also binds to C5a.

Based on these encouraging *in vitro* findings, we performed a Phase II pilot clinical trial with avacopan in six patients with aHUS who were on dialysis. The primary efficacy objective of the trial was to evaluate whether treatment with avacopan may reduce thrombosis formation in chronic dialysis patients with aHUS. In 2016, we reported positive data from five patients from this clinical trial. After 14 days of dosing in aHUS patients, the mean decrease in thrombus size was 83%, with 100% inhibition in three of these patients. Treatment appeared to be mechanism specific, as the thrombus size returned to baseline levels when avacopan treatment was stopped. There was one serious adverse event, not considered related to avacopan use, in a patient with long-standing cardiovascular and renal disease of cardiac asystole. Two patients in the study had relatively low platelet counts which appeared to improve on avacopan treatment. We are actively dosing aHUS patients under compassionate use protocols as we explore appropriate doses and dosing regimen for that indication.

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 all three orphan 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 and China, 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. 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 and China. 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.

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 patients across seven clinical trials. These clinical studies 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.

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 m²), 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 m².

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) Non-nephrotic primary FSGS patients with moderate proteinuria (>1 g/g); and
- (ii) Primary FSGS patients with nephrotic syndrome (heavy proteinuria >3.5 g/g).

The first trial is a randomized controlled, dose ranging study with the aim to evaluate the effect of treatment with CCX140 in subjects with non-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 the study is to evaluate the effect of CCX140 on proteinuria in patients with primary FSGS with nephrotic syndrome, 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 United 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 certain territories outside 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 retain marketing rights for rare renal disease in the United States and China, while Vifor has commercialization rights in the rest of the world. We will be 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.

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 2018, approximately 55,440 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 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, CCR5, 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 programmed cell death protein-1, or 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 December 2017, 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-CT26 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.

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 newly 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 expect to nominate a development candidate in 2018.

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 2017, 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. Results from this work were published in a peer reviewed journal article in October 2017. CCR6 antagonists present a novel therapeutic approach to treating 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.

Our patent estate, on a worldwide basis, includes approximately 824 issued or allowed patents and approximately 197 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. There are approximately 87 issued or allowed patents and 41 patent applications pending for avacopan, our lead drug candidate in the C5aR program. With respect to our drug candidates in the CCR2 program, we have approximately 77 issued or allowed patents and 26 patents pending worldwide relating to their chemical composition or use thereof. With respect to the CCR1 and CCR9 chemokine receptors, we have approximately 468 issued or allowed patents and 120 patents pending worldwide relating to their chemical composition or use thereof. We have approximately 143 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs. We have approximately 49 issued patents and 10 pending applications relating to other small molecule compounds and approximately 140 issued patents relating to our novel biological discoveries and our proprietary screening and drug development technologies.

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, China, Hong Kong, Israel, Mexico, Singapore 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 and Canada 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.

CCX140 is covered by two patent families with patents in the United States for the composition-of-matter of CCX140 and pharmaceutical compositions thereof that will expire in 2026 and 2028, respectively (not including

patent term adjustment and 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). Patent applications are pending in a few countries including Brazil and the United States, which, if issued, are anticipated to expire in 2028 (not including patent term extensions or supplementary protection certificates that may be available).

CCX872 is covered by a patent family in the United States for the composition-of-matter of CCX872 and pharmaceutical compositions thereof that will expire in 2026, not including patent term extension that may be available to extend the term of the granted patents.

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 to determine priority of invention.

Competition

We compete in the pharmaceutical, biotechnology and other related markets that address AAV, aHUS, HS, C3G, DN, 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[®]).

CCX140, our first CCR2 inhibitor, if approved for marketing by the FDA or other regulatory agencies for the treatment of DN, 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 DN. 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, Biolinex, 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 compliance with an international guideline for the ethical conduct of clinical research known as the Declaration of Helsinki and/or the laws and regulations of the country or countries in which the clinical trial is performed, whichever provides the greater protection to the participants in the clinical trial. 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 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 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 \$74,792 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 require 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 \$11,052 per failure up to an aggregate of \$165,786 per year (or up to an aggregate of \$1.105 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 \$55,910 per violation, not to exceed \$1.68 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 marketing 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 marketing 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, 2017, we had 66 full-time employees, 30 of whom hold Ph.D.s, M.D.s or both. Of our total workforce, 52 employees are engaged in research and development, and 14 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.

Research and Development

We invested \$49.5 million, \$37.9 million and \$33.2 million in research and development in the years 2017, 2016, and 2015, respectively.

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 a wholly owned subsidiary, ChemoCentryx Limited, organized under the laws of the United Kingdom that is currently inactive.

Financial Information about Segments

We operate only in one business segment, which is the commercialization and development of pharmaceutical products. See Note 1 to our consolidated financial statements included in this Annual Report on Form 10-K. For financial information regarding our business, see “Item 7. Management’s Discussion and Analysis of Financial Condition and Results of Operations” and our financial statements and related notes.

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 public may read or copy any materials we file with the SEC at the SEC's Public Reference Room at 100 F Street NE, Washington, D.C. 20549. The public may obtain information on the operation of the Public Reference Room by calling the SEC at 1-800-SEC-0330. 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, 2017, 2016 and 2015 was \$17.9 million, \$(40.0) million and \$(47.3) million, respectively. As of December 31, 2017, we had an accumulated deficit of \$289.2 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 by the applicable regulatory authorities on the time schedule we have planned, or at all.

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, only six 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 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 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 example, in August 2013, our former collaboration partner, Glaxo Group Limited, or GSK, an affiliate of GlaxoSmithKline, reported the first of four Phase III studies, the SHIELD-1 study, investigating vécirnon in patients with moderate-to-severe Crohn's disease, did not achieve the primary endpoint of improvement in clinical response and the key secondary endpoint of clinical remission, and GSK subsequently reverted to us all

rights to vercirnon and its two identified back-up compounds. In addition, in November 2013, GSK reverted to us all rights to CCX354 and its two identified back-up compounds, and GSK declined its last option to license avacopan under our agreement with GSK.

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 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 or other foreign regulatory agencies after submission of our results;
- an unfavorable FDA inspection of our contract manufacturers of API or drug product; and
- inspection of the clinical trial 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 CCX140 and avacopan. As of the date of this Annual Report on Form 10-K, six 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.

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 commercial 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, 2017, we had approximately \$135.2 million in cash, cash equivalents and investments, excluding the \$50.0 million milestone payment in connection with the Avacopan Agreement and the \$10.0 million upfront commitment in connection with the February 2017 amendment to the Avacopan Agreement. 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 12, 2018. 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, 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 \$5.0 million at December 31, 2017.

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 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. 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, aHUS 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, aHUS 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 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. 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 currently have no 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 currently have no sales, marketing or distribution capabilities or experience. If our products are approved for sale, we intend to rely on third parties to assist us in the marketing and distribution of our products. To the extent we rely on third parties 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, 2017, we had 66 full-time employees. We will need to continue to expand our 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:

- 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, aHUS, FSGS, DN 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 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 recently enacted U.S. tax legislation, although the treatment of tax losses generated before December 31, 2017 has generally not changed, tax losses generated in calendar year 2018 and beyond may be used to offset only 80% of taxable income. 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.

Recently enacted U.S. tax legislation has 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 are 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. While our analysis and interpretation of this legislation is ongoing, based on our current

evaluation, we have reflected a \$36.0 million write-down of our federal deferred income tax assets (including the value of our net operating loss carryforwards and our tax credit carryforwards) due to the reduction of the U.S. corporate income tax rate. This amount may be subject to further adjustment in subsequent periods throughout 2018 in accordance with subsequent interpretive guidance issued by the SEC or the IRS. 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 the Company 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 received 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

Patent litigation could absorb significant management time and financial resources, and, if we do not prevail, could materially and adversely impact our financial position and results of operations.

Intellectual property litigation and patent litigation in particular, is expensive, complex and lengthy and its outcome is difficult to predict. A court could enter orders that temporarily, preliminarily or permanently enjoin us or our strategic partners from using, selling, offering to sell or importing our current or future drug candidates or could enter an order mandating that we undertake certain remedial activities. We may also be subject to negative publicity due to litigation. Pending or future patent litigation against us or any strategic partners may force us or any strategic partners to stop or delay developing, manufacturing or selling potential drug candidates that are claimed to infringe a third party's intellectual property, unless that party grants us or any strategic partners rights to use its intellectual property. U.S. patents are entitled to a presumption of validity and the burden of proving invalidity would be heavily weighted against us. Such decisions on patent validity often favor the patent owner because of the presumption of validity.

The cost to us of any patent litigation or other proceedings, such as interference proceedings, which are meant to determine who first invented any of the claims covered by the patent even if resolved in our favor, 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. Patent 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.

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 824 issued or allowed patents and approximately 197 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. There are approximately 87 issued or allowed patents and 41 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 77 issued or allowed patents and 26 patents pending worldwide relating to their chemical composition or use thereof. With respect to the CCR1 and CCR9 chemokine receptors, we have approximately 468 issued or allowed patents and 120 patents pending worldwide relating to their chemical composition or use thereof. We have approximately 143 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs. We have approximately 49 issued patents and 10 pending applications relating to other small molecule compounds and approximately 140 issued patents relating to our novel biological discoveries and our proprietary screening and drug development technologies. 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. 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.

In addition, there are numerous recent changes to the patent laws and proposed changes to the rules of the U.S. Patent and Trademark Office, or USPTO, which may have a significant impact on our ability to protect our technology and enforce our intellectual property rights. For example, in September, 2011, the President signed the America Invents Act, which codifies several significant changes to the U.S. patent laws, including, among

other things, changing from a “first to invent” to a “first inventor to file” system, limiting where a patentee may file a patent suit, requiring the apportionment of patent damages, replacing interference proceedings with derivation actions, and creating a post-grant opposition process to challenge patents after they have issued. The effects of these changes are currently unclear as the USPTO must still implement various regulations, the courts have yet to address any of these provisions, and the applicability of the act and new regulations on specific patents discussed herein have not been determined and would need to be reviewed.

We rely upon unpatented trade secrets, unpatented know-how and continuing technological innovation to develop and maintain our competitive position, which we seek to protect, in part, by confidentiality agreements with our employees and our collaborators and consultants. We also have agreements with our employees and selected consultants that obligate them to assign their inventions to us. However, it is possible that technology relevant to our business will be independently developed by a person that is not a party to such an agreement. Furthermore, if the employees and consultants that are parties to these agreements breach or violate the terms of these agreements, we may not have adequate remedies for any such breach or violation, and we could lose our trade secrets through such breaches or violations. Further, our trade secrets could otherwise become known or be independently discovered by our competitors. In addition, intellectual property laws in foreign countries may not protect our intellectual property to the same extent as the laws of the United States.

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.

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.

In addition to infringement claims against 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 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.

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 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 (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 Food and Drug Administration Safety and Innovation Act of 2012 required the FDA to issue guidance on permissible forms of internet and social media promotion of regulated medical products, and the FDA issued a number of draft guidance documents relating to social media and may soon specify new restrictions on this form of product promotion. In addition, in December 2016, 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, but its ultimate implementation is unclear. 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 Trump 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.

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 European Union 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 European Union'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 January 2018, the EMA validated our CMA application for avacopan in the treatment of patients with AAV. With the validation of a CMA application the EMA simply confirms that the application is complete and that the CHMP can initiate its assessment, which may or may not conclude with a positive opinion recommending the CMA of the product, and we may not be able to meet all conditions imposed on us by the EMA in the context of a CMA approval for avacopan.

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 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; 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. Recently, the Tax Cuts and 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, the 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, 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.

Heightened Congressional scrutiny on the adequacy of the FDA's drug approval process and the agency's efforts to assure the safety of marketed drugs resulted in the enactment of legislation addressing drug safety issues, the FDA Amendments Act of 2007, or FDAAA. This legislation provided the FDA with expanded authority over drug products after approval, including the authority to impose the requirement for a REMS to assure the safe use of the drug, either as a condition for product approval or after a product is approved on the basis of new safety information. A REMS could include medication guides, physician communication plans, or elements to assure safe use, such as restricted distribution methods, patient registries and other risk minimization tools. Any of these limitations on approval or marketing could restrict the commercial promotion, distribution, prescription or dispensing of our drug candidates, if approved. The FDA's exercise of this authority under FDAAA has resulted in delays or increased costs during the period of product development, clinical trials and regulatory review and approval, and increased costs to assure compliance with new post-approval regulatory requirements. Given the serious public health risks of high profile adverse safety events with certain drug products, the FDA may require, as a condition of approval, costly risk management programs which may include safety surveillance, restricted distribution and use, patient education, enhanced labeling, special packaging or labeling, expedited reporting of certain adverse events, preapproval of promotional materials and restrictions on direct-to-consumer advertising.

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 \$74,792 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 \$55,910 per violation, not to exceed \$1.68 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 require 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 \$11,052 per failure up to an aggregate of \$165,786 per year (or up to an aggregate of \$1.105 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, 2017, the price per share for our common stock on the Nasdaq Global Select Market ranged from a low sale price of \$5.42 to a high sale price of \$10.80. 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 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 February 28, 2018, our officers and directors, together with holders of 5% or more of our outstanding common stock and their respective affiliates, beneficially owned approximately 65% 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, 2017, we had 48,837,060 shares of common stock outstanding. Of these shares, approximately 36,510,319 are freely tradeable, without restriction, in the public market. In addition, approximately 16,104,533 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. If these 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. 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 \$4.0 million for severance and other benefits and acceleration of vesting of stock awards with an intrinsic value of \$3.4 million as of December 31, 2017 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 2018, we had research coverage by only two 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 European Union 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 two years after the government of the United Kingdom formally initiates a withdrawal process. Nevertheless, the referendum has created significant uncertainty about the future relationship between the United Kingdom and the European Union, including with respect to the laws and regulations that will apply as the United Kingdom determines which European Union laws to replace or replicate in the event of a withdrawal. The referendum has also given rise to calls for the governments of other European Union 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 has been traded on the Nasdaq Global Select Market since February 8, 2012 under the symbol “CCXI.” Prior to such time, there was no public market for our common stock. The following table sets forth the high and low sales prices per share of our common stock on the Nasdaq Global Select Market for the quarterly periods indicated.

	Sales Price of Common Shares	
	High	Low
<i>Fiscal 2017</i>		
First Quarter	\$ 8.58	\$ 6.13
Second Quarter	\$ 9.47	\$ 6.51
Third Quarter	\$10.80	\$ 6.41
Fourth Quarter	\$ 8.60	\$ 5.42
<i>Fiscal 2016</i>		
First Quarter	\$ 7.96	\$ 2.16
Second Quarter	\$ 5.69	\$ 1.92
Third Quarter	\$ 6.20	\$ 3.95
Fourth Quarter	\$ 9.10	\$ 5.19

Holder of Common Stock

As of February 28, 2018, there were approximately 44 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, 2017.

<u>Plan Category</u>	<u>Shares Issuable Upon Exercise of Outstanding Options, Warrants and Rights⁽²⁾</u>	<u>Weighted Average Exercise Price of Outstanding Options, Warrants and Rights⁽³⁾</u>	<u>Number of Securities Available for Future Issuance⁽⁴⁾</u>
Equity compensation plans approved by security holders: ⁽¹⁾	10,626,584	\$7.68	2,362,215
Equity compensation plans not approved by security holders:	—	—	—
Total	<u>10,626,584</u>	<u>\$7.68</u>	<u>2,362,215</u>

- (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 10,203,571 shares subject to outstanding stock option and 423,013 shares subject to outstanding restricted stock unit as of December 31, 2017.
- (3) Calculated exclusive of outstanding restricted stock unit awards.
- (4) Of these shares, 2,028,880 shares were available for stock option awards, restricted stock units and restricted stock awards, and 333,335 were available for the ESPP, in each case as of December 31, 2017.

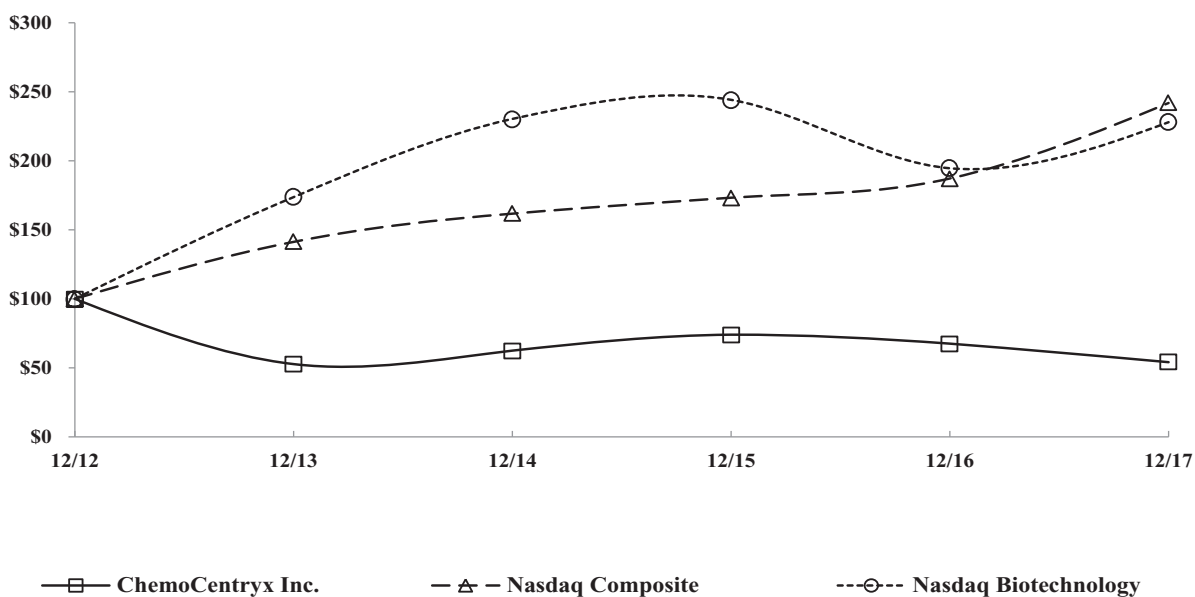
Performance Graph

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, 2012 (the date our common stock commenced trading on the Nasdaq Global Select Market) through December 31, 2017 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.

COMPARISON OF 5 YEAR CUMULATIVE TOTAL RETURN*

Among ChemoCentryx Inc., the Nasdaq Composite Index and the Nasdaq Biotechnology Index



*\$100 invested on 12/31/12 in stock or index, including reinvestment of dividends.
Fiscal year ending December 31.

	<u>12/12</u>	<u>12/13</u>	<u>12/14</u>	<u>12/15</u>	<u>12/16</u>	<u>12/17</u>
ChemoCentryx Inc.	100.00	52.93	62.43	74.04	67.64	54.39
Nasdaq Composite	100.00	141.63	162.09	173.33	187.19	242.29
Nasdaq Biotechnology	100.00	174.05	230.33	244.29	194.95	228.29

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,				
	2017	2016	2015	2014	2013
	(in thousands, except share and per share data)				
Consolidated Statements of Operations Data:					
Revenue:					
Collaboration and license revenue	\$ 82,497	\$ 11,435	\$ —	\$ —	\$ 6,060
Grant revenue	—	500	—	—	—
Total revenue	82,497	11,935	—	—	6,060
Operating expenses:					
Research and development	49,495	37,945	33,183	33,815	33,541
General and administrative	16,509	14,710	14,506	13,584	11,634
Total operating expenses	66,004	52,655	47,689	47,399	45,175
Income (loss) from operations	16,493	(40,720)	(47,689)	(47,399)	(39,115)
Interest income	1,370	757	384	494	501
Interest expense	(4)	—	—	(24)	(59)
Net income (loss)	\$ 17,859	\$ (39,963)	\$ (47,305)	\$ (46,929)	\$ (38,673)
Net income (loss) per share, basic ⁽¹⁾	\$ 0.37	\$ (0.86)	\$ (1.08)	\$ (1.08)	\$ (0.95)
Net income (loss) per share, diluted ⁽¹⁾	\$ 0.36	\$ (0.86)	\$ (1.08)	\$ (1.08)	\$ (0.95)
Shares used to compute net income (loss) per share, basic	48,412,531	46,431,501	43,889,677	43,275,276	40,916,138
Shares used to compute net income (loss) per share, diluted	49,615,406	46,431,501	43,889,677	43,275,276	40,916,138

- (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,				
	2017	2016	2015	2014	2013
	(in thousands)				
Consolidated Balance Sheets Data:					
Cash, cash equivalents and investments	\$ 135,220	\$ 123,761	\$ 76,289	\$ 114,620	\$ 149,874
Accounts receivable ⁽²⁾	51,090	30,205	—	—	393
Working capital	146,893	110,356	66,541	66,139	127,430
Total assets	189,328	155,872	78,155	116,981	152,422
Long-term debt, net	4,676	—	—	—	16
Accumulated deficit	(289,200)	(307,059)	(267,096)	(219,791)	(172,862)
Total stockholders’ equity	79,267	49,889	72,507	108,606	145,308

- (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. Accounts receivable at December 31, 2017 included \$1.1 million of unbilled receivable related to development costs to be reimbursed by Vifor under the CCX140 Agreement. 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 10, Collaboration and License Agreements” 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 selectively blocks a specific chemoattractant receptor, leaving the rest of the immune system intact. Our drug candidates are small molecules, which are orally administered, offering 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 commitments and milestones to support the clinical development of our leading two 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 kidney disease, particularly 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) — Complement Inhibition 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; (ii) complement 3 glomerulopathy, or C3G, a debilitating disease that can lead to kidney failure; and (iii) atypical hemolytic uremic syndrome, or aHUS, a rare, life threatening disease.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV, C3G and aHUS 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 enrolling approximately 300 patients at approximately 200 sites in the United States, Canada, Europe, Australia and New Zealand. We expect to complete patient enrollment of

the Phase III ADVOCATE trial in mid-2018. We recently launched a registration-supporting clinical trial to study avacopan for the treatment of patients with C3G and plan to initiate clinical studies for the treatment of patients with hidradenitis suppurativa, or HS, in 2018. Meanwhile, we are actively dosing aHUS patients under compassionate use protocols as we explore appropriate doses and dosing regimen for that indication.

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.

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 highest proteinuric patients. Based on the safety and efficacy data related to reduction in proteinuria observed in the Phase II trial in DN, we launched our clinical development program of CCX140 for the treatment of patients with FSGS, 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 commercial 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 this second agreement, we again retained all rights to the United States and China and we granted Vifor worldwide 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.

We have secured \$205 million in upfront cash payments and commitments and milestones pursuant to our agreements with Vifor and are eligible for additional substantial milestone payments. Through our alliance, we maintain the commercial rights to avacopan and CCX140 in the United States and China, and also retain control of the clinical development programs for orphan renal disease. Vifor gained the commercial 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 the science has led us to focus 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 other 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 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, 2017, we had an accumulated deficit of \$289.2 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

Appointment of William J. Fairey Jr. as Executive Vice President and Chief Operating Officer

In January 2018, we announced the appointment of William J. Fairey, Jr. as Executive Vice President and Chief Operating Officer. Mr. Fairey will lead our commercial strategy, including the development and execution of our commercialization plans for our orphan disease drug candidates avacopan and CCX140, as well as oversee other key operational functions.

Avacopan Conditional Marketing Authorization Application Accepted for Review by the European Medicines Agency

In December 2017, our CMA application for avacopan in the treatment of patients with AAV was accepted for review by the EMA's CHMP. Under the terms of our kidney health alliance with Vifor, this validation of the avacopan CMA application by the EMA resulted in a \$50.0 million milestone.

CCX872 Positive Overall Survival Results for Second CCR2 Inhibitor in the Treatment of Locally Advanced/Metastatic Pancreatic Cancer

In January 2018, we announced positive overall survival, or OS, results from an ongoing Phase Ib clinical trial of CCX872, our second CCR2 inhibitor, in the treatment of locally advanced/metastatic pancreatic cancer. The study reported OS of 29% at 18 months with CCX872 and FOLFIRINOX combination therapy in all randomized patients treated. This result compares favorably with previously published data of OS of 18.6% at 18 months using FOLFIRINOX regimen alone to treat pancreatic cancer patients with metastatic disease.

\$50 Million Loan and Security Agreement with Hercules Capital, Inc.

In December 2017, we entered into a \$50 million growth capital financing agreement with Hercules Capital, Inc., or Hercules. The borrowings under this facility are available in three tranches through June 15, 2019, subject to certain conditions. Advances under the financing agreement 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%. The financing agreement has a 24-month interest-only period from initial funding, which is extendable to 30 months upon satisfaction of certain milestones and matures in 48 months.

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

We enter into corporate collaboration and license agreements under which we may obtain upfront license fees, research and development funding and, contingent milestone and royalty payments. Our deliverables under these arrangements may include intellectual property rights, distribution rights, delivery of manufactured product, participation on joint steering committees and/or research and development services. In order to account for the multiple-element arrangements, we identify the deliverables included within the arrangement and evaluate whether the delivered elements under these arrangements have value to our collaboration partner on a stand-alone basis and represent separate units of accounting. Analyzing the arrangement to identify deliverables requires the use of judgment, and each deliverable may be an obligation to deliver future goods or services, a right or license to use an asset, or another performance obligation. If we determine that multiple deliverables exist, the consideration is allocated to one or more units of accounting based upon the best estimate of the selling price of each deliverable. The selling price used for each deliverable will be based on vendor-specific objective evidence, if available, third-party evidence if vendor-specific objective evidence is not available, or estimated selling price if neither vendor-specific or third-party evidence is available. A delivered item or items that do not qualify as a separate unit of accounting within the arrangement shall be combined with the other applicable undelivered items within the arrangement. The allocation of arrangement consideration and the recognition of revenue then shall be determined for those combined deliverables as a single unit of accounting. For a combined unit of accounting, non-refundable upfront fees are recognized in a manner consistent with the final deliverable, which has generally been ratably over the estimated period of continued involvement. We periodically review the basis for our estimates, and we may change the estimates if circumstances change. These changes can significantly change the timing of revenue recognized. Amounts received in advance of performance are recorded as deferred revenue. Revenues derived from funding of development costs are recognized when the related costs are incurred and when collectability is reasonably assured. Revenues from upfront fees and development services are classified as collaboration and license revenue in our Consolidated Statements of Operations.

We consider sales-based contingent payments to be royalty revenue which is generally recognized at the date the contingency is achieved. Research and development funding related to collaborative research and development efforts is recognized as revenue as the related services are performed or delivered, in accordance with contract terms.

For certain contingent payments under collaboration and license arrangements, we recognize revenue using the milestone method. Under the milestone method, a payment that is contingent upon the achievement of a substantive milestone is recognized in its entirety in the period in which the milestone is achieved. A milestone is an event: (i) that can be achieved based in whole or in part on either our performance or on the occurrence of a specific outcome resulting from our performance, (ii) for which there is substantive uncertainty at the date the arrangement is entered into that the event will be achieved and (iii) that would result in additional payments being due to us. The determination that a milestone is substantive requires estimation and judgment and is made at the inception of the arrangement. Milestones are considered substantive when the consideration earned from the achievement of the milestone is: (i) commensurate with either our performance to achieve the milestone or the enhancement of value of the item delivered as a result of a specific outcome resulting from our performance

to achieve the milestone, (ii) related solely to past performance and (iii) reasonable relative to all deliverables and payment terms in the arrangement. In making the determination as to whether a milestone is substantive or not, we consider all facts and circumstances relevant to the arrangement, including factors such as the scientific, regulatory, commercial and other risks that must be overcome to achieve the respective milestone, the level of effort and investment required to achieve the respective milestone and whether any portion of the milestone consideration is related to future performance or deliverables. Contingency and milestones payments, when recognized as revenue, are classified as collaboration and license revenues in our Consolidated Statements of Operations.

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 \$8.7 million, \$8.5 million and \$9.0 million for the years ended December 31, 2017, 2016, and 2015, respectively. At December 31, 2017 and 2016, we had \$8.2 million and \$9.6 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 and 2.4 years, respectively. 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 year ended December 31, 2017, our revenues were derived from (i) the recognition of the milestone payment related to the Avacopan Agreement;

(ii) the recognition of the upfront payments related to the Avacopan Agreement, Avacopan Amendment and CCX140 Agreement and (iii) collaboration revenue under the CCX140 Agreement. 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. No revenue was recorded in 2015.

Total revenue were as follows (in thousands):

	Year Ended December 31,		
	2017	2016	2015
Collaboration and license revenue	\$82,497	\$11,435	\$—
Grant revenue	—	500	—
Total revenue	<u>\$82,497</u>	<u>\$11,935</u>	<u>\$—</u>
Dollar increase	\$70,562	\$11,935	\$—
Percentage increase	591%	100%	0%

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,		
	2017	2016	2015
Research and development expenses	\$49,495	\$37,945	\$33,183
Dollar increase	\$11,550	\$ 4,762	
Percentage increase	30%	14%	

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 increase in research and development expenses from 2015 to 2016 was primarily attributable to higher Phase I and Phase III clinical development expenses in 2016 partially offset by a decrease in Phase II

development expenses. The increase in Phase III development expense was due to the initiation of Phase III development program for avacopan in patients with AAV in 2016. The increase in Phase I development expense was driven by the completion of ancillary Phase I clinical trials for avacopan in support of end of Phase II meetings with regulatory agencies, as well as higher expenses associated with CCX872 for our ongoing Phase I clinical trial in patients with advanced pancreatic cancer. The decrease in Phase II development expense was due to the completion of the CLEAR and CLASSIC clinical trials for avacopan for the treatment of AAV in 2016.

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

	Year Ended December 31,		
	2017	2016	2015
Phase I	\$ 785	\$ 5,959	\$ 3,686
Phase II	8,854	10,866	16,151
Phase III	26,198	8,019	—
Research and drug discovery	13,658	13,101	13,346
Total R&D	<u>\$49,495</u>	<u>\$37,945</u>	<u>\$33,183</u>

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,		
	2017	2016	2015
General and administrative expenses	\$16,509	\$14,710	\$14,506
Dollar increase	\$ 1,799	\$ 204	
Percentage increase	12%	1%	

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 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. The increase from 2015 to 2016 was primarily due to increase in professional fees relating to our business development efforts.

We expect that general and administrative expenses will increase in the future as we expand our operating activities and incur additional costs associated with being a public company. These public company related increases will likely include, but not be limited to, investor and public relations expenses and legal and accounting related fees. We also expect that general and administrative expenses will increase as we plan to commercialize our orphan disease drug candidates such as avacopan in the United States.

Other income (expense), net

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

	Year Ended December 31,		
	2017	2016	2015
Interest income	\$1,370	\$757	\$384
Interest expense	(4)	—	—
Total other income, net	<u>\$1,366</u>	<u>\$757</u>	<u>\$384</u>
Dollar increase	\$ 609	\$373	
Percentage increase	80%	97%	

The increase in total other income (expense), 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.

The increase in total other income from 2015 to 2016 was primarily due to higher cash and investment balances in 2016 due to the \$85.0 million upfront payment received from Vifor in connection with the Avacopan Agreement.

We expect that interest expense will increase in the future due to our advances from the loan and security agreement, or Credit Agreement, with Hercules.

Liquidity and Capital Resources

As of December 31, 2017, we had approximately \$135.2 million in cash, cash equivalents and investments. Such amounts exclude the \$50.0 million milestone payment in connection with the Avacopan Agreement and \$10.0 million in remaining upfront commitment in connection with the February 2017 Avacopan Amendment, which is due on the first anniversary of the agreement. The following table shows a summary of our cash flows for each of the three years ended December 31, 2017, 2016, and 2015 (in thousands):

	<u>2017</u>	<u>2016</u>	<u>2015</u>
Cash provided by (used in)			
Operating activities	\$ 4,878	\$ 39,145	\$(39,327)
Investing activities	\$15,602	\$(48,764)	\$ 33,884
Financing activities	\$ 7,516	\$ 8,820	\$ 2,191

Operating activities. Net cash provided by operating activities was \$4.9 million for the year ended December 31, 2017, compared to \$39.2 million provided by operating activities for the same period in 2016. This change was primarily due to changes in working capital items, which was partially offset by an increase in net income. 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. Net cash provided by operating activities was \$39.2 million for the year ended December 31, 2016, compared to cash used of \$39.3 million for the same period in 2015. This change was primarily due to changes in working capital items in 2016 primarily due to \$78.0 million in connection with the Avacopan Agreement, as well as a lower net loss.

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 2016 represents the investment of funds received under the Avacopan Agreement.

Financing activities. Net cash provided by financing activities was \$7.5 million for the year ended December 31, 2017, which included net proceeds of \$4.7 million received under our credit facility (discussed below), partially offset by cash used of \$0.4 million (the value of withheld shares) for tendered ChemoCentryx, Inc. common stock to satisfy employee tax withholding requirements upon vesting of restricted stock units. 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 was \$2.2 million for the year ended December 31, 2015. Net cash provided by financing activities for the years presented included proceeds from the exercise of stock options and from employee purchases of stock under our 2012 Employee Stock Purchase Plan.

In December 2017, we entered into a Credit Agreement with Hercules, under which we may borrow up to \$50.0 million in three tranches, subject to certain terms and conditions, which we refer to as the Credit Facility. Under the first tranche, we may borrow up to \$15.0 million, of which we advanced \$5.0 million in December 2017. Upon satisfaction of certain milestones, the second tranche is available under the Credit Facility, which would allow us to borrow an additional amount up to \$10.0 million through December 15, 2018. The third tranche, which would allow us to borrow an additional \$25.0 million, will be available upon 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, 2017. See "Note 7. 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, 2017, we had approximately \$135.2 million in cash, cash equivalents and investments, excluding the \$50.0 million milestone payment in connection with the Avacopan Agreement and \$10.0 million in remaining upfront commitment in connection with the February 2017 Avacopan Amendment. 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 12, 2018. 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, 2017 (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⁾	\$5,000	\$—	\$2,394	\$2,606	\$—
Operating lease ⁽²⁾	\$2,753	\$937	\$1,816	\$ —	\$—
Total contractual obligations	<u>\$7,753</u>	<u>\$937</u>	<u>\$4,210</u>	<u>\$2,606</u>	<u>\$—</u>

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

(2) 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 in market rates would have any significant impact on the realized value of our marketable securities.

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-31 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, 2017, 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, 2017, 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, 2017 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, 2017.

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, 2017, 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, 2017, 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 2018 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, 2017, 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:

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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.

ChemoCentryx, Inc.

**Consolidated Financial Statements
As of December 31, 2017 and 2016
and for each of the three years in the period ended December 31, 2017**

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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, 2017 and 2016, 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, 2017, 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, 2017 and 2016, and the results of its operations and its cash flows for each of the three years in the period ended December 31, 2017, 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, 2017, 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 12, 2018 expressed an unqualified opinion thereon.

Basis for Opinion

These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on these 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 12, 2018

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, 2017, 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, 2017, 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, 2017 and 2016, 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, 2017, and the related notes of the Company and our report dated March 12, 2018 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 12, 2018

CHEMOCENTRYX, INC.
Consolidated Balance Sheets
(In thousands, except share data)

	December 31,	
	2017	2016
Assets		
Current assets:		
Cash and cash equivalents	\$ 40,020	\$ 12,024
Short-term investments	87,271	105,740
Accounts receivable	51,090	30,205
Prepaid expenses and other current assets	1,449	722
Total current assets	179,830	148,691
Property and equipment, net	1,210	905
Long-term investments	7,929	5,997
Other assets	359	279
Total assets	\$ 189,328	\$ 155,872
Liabilities and Stockholders' Equity		
Current liabilities:		
Accounts payable	\$ 1,400	\$ 671
Accrued liabilities	8,575	8,645
Deferred revenue	22,962	29,019
Total current liabilities	32,937	38,335
Long-term debt, net	4,676	—
Noncurrent deferred revenue	72,197	67,547
Other non-current liabilities	251	101
Total liabilities	110,061	105,983
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; 48,837,060 shares and 48,057,920 shares issued and outstanding at December 31, 2017 and December 31, 2016, respectively	49	48
Additional paid-in capital	368,553	356,966
Note receivable	(16)	(16)
Accumulated other comprehensive loss	(119)	(50)
Accumulated deficit	(289,200)	(307,059)
Total stockholders' equity	79,267	49,889
Total liabilities and stockholders' equity	\$ 189,328	\$ 155,872

See accompanying notes.

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

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Revenue:			
Collaboration and license revenue	\$82,497	\$ 11,435	\$ —
Grant revenue	—	500	—
Total revenue	<u>82,497</u>	<u>11,935</u>	<u>—</u>
Operating expenses:			
Research and development	49,495	37,945	33,183
General and administrative	<u>16,509</u>	<u>14,710</u>	<u>14,506</u>
Total operating expenses	<u>66,004</u>	<u>52,655</u>	<u>47,689</u>
Income (loss) from operations	16,493	(40,720)	(47,689)
Other income (expense):			
Interest income	1,370	757	384
Interest expense	<u>(4)</u>	<u>—</u>	<u>—</u>
Total other income, net	<u>1,366</u>	<u>757</u>	<u>384</u>
Net income (loss)	<u>\$17,859</u>	<u>\$(39,963)</u>	<u>\$(47,305)</u>
Net income (loss) per common share			
Basic	<u>\$ 0.37</u>	<u>\$ (0.86)</u>	<u>\$ (1.08)</u>
Diluted	<u>\$ 0.36</u>	<u>\$ (0.86)</u>	<u>\$ (1.08)</u>
Shares used to compute net income (loss) per common share			
Basic	<u>48,413</u>	<u>46,432</u>	<u>43,890</u>
Diluted	<u>49,615</u>	<u>46,432</u>	<u>43,890</u>

See accompanying notes.

CHEMOCENTRYX, INC.
Consolidated Statements of Comprehensive Income (Loss)
(In thousands)

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Net income (loss)	\$17,859	\$(39,963)	\$(47,305)
Unrealized (loss) gain on available-for-sale securities	<u>(69)</u>	<u>(10)</u>	<u>30</u>
Comprehensive income (loss)	<u>\$17,790</u>	<u>\$(39,973)</u>	<u>\$(47,275)</u>

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, 2014	43,446,096	\$ 43	\$328,440	\$ (16)	\$ (70)	\$(219,791)	\$108,606
Net loss	—	—	—	—	—	(47,305)	(47,305)
Unrealized gain / (loss) on investments	—	—	—	—	30	—	30
Issuance of common stock under equity incentive and employee stock purchase plans	739,410	1	2,190	—	—	—	2,191
Employee stock-based compensation	—	—	8,860	—	—	—	8,860
Compensation expense related to options granted to consultants	—	—	125	—	—	—	125
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	<u>48,837,060</u>	<u>\$ 49</u>	<u>\$368,553</u>	<u>\$ (16)</u>	<u>\$(119)</u>	<u>\$(289,200)</u>	<u>\$ 79,267</u>

See accompanying notes.

CHEMOCENTRYX, INC.
Consolidated Statements of Cash Flows
(In thousands)

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Operating activities			
Net income (loss)	\$ 17,859	\$ (39,963)	\$(47,305)
Adjustments to reconcile net income (loss) to net cash provided by (used in) operating activities:			
Depreciation of property and equipment	418	348	477
Stock-based compensation	8,748	8,535	8,985
Noncash interest expense, net	143	179	1,007
Changes in assets and liabilities:			
Accounts receivable	(20,885)	(30,205)	—
Prepays and other current assets	(727)	35	236
Other assets	(80)	(119)	—
Accounts payable	729	(4)	(73)
Other liabilities	80	3,773	(2,654)
Deferred revenue	(1,407)	96,566	—
Net cash provided by (used in) operating activities	<u>4,878</u>	<u>39,145</u>	<u>(39,327)</u>
Investing activities			
Purchases of property and equipment, net	(723)	(304)	(218)
Purchases of investments	(133,845)	(136,234)	(24,372)
Sales of investments	—	—	4,051
Maturities of investments	150,170	87,774	54,423
Net cash provided by (used in) investing activities	<u>15,602</u>	<u>(48,764)</u>	<u>33,884</u>
Financing activities			
Proceeds from issuance of common stock	—	7,000	—
Proceeds from exercise of stock options and employee stock purchase plan	3,269	1,820	2,191
Employees' tax withheld and paid for restricted stock units	(429)	—	—
Borrowings under credit facility agreement, net of issuance costs	4,676	—	—
Net cash provided by financing activities	<u>7,516</u>	<u>8,820</u>	<u>2,191</u>
Net increase (decrease) in cash and cash equivalents	27,996	(799)	(3,252)
Cash and cash equivalents at beginning of period	12,024	12,823	16,075
Cash and cash equivalents at end of period	<u>\$ 40,020</u>	<u>\$ 12,024</u>	<u>\$ 12,823</u>

See accompanying notes.

CHEMOCENTRYX, INC.
Notes to Consolidated Financial Statements
December 31, 2017

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 subsidiary, ChemoCentryx Limited. The operations of 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 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.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

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, 2017 and 2016, 100% and 96%, respectively, of the Company's total revenue were derived from the Company's collaboration with Vifor (International) Ltd., and/or its affiliates, or collectively, Vifor. The Company did not generate any revenue in 2015. 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,	
	2017	2016
Vifor ⁽¹⁾	\$51,090	\$30,000
U.S. Food and Drug Administration	—	205
	<u>\$51,090</u>	<u>\$30,205</u>

- (1) As of December 31, 2017, accounts receivable excluded the \$10.0 million cash commitments due from Vifor in February 2018 in connection with the Avacopan Amendment. Accounts receivable at December 31, 2017 included \$1.1 million of unbilled receivable related to development costs to be reimbursed by Vifor under the CCX140 Agreement. As of December 31, 2016, accounts receivable excluded the \$20.0 million cash commitment due from Vifor in December 2017 in connection with the CCX140 Agreement. See "Note 10. Collaboration and License Agreements" 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

The Company enters into corporate collaborations under which it may obtain upfront license fees, research and development funding and contingent milestones and royalty payments. The Company's deliverables under

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

these arrangements may include intellectual property rights, distribution rights, delivery of manufactured product, participation on joint steering committees and/or research and development services. In order to account for the multiple-element arrangements, the Company identifies the deliverables included within the arrangement and evaluate whether the delivered elements under these arrangements have value to its collaboration partner on a stand-alone basis and represent separate units of accounting. If the Company determines that multiple deliverables exist, the consideration is allocated to one or more units of accounting based upon the best estimate of the selling price of each deliverable. The selling price used for each deliverable will be based on vendor-specific objective evidence, if available, third-party evidence if vendor-specific objective evidence is not available, or estimated selling price if neither vendor-specific nor third-party evidence is available. A delivered item or items that do not qualify as a separate unit of accounting within the arrangement shall be combined with the other applicable undelivered items within the arrangement. The allocation of arrangement consideration and the recognition of revenue then shall be determined for those combined deliverables as a single unit of accounting. For a combined unit of accounting, non-refundable upfront fees are recognized in a manner consistent with the final deliverable, which has generally been ratably over the period of the performance obligation. Amounts received in advance of performance are recorded as deferred revenue. Revenues derived from funding of development costs are recognized when the related costs are incurred and when collectability is reasonably assured. Revenues from upfront fees and development services are classified as collaboration and license revenue in the Consolidated Statements of Operations.

The Company considers sales-based contingent payments to be royalty revenue which is generally recognized at the date the contingency is achieved. Research and development funding related to collaborative research and development efforts is recognized as revenue as the related services are performed or delivered, in accordance with contract terms.

For certain contingent payments under collaboration and license arrangements, the Company recognizes revenue using the milestone method. Under the milestone method a payment that is contingent upon the achievement of a substantive milestone is recognized in its entirety in the period in which the milestone is achieved. A milestone is an event: (i) that can be achieved based in whole or in part on either the Company's performance or on the occurrence of a specific outcome resulting from the Company's performance, (ii) for which there is substantive uncertainty at the date the arrangement is entered into that the event will be achieved and (iii) that would result in additional payments being due to the Company. Milestones are considered substantive when the consideration earned from the achievement of the milestone is: (i) commensurate with either the Company's performance to achieve the milestone or the enhancement of value of the item delivered as a result of a specific outcome resulting from the Company's performance to achieve the milestone, (ii) related solely to past performance and (iii) reasonable relative to all deliverables and payment terms in the arrangement. In making the determination as to whether a milestone is substantive or not, the Company considers all facts and circumstances relevant to the arrangement, including factors such as the scientific, regulatory, commercial and other risks that must be overcome to achieve the respective milestone, the level of effort and investment required to achieve the respective milestone and whether any portion of the milestone consideration is related to future performance or deliverables. Contingency and milestones payments, when recognized as revenue, are classified as collaboration and license revenues in the Consolidated Statements of Operations.

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 income (loss) consists of unrealized gains and losses on the Company's available-for-sale securities. For the year ended December 31, 2015, amounts reclassified from accumulated other income to net loss for unrealized gains (losses) on available-for-sale securities were not significant, and were recorded as part of other income (expense), net in the Consolidated Statements of Operations. For the years ended December 31, 2017 and 2016, there were no sales of investments, and therefore there were no reclassifications.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

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 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,		
	2017	2016	2015
	(in thousands, except per share data)		
Numerator:			
Net income (loss)	\$17,859	\$(39,963)	\$(47,305)
Denominator:			
Weighted average shares outstanding - basic	48,413	46,432	43,890
Dilutive stock options, RSUs and RSAs	1,202	—	—
Weighted average shares outstanding - diluted	49,615	46,432	43,890
Net income (loss) per common share			
Basic	\$ 0.37	\$ (0.86)	\$ (1.08)
Diluted	\$ 0.36	\$ (0.86)	\$ (1.08)

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,		
	2017	2016	2015
Options to purchase common stock, including purchases from contributions to ESPP	6,320,038	9,358,389	7,861,953
Restricted stock units	—	440,344	67,481
Restricted stock awards	—	31,306	—
Warrants to purchase common stock	150,000	150,000	150,000
	6,470,038	9,980,039	8,079,434

Recent Accounting Pronouncements

In May 2014, the Financial Accounting Standard Board (FASB) issued Accounting Standards Update (ASU) No. 2014-09, which supersedes the revenue recognition requirements in ASC Topic 605, Revenue Recognition, and creates a new Topic 606, Revenue from Contracts with Customers. The new standard's core principle is that a reporting entity will recognize revenue when it transfers promised goods or services to customers in an amount that reflects the consideration to which the entity expects to be entitled in exchange for those goods or services. On July 9, 2015, the FASB voted to delay the effective date of the new standard by one year. The standard would become effective for the Company beginning in the first quarter of 2018. Early application would be permitted in 2017. Entities would have the option of using either a full retrospective or a modified retrospective approach to adopt this new guidance. In 2016, the FASB updated the guidance for reporting revenue gross versus net to improve the implementation guidance on principal versus agent considerations, and for identifying performance obligations and the accounting of intellectual property licenses. In addition, the FASB introduced practical expedients and made narrow scope improvements to the new accounting guidance.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

During 2016, the Company entered into two license and collaboration agreements. During 2017 and 2016, the Company primarily derived its revenue from license and collaboration agreements. The consideration the Company is eligible to receive under these agreements includes upfront payments, research and development funding, milestone payments, and royalties. The Company assessed each collaboration agreement under the five-step process under the new standard. The Company reviewed the guidance as it relates to the selection of a measure of progress on the complete satisfaction of performance obligation as this may impact the estimation of, and the determination of the timing of revenue recognition. ASC 606 notes the objective when measuring progress is to depict an entity's performance in transferring control of goods or services promised to a customer.

The Company recognized upfront fees straight-line under ASC 605 over the estimated performance period and recognized milestone when earned under the Milestone Method of accounting. Under ASC 606, the Company's upfront fees and milestones will be recognized using an input method to measure its progress toward the completion of its performance obligations. The Company determined that external R&D costs are a reasonable measure of progress to measure the value that will be transferred to Vifor. These factors will yield the material changes in the Company's revenue recognition between ASC 605 and 606.

On January 1, 2018, the Company adopted the accounting standard update using the modified retrospective approach. Management is finalizing the measure of progress calculations related to the Company's two license and collaboration agreements. Given the factors discussed above, the impact upon adoption is anticipated to increase accumulated deficit in a range of \$45 million to \$50 million. The actual, final quantitative effect of the adoption of Topic 606 will be completed in the first quarter of 2018.

Finally, Topic 606 requires more robust disclosures than required by previous guidance, including disclosures related to disaggregation of revenue into appropriate categories, performance obligations, the judgments made in revenue recognition determinations, adjustments to revenue which relate to activities from previous quarters or years, any significant reversals of revenue, and costs to obtain or fulfill contracts. The Company plans to address these disclosure requirements in its Form 10-Q for the three-month period ending March 31, 2018.

In February 2016, the FASB issued ASU No. 2016-12, 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. The new standard will be effective for the Company on January 1, 2019. The Company is currently evaluating the impact of the adoption of this standard on its financial statements. However, the Company expects the adoption of this accounting guidance to result in an increase in lease assets and a corresponding increase in lease liabilities on its balance sheets.

In March 2016, the FASB issued ASU No. 2016-09, Improvements to Employee Share-Based Payment Accounting, which is intended to simplify several aspects of the accounting for share-based payment award transactions, including the income tax consequences, an option to recognize gross stock compensation expense with actual forfeitures recognized as they occur, as well as certain classifications on the statement of cash flows. Under this guidance, on a prospective basis, companies will no longer record excess tax benefits and tax deficiencies from stock option exercises in additional paid-in capital (APIC). Instead, they will record all excess tax benefits and tax deficiencies as income tax expense or benefit in the statement of operations. In addition, the guidance eliminates the requirement that excess tax benefits be realized before companies can recognize them. The ASU requires a cumulative-effect adjustment for previously unrecognized excess tax benefits in opening retained earnings in the annual period of adoption. The Company adopted ASU No. 2016-09 on January 1, 2017. Upon adoption, the Company recognized the excess tax benefit balance of \$2.1 million as of January 1, 2017 as a deferred tax asset with a corresponding increase to the Company's deferred tax asset valuation allowance.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

Additionally, as provided for under this new guidance, the Company elected to continue to estimate forfeitures. The adoption of this aspect of the guidance did not have a material impact on the Company's 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.

3. Cash Equivalents and Investments

The amortized cost and fair value of cash equivalents and investments at December 31, 2017 and 2016 were as follows (in thousands):

	December 31, 2017			
	Amortized Cost	Gross Gains	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	\$—	\$(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

	December 31, 2016			
	Amortized Cost	Gross Gains	Unrealized Losses	Fair Value
Money market fund	\$ 9,746	\$—	\$—	\$ 9,746
U.S. treasury securities	49,693	1	(22)	49,672
Commercial paper	16,183	—	—	16,183
Corporate debt securities	45,911	—	(29)	45,882
Total available-for-sale securities	\$121,533	\$ 1	\$(51)	\$121,483

Classified as:

Cash equivalents				\$ 9,746
Short-term investments				105,740
Long-term investments				5,997
Total available-for-sale securities				\$121,483

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

3. Cash Equivalents and Investments (continued)

Cash equivalents in the tables above exclude cash of \$3.2 million and \$2.3 million as of December 31, 2017 and 2016, respectively. All available-for-sale securities held as of December 31, 2017, 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 available-for-sale securities held as of December 31, 2017 have been in a continuous unrealized loss position for more than 12 months. As of December 31, 2017, 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.

The Company's financial assets and liabilities subject to fair value measurements on a recurring basis and the level of inputs used in such measurements are as follows as of December 31, 2017 and 2016 (in thousands):

Description	December 31, 2017			
	Level 1	Level 2	Level 3	Total
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	<u>\$29,848</u>	<u>\$102,165</u>	<u>\$—</u>	<u>\$132,013</u>
	December 31, 2016			
	Level 1	Level 2	Level 3	Total
Money market fund	\$ 9,746	\$ —	\$—	\$ 9,746
U.S. treasury securities	—	49,672	—	49,672
Commercial paper	—	16,183	—	16,183
Corporate debt securities	—	45,882	—	45,882
Total assets	<u>\$ 9,746</u>	<u>\$111,737</u>	<u>\$—</u>	<u>\$121,483</u>

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

4. Fair Value Measurements (continued)

During the year ended December 31, 2017 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.

5. Property and Equipment

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

	December 31,	
	2017	2016
Lab equipment	\$ 5,897	\$ 5,950
Computer equipment and software	1,688	1,511
Furniture and fixtures	551	528
Tenant improvements	893	866
	9,029	8,855
Less: accumulated depreciation	(7,819)	(7,950)
	\$ 1,210	\$ 905

6. Accrued Liabilities

Accrued liabilities consist of the following (in thousands):

	December 31,	
	2017	2016
Research and development related	\$4,962	\$5,482
Compensation related	2,345	2,460
Consulting and professional services	1,012	421
Other	256	282
	\$8,575	\$8,645

7. Long-term Debt

On December 28, 2017 (the Closing Date), the Company entered into a Loan and Security Agreement (the Credit Agreement) with Hercules Capital, Inc. (Hercules) 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. Under the first tranche, the Company may borrow through June 2018 an amount up to \$15.0 million, of which \$5.0 million was advanced to the Company on the Closing Date. Upon satisfaction of

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

7. Long-term Debt (continued)

certain terms and conditions, the second tranche is available under the Credit Facility, which would allow the Company to borrow an additional amount up to \$10.0 million through December 15, 2018. The third tranche, which would allow the Company to borrow an additional \$25.0 million, would be available upon 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%. For advances under the first tranche, the Company will make interest-only payments through January 1, 2020, extended to July 1, 2020 upon satisfaction of certain milestones, and will then repay the principal balance and interest on the advances in equal monthly installments after the interest-only period and continuing through December 1, 2021. For advances made under the second and third tranches, the Company will make interest only payments in the first 24 months, extended to 30 months upon satisfaction of certain conditions, and will then repay the principal balance and interest on the advances in equal monthly installments after the interest only period with the entire term loan advances repaid 48 months after the advance under the applicable tranche is drawn.

The Company may prepay advances under the Credit Agreement, in whole or in part, at any time, subject to a prepayment charge equal to: (a) 2.0% of amounts so prepaid, if such prepayment occurs during the first year following the Closing Date; (b) 1.5% of the amount so prepaid, if such prepayment occurs during the second year following the Closing Date; and (c) 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 as of December 31, 2017.

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 the greater of (a) 6.25% of the aggregate amount of the advances and (b) 6.25% of the aggregate amount of the advances plus 50% of the unfunded portion of the first tranche. 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 Agreement. 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. Amortization of the debt discounts were immaterial for the year ended December 31, 2017.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

7. Long-term Debt (continued)

As of December 31, 2017, the Company had outstanding borrowings under the Credit Agreement of \$4.7 million, net of discounts of \$0.3 million. Future minimum principal payments, which exclude the end of term charge, related to the Credit Agreement as of December 31, 2017 are as follows (in thousands):

	<u>Amounts</u>
Year ending December 31:	
2018	\$ —
2019	—
2020	2,394
2021	<u>2,606</u>
Total minimum payments	5,000
Less current portion	—
Noncurrent portion	<u>\$5,000</u>

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, 2017, 2016, and 2015 were \$1.3 million, \$1.2 million and \$1.1 million, respectively.

Future minimum lease payments under all noncancelable operating leases as of December 31, 2017, are as follows (in thousands):

	<u>Amounts</u>
Year ending December 31:	
2018	\$ 937
2019	1,316
2020	<u>500</u>
Total minimum lease payments	<u>\$2,753</u>

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, 2017. 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, 2017 and 2016.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Related-Party Transactions (continued)

For the years ended December 31, 2017, 2016, and 2015, the Company paid Bio-Techne \$96,000, \$114,000 and \$62,000, respectively, for research materials. As of December 31, 2017 and 2016, the Company had an accounts payable balance due to Bio-Techne for the purchase of research materials of \$6,000 and \$25,000, respectively.

10. Collaboration and License Agreements

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 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 and other rare diseases. The Company retained 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 and China. 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. The \$85.0 million upfront consideration was initially allocated as of June 30, 2016 as follows:

- \$7.0 million for 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.
- \$12.5 million, which was creditable against an upfront fee payable by Vifor, should the parties enter into a worldwide license agreement for CCX140. The amount creditable decreased ratably into the fourth quarter of 2016. In October 2016, the amount creditable expired and was reclassified to the amortizable portion of deferred revenue as discussed below.
- The remaining upfront consideration of \$65.5 million will be recognized over the estimated period of performance under the Avacopan Agreement, which was initially estimated to approximate 4.2 years, ending in June 2020. The deliverables under the Avacopan Agreement consist of intellectual property licenses, development and regulatory services for the submission of the Marketing Authorization Application (MAA). The Company considered the provisions of the revenue recognition multiple-element arrangement guidance and concluded that the license and the development and regulatory activities for the submission of the MAA do not have stand-alone value because the rights conveyed do not permit Vifor to perform all efforts necessary to use the Company's technology to bring the compound through development and, upon regulatory approval, commercialization of the compound. Accordingly, the Company combined these deliverables and allocated the remaining upfront consideration of \$65.5 million into a single unit of accounting.

Following the October 2016 expiration of the \$12.5 million potentially creditable towards a CCX140 license agreement, such amount was reclassified to the amortizable portion of deferred revenue, which continues to be recognized over the estimated period of performance under the Avacopan Agreement.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

10. Collaboration and License Agreements (continued)

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). 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 and China. In connection with this arrangement, the Company received a \$20.0 million upfront cash commitment for the expanded rights, \$10.0 million of which was received in February 2017. The remaining \$10.0 million is due in February 2018 and is not reflected in accounts receivable as of December 31, 2017. The February 2017 Avacopan Amendment and the original May 2016 Avacopan Agreement are accounted for as a combined agreement. The February 2017 Avacopan Amendment did not represent a material modification given, among other factors, there were no changes to the Company's deliverables under the arrangement. As such, the additional upfront commitment of \$20.0 million under the Avacopan Amendment will be recognized over the remaining estimated period of performance.

In the fourth quarter of 2017, the Company changed its estimated period of performance from 4.2 years to 5.4 years and is recognizing the remaining unamortized portion of the upfront payment over the revised expected remaining performance period, which is estimated to end in September 2021.

For the years ended December 31, 2017 and 2016, the Company recognized \$72.5 million and \$11.4 million, respectively, of collaboration and license revenue under the Avacopan Agreement and the Avacopan Amendment. In December 2017, the Company recognized \$50.0 million of revenue upon achievement of a regulatory milestone when the European Medicines Agency validated the Company's Conditional MAA for avacopan for the treatment of anti-neutrophil cytoplasmic auto-antibody-associated vasculitis. This amount was recorded as accounts receivable as of December 31, 2017.

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.

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 will be responsible for the clinical development of CCX140 in rare renal diseases and will be 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 in CKD in the United States.

Under the terms of the CCX140 Agreement, the Company received a non-refundable upfront commitment of \$50.0 million, all of which was received in 2017. The deliverables under the CCX140 Agreement consist of intellectual property licenses, development and regulatory services for the submission of the MAA. The Company considered the provisions of the revenue recognition multiple-element arrangement guidance and concluded that the license and the development and regulatory activities for the submission of the MAA do not have stand-alone value because the rights conveyed do not permit Vifor to perform all efforts necessary to use

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

10. Collaboration and License Agreements (continued)

the Company's technology to bring the compound through development and, upon regulatory approval, commercialization of the compound. Accordingly, the Company combined these deliverables and allocated the upfront consideration of \$50.0 million into a single unit of accounting. The upfront commitment of \$50.0 million will be recognized over the estimated period of performance under the CCX140 Agreement, which was initially estimated to approximate 5.0 years, ending in December 2021. In the fourth quarter of 2017, the Company changed its estimated period of performance from 5.0 years to 8.0 years and is recognizing the remaining unamortized portion of the upfront payment over the revised expected remaining performance period, which was estimated to end in December 2024.

For the year ended December 31, 2017, the Company recognized \$10.0 million of collaboration and license revenue under the CCX140 Agreement, of which \$9.0 million was associated with the recognition of upfront commitment. The remaining amount represented collaboration revenue derived from funding of CCX140 development services from Vifor. 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.

Under the Avacopan Agreement and the CCX140 Agreement, the Company determined that future contingent payments related to regulatory milestones meet the definition of a substantive milestone under the accounting guidance. Accordingly, revenue for the achievement of these milestones will be recognized in the period when the milestone is achieved. The Company will be eligible to receive contingent payments related to commercial milestones based on the performance of Vifor and these payments are not considered to be milestones under the accounting guidance. These contingent commercial milestone payments will be included in the allocation of arrangement consideration if and when achieved, resulting in an accounting treatment similar to the upfront payment. The Company expects to recognize royalty revenue in the period of sale of the related product, based on the underlying contract terms. The Avacopan Agreement and the CCX140 Agreement are accounted for as separate arrangements.

11. 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.

12. 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, 2017, a total of 11,500,000 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 October 2017, the Board of Directors approved an increase to the number of shares reserved for issuance under the 2012 Plan by 1,940,000 shares effective January 1, 2018. 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)

12. Equity Incentive Plans (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, 2016 and 2015, the weighted average grant date fair value for restricted stock granted was \$4.48 and \$7.88, respectively. The total fair value of restricted stock vested during the years ended December 31, 2017, 2016 and 2015 was \$1.7 million, \$0.2 million and \$1.0 million, respectively.

The activity for restricted stock is summarized as follows:

	<u>Shares</u>	<u>Weighted Average Grant-Date Fair Value</u>
Balance at December 31, 2016	471,650	\$4.60
Granted	279,738	6.72
Vested	(229,610)	4.60
Canceled	(13,334)	3.57
Unvested at December 31, 2017	<u>508,444</u>	\$5.79

As of December 31, 2017, there was \$1.7 million of unrecognized compensation expense associated with unvested restricted stock, which is expected to be recognized over a weighted-average period of 1.4 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)

12. Equity Incentive Plans (continued)

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

	<u>Available for Grant</u>	<u>Shares</u>	<u>Weighted Average Exercise Price</u>	<u>Weighted Average Remaining Contractual Term (in years)</u>	<u>Aggregate Intrinsic Value</u>
Balance at December 31, 2016	1,655,524	9,345,515	\$7.72		
Shares authorized	1,900,000	—			
Granted ⁽¹⁾	(2,212,138)	1,932,400	6.80		
Exercised ⁽²⁾	58,967	(461,151)	5.84		
Forfeited and expired ⁽³⁾	626,527	(613,193)	7.03		
Outstanding at December 31, 2017	<u>2,028,880</u>	<u>10,203,571</u>	\$7.68	6.19	\$3,773,137
Vested and expected to vest, net of estimated forfeiture at December 31, 2017		<u>9,967,860</u>	\$7.72	6.13	\$3,636,358
Exercisable at December 31, 2017		<u>7,099,052</u>	\$8.37	5.10	\$1,798,555

- (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.
- (3) The difference between shares forfeited and expired in the number of shares available for grant and outstanding options represents the RSUs canceled for the period.

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 \$1.3 million, \$0.7 million and \$2.1 million during 2017, 2016, and 2015, respectively. As of December 31, 2017, there was \$8.2 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.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

12. Equity Incentive Plans (continued)

As of December 31, 2017, stock options outstanding were as follows:

<u>Exercise Price Range</u>	<u>Options Outstanding</u>	
	<u>Shares</u>	<u>Weighted-Average Contractual Life</u>
\$2.10 - \$3.29	214,050	8.32
\$3.57	1,151,175	8.08
\$3.72 - \$5.95	486,090	8.52
\$6.00	1,054,514	1.18
\$6.08 - \$6.30	1,134,724	4.61
\$6.50 - \$6.60	96,087	9.30
\$6.62	1,175,900	9.10
\$6.90 - \$7.10	1,113,845	5.90
\$7.12 - \$7.85	418,100	8.87
\$8.19 - \$15.90	3,359,086	5.84
	<u>10,203,571</u>	<u>6.19</u>

Employee Stock Purchase Plan

In February 2012, the stockholders approved the ESPP. As of December 31, 2017, a total of 950,000 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 October 2017, the Board of Directors approved an increase to the number of shares reserved for issuance under the ESPP by 150,000 shares effective January 1, 2018.

The Company issued 93,221 shares, 157,893 shares and 134,579 shares under the ESPP in 2017, 2016 and 2015, respectively. As of December 31, 2017, 333,335 shares were available for issuance under the ESPP. As of December 31, 2017, 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):

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Research and development	\$3,154	\$3,245	\$3,240
General and administrative	4,965	4,977	5,620
Total	<u>\$8,119</u>	<u>\$8,222</u>	<u>\$8,860</u>

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

12. Equity Incentive Plans (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 2017, 2016, and 2015 were estimated at the date of grant using the Black-Scholes option-pricing model with the following assumptions:

	<u>Employee Stock Options</u>			<u>Employee Stock Purchase Plan</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>	<u>2017</u>	<u>2016</u>	<u>2015</u>
Dividend yield	0%	0%	0%	0%	0%	0%
Volatility	68.3%	65.6%	67.6%	52.9%	99.7%	60.1%
Weighted-average expected life (in years)	6.0	6.0	6.0	0.5	0.5	0.5
Risk-free interest rate	2.04%	1.58%	1.70%	1.22%	0.47%	0.21%
Weighted average grant date fair value	\$4.30	\$2.43	\$5.00	\$2.16	\$2.29	\$2.22

Stock Options Granted to Nonemployees

During 2017, 2016 and 2015, the Company granted to consultants options to purchase 239,266, 15,000, and 90,300 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):

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Research and development	\$629	\$313	\$105
General and administrative	—	—	20
Total	<u>\$629</u>	<u>\$313</u>	<u>\$125</u>

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:

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Dividend yield	0%	0%	0%
Volatility	69-70%	65-68%	65-66%
Weighted-average expected life (in years)	5.5-10.0	6.1-9.9	5.6-9.9
Risk-free interest rate	1.9-2.5%	1.3-2.4%	1.7-2.4%

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

13. 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, 2017, 2016, and 2015.

14. 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):

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
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	<u>\$—</u>	<u>\$—</u>	<u>\$—</u>

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

	<u>Year Ended December 31,</u>		
	<u>2017</u>	<u>2016</u>	<u>2015</u>
Federal statutory income tax rate	34.0%	(34.0%)	(34.0%)
State income taxes, net of federal benefit	—	(5.8)	(5.8)
Permanent items	5.5	2.1	1.9
Excess tax benefit for stock-based compensation	(2.0)	—	—
Research and development credits	(7.2)	(2.8)	(2.1)
Change in valuation allowance	(224.1)	38.1	40.0
Change in tax rate	193.8	—	—
Other	—	2.4	—
(Benefit from) provisions for income taxes	<u>— %</u>	<u>— %</u>	<u>— %</u>

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

14. 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,	
	2017	2016
Net operating loss carryforwards	\$ 55,569	\$ 102,269
Research and development credit	10,470	8,719
Amortization of deferred stock compensation - non-qualified	6,385	10,567
Reserves and accruals	785	1,747
Deferred revenue	10,297	—
Depreciation and amortization	231	445
Net deferred tax assets	83,737	123,747
Less: valuation allowance	(83,737)	(123,747)
Net deferred tax assets	\$ —	\$ —

On December 22, 2017, the Tax Cuts and Jobs Act (“the Act”) was enacted into law. The new tax legislation 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. The Company has analyzed the potential impacts of the Act and due to the federal rate reduction it is expecting a reduction in its federal deferred tax assets by \$36.0 million with an offset to the valuation allowance, which has been reflected in the financial statements for the tax year ended December 31, 2017.

Accounting Standards Codification (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 assets based on its current understanding of the provisions of the new law. The Company considers its accounting for the impacts of the new law to be provisional and the Company will continue to assess the impact of the recently enacted tax law (and expected further guidance from federal and state tax authorities as well as further guidance for the associated income tax accounting) on its business and consolidated financial statements over the next 12 months.

The Company has concluded that it is more likely than not that its deferred tax assets will not be realized. Accordingly, the total deferred tax assets have been fully offset by a valuation allowance. The Company’s valuation allowance decreased by approximately \$40.0 million in 2017 and increased by approximately \$15.2 million and \$19.3 million during 2016 and 2015 respectively.

At December 31, 2017, the Company had federal and state net operating loss carryforwards of approximately \$192.5 million and \$247.9 million, respectively. The federal net operating loss carryforwards will begin to expire in 2031. The state net operating loss carryforwards will begin to expire in 2018.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

14. Income Taxes (continued)

The Company has federal and state research and development credit carryforwards of \$11.2 million and \$6.1 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, 2017, 2016, and 2015, is as follows (in thousands):

	Unrecognized Income Tax Benefits
Balance as of December 31, 2015	\$4,872
Additions for current tax positions	<u>558</u>
Balance as of December 31, 2016	\$5,430
Additions for current tax positions	603
Additions for prior tax positions	<u>2,753</u>
Balance as of December 31, 2017	<u>\$8,786</u>

As of December 31, 2017 and 2016, the Company had approximately \$8.8 million and \$5.4 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. 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 2014 and 2013, 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, 2017, there has been no interest expense or penalties related to unrecognized tax benefits.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

15. Selected Quarterly Financial Data (unaudited)

Selected quarterly results from operations for the years ended December 31, 2017 and 2016 are as follows (in thousands except per share amounts):

	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
	2016 Quarter Ended			
	March 31	June 30	September 30	December 31
Revenue	\$ —	\$ 2,795	\$ 4,251	\$ 4,889
Net loss	\$(15,243)	\$(9,983)	\$(7,072)	\$(7,665)
Basic net loss per share	\$ (0.34)	\$ (0.22)	\$ (0.15)	\$ (0.16)
Diluted net loss per share	\$ (0.34)	\$ (0.22)	\$ (0.15)	\$ (0.16)

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.

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 12, 2018

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.

<u>Signature</u>	<u>Title</u>	<u>Date</u>
<u>/s/ Thomas J. Schall, Ph.D.</u> Thomas J. Schall, Ph.D.	President, Chief Executive Officer and Director (Principal Executive Officer)	March 12, 2018
<u>/s/ Susan M. Kanaya</u> Susan M. Kanaya	Executive Vice President, Chief Financial and Administrative Officer and Secretary (Principal Financial and Accounting Officer)	March 12, 2018
<u>/s/ Pui San Kwan</u> Pui San Kwan	Vice President, Finance (Principal Accounting Officer)	March 12, 2018
<u>/s/ Thomas A. Edwards</u> Thomas A. Edwards	Director	March 12, 2018
<u>/s/ Joseph M. Feczko, M.D.</u> Joseph M. Feczko, M.D.	Director	March 12, 2018
<u>/s/ Roger C. Lucas, Ph.D.</u> Roger C. Lucas, Ph.D.	Director	March 12, 2018
<u>/s/ Henry A. McKinnell, Jr., Ph.D.</u> Henry A. McKinnell, Jr., Ph.D.	Director	March 12, 2018
<u>/s/ Geoffrey M. Parker</u> Geoffrey M. Parker	Director	March 12, 2018
<u>/s/ James L. Tyree</u> James L. Tyree	Director	March 12, 2018

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CORPORATE INFORMATION

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TRANSFER AGENT

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<http://www.amstock.com/new/InvestorServices/ShareholderServices.aspx>

INDEPENDENT REGISTERED PUBLIC ACCOUNTING FIRM

Ernst & Young LLP
Redwood City, CA

FORM 10-K

A copy of the ChemoCentryx annual report on Form 10-K filed with the Securities and Exchange Commission is available free of charge from the company's Corporate Communications Department by calling 650.210.2900.

STOCK INFORMATION

The common stock of the company has traded on the Nasdaq Global Select Market under the symbol "CCXI" since February 8, 2012. No dividends have been paid on the common stock since the company's inception.

BOARD OF DIRECTORS

Thomas J. Schall, PhD
President, Chief Executive Officer and Chairman of the Board
ChemoCentryx, Inc.

Roger C. Lucas, PhD
Former Vice Chairman, Bio-Techne Corporation
Lead Independent Director, ChemoCentryx, Inc.

Thomas A. Edwards
Retired Partner, Latham & Watkins, LLP
Chairman of the Nominating and Corporate Governance Committee,
ChemoCentryx, Inc.

Joseph M. Feczko, MD
Former Senior Vice President and Chief Medical Officer, Pfizer Inc.

Henry A. McKinnell, Jr., PhD
Retired Chairman and Chief Executive Officer, Pfizer Inc.

Geoffrey M. Parker
Senior Vice President and Chief Financial Officer, Tricida, Inc.

James L. Tyree
Former Executive Vice President, Abbott Laboratories
Chairman of the Audit Committee, ChemoCentryx, Inc.
Chairman of the Compensation Committee, ChemoCentryx, Inc.

MANAGEMENT

Thomas J. Schall, PhD
President, Chief Executive Officer and Chairman of the Board

William C. Fairey, Jr.
Executive Vice President, Chief Operating Officer

Susan M. Kanaya
Executive Vice President, Chief Financial and Administrative Officer
and Secretary

Markus J. Cappel, PhD
Chief Business Officer and Treasurer

Jan Hillson, MD, PhD
Senior Vice President, Drug Development

Rajinder Singh, PhD
Senior Vice President, Research



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