



2016

ANNUAL REPORT

GREAT SCIENCE | GREAT MEDICINE



CHEMOCENTRYX

Delivering on important promises at ChemoCentryx

At ChemoCentryx (CCXI) we are creating value for patients and shareholders by developing new medicines for diseases where therapeutic innovation have been previously lacking.

In 2016 our mission became clearer, our focus sharper, and our balance sheet stronger. We are an emerging power in orphan and rare disease, particularly rare renal disease.

In this letter, I'll review some of the reasons for our corporate focus. I'll cover some of the progress of the past year, since that guides us in taking our next step in bringing our unique medications to an awaiting community. We'll discuss the next steps, as well as some of the challenges.

Our mission and commitments are unwavering: we aim to create value for patients and also for shareholders of ChemoCentryx, by moving our therapeutic agents now through late stage development, regulatory approvals and product launch, and marketing those new medications to specialists who treat the rare diseases on which we focus. Key pieces of this plan have already been accomplished. If successful with the next step, the Company should be on a path to self-sustainability and eventual profitability.

We're proud of the progress we've made in the execution of our long term plan

As I consider the history of our enterprise and its continued progress, the evidence shows that we are executing on our master plan. This plan might be thought of as ascending in four key steps:

1. Discovery of novel drug candidates from our unique research platform;
2. Clinical development (indeed, now including late stage clinical development) of these unique proprietary compounds;
3. Partnership formation in our area of clinical focus (orphan and rare renal disease), providing good cash inflows, and retention of key rights; and
4. Forward integration to marketing, by building CCXI's capacity to market our drugs at home, and through our partners abroad.

By the end of 2016 we had scaled the first three major steps in the plan; with those accomplishments representing promises fulfilled. We now have sights set firmly on the added promises that we now need to keep: building capacity to market our products that are approved; achieving success in the fourth step.

2016 was a year of marked accomplishment for the Company

In previous communications to shareholders I committed, on behalf of all of CCXI, to some important and challenging goals: (i) to being a late stage development company by the end of 2016, including the launch of a Phase III clinical trial in our lead program, (ii) to further develop our emerging priorities in orphan and rare disease, and (iii) to raise capital by managing our pipeline and pursuing strategically appropriate business development deals.

The Company has delivered on those commitments.

We successfully funded the Company in 2016, strengthening the balance sheet considerably. While financial headwinds buffeted the life science industry throughout most of 2016, CCXI was able to sail against the trend. In the seven month period from May to December 2016, the Company brought in \$135M in upfront cash and commitments via business development agreements. Importantly, the agreements, outlined below, retain for CCXI central rights (e.g., U.S. marketing entirely), while also providing excellent near-term and downstream economics.

We also delivered on our other promises: we became a Phase III clinical development company by launching the ADVOCATE trial with our drug avacopan (formerly CCX168) in the treatment of the autoimmune disease known as anti-neutrophil cytoplasmic autoantibody-associated vasculitis (ANCA-vasculitis or AAV). Moreover, we have

considerably expanded our efforts in orphan and rare disease, particularly in rare renal disease.

This last point raises the questions of 'Why renal disease?', and 'Why rare disease'?

The answers relate to the unique science we practice at CCXI, and where that science and medicine reveals the greatest potential to create value for patients and shareholders.

Our discovery platform is based on the science of cellular chemoattraction, which is the directed migration of cells in the body. At CCXI we are concerned mostly with the chemoattraction of different varieties of destructive inflammatory cells in autoimmune diseases. Damaging inflammatory cells are guided to the sites they damage by cues in the body, known as chemoattractants, which bind specifically to chemoattractant receptors. The receptors (frequently referred to simply as chemokine receptors) are the molecular 'guidance system' for inflammatory cells. Our discovery technology is designed specifically to jam the chemoattractant receptor guidance systems with orally administered compounds that bind to the chemoattractant receptors, thus rendering those targeted receptors non-functional. By eliminating the attraction, accumulation, and persistent activation of the disease-causing cells in a given pathology, we aim to stop ongoing tissue damage, providing the opportunity for organs to repair and rebuild and function more healthily.

The CCXI approach offers distinct advantages: our therapeutics have more specific (and not broadly immune-suppressive) modes of action than current standard of care therapies (SOCs). Also, our medications are orally-administered. By contrast, SOC's in the indications in which we are interested are frequently injectables or infusibles, and/or immunosuppressive agents with very serious safety issues.

The CCXI discovery platform is quite powerful. We have employed it to develop a broad pipeline of clinical candidates for various indications, and we are still using the platform to discover potential novel therapies.

Using our discovery platform, we at CCXI also thoroughly interrogate the pathobiology of the different chemoattractant receptors and the disease-inducing cells they guide. Science at CCXI, and amassing science from other sources, has provided a growing body of evidence for the involvement of two important chemoattractant receptors in various chronic kidney diseases (CKDs). The chemoattractant receptors most involved in CKDs are the complement C5a receptor (C5aR), the target of our drug candidate avacopan, and another chemokine receptor, CCR2, which is the target of our drug candidate CCX140.

Identifying C5aR and CCR2 as drivers of kidney pathology had important implications. CKDs cause immense human suffering and are a staggering economic burden to health care systems around the world. Kidney diseases can ultimately result in end stage renal disease (ESRD), that is, the need for dialysis or the need for renal transplant. In the U.S., ESRD expenses comprise approximately 8% of the Medicare budget, a staggering sum of many tens of billions of dollars. It is ironic, however, that clinical development in large prevalence CKDs (such as the kidney disease common in diabetes) can be very difficult and extremely expensive, thus discouraging therapeutic innovation. This is a regulatory and economic puzzle that society has yet to solve. For the present, independent clinical development of a new agent in large prevalence CKD is out of reach for an organization of the scale and scope of CCXI.

But, C5aR and CCR2 are also very important in orphan and rare renal disease.

Orphan and rare diseases provide attractive routes for product development at CCXI. Orphan diseases can provide a clinical and regulatory development path to eventual approval that is tractable and affordable for a company of CCXI's size and stage of corporate maturity.

Importantly, too, in rare disease the human need is very high. Often there are no FDA-approved therapies at all, and when current SOC therapies do exist, frequently they are inadequate or carry with them toxic side effects. For example, chronic high dose corticosteroid therapy is used in the treatment of AAV. Steroid usage of this kind has many toxic side effects; also it must be tapered, contributing to relapse and ongoing accumulated tissue damage.

While clinical development of orphan disease does carry with it unique challenges (small patient numbers for trials, a dearth of defined and validated regulatory endpoints,

etc.), innovation can be amply rewarded. Investment risk in rare disease clinical development is relatively lower than in larger indications, and reasonable pricing for orphan and rare disease is still supported by payers and society. For these reasons, CCXI believes that orphan and rare diseases provide an ideal path for some of the unique assets in our pipeline, particularly for avacopan and CCX140 in rare renal disease.

Accordingly, our focus at CCXI has evolved. We at CCXI intend to be a force to be reckoned with in rare renal disease, and we have a late stage pipeline tailor-made for this purpose. Avacopan and CCX140 are moving forward in what we believe to be high value creation areas. Avacopan is currently in Phase III for AAV. Furthermore, we aim to expand the clinical development footprint of avacopan this year with trials in additional kidney diseases, including atypical hemolytic uremic syndrome (aHUS), a rare and very serious kidney disease where the SOC is not always accessible to patients, and C3 glomerulopathy (C3G disease), a rare disorder that strikes adolescents and young adults, requiring dialysis and transplant, and for which there is no currently approved therapy. The expanded clinical development footprint for avacopan represents the execution of our 'pipeline in a drug' strategy for this unique molecule.

Separately, CCX140 has completed a successful Phase II trial in diabetic chronic kidney disease (D-CKD), and we intend now to launch a study this year in a rare form of CKD known as focal segmental glomerulosclerosis (FSGS). FSGS is an orphan kidney disease also resulting in ESRD and for which there is no FDA approved therapy.

Our intent is that all of the studies with avacopan and CCX140 that we have ongoing (or plan to initiate this year) will be designed such that if they are successful, the data could support registration of the drug in its trialed indication.

The maturing of our pipeline and focus on rare renal diseases exemplifies the broader achievements of 2016, a year of marked progress. Let's consider even a partial list of 2016 achievements:

- Successful completion of the Phase II development program for avacopan in AAV (with positive results in the CLEAR and CLASSIC trials)
- Signing of regional marketing deal with Vifor on avacopan for rare renal disease
- Successful end of phase 2 meetings with the EMA and the FDA with buy-in on single Phase III registration trial design for avacopan in AAV
- Initiation of Phase III 'ADVOCATE' registration trial in AAV
- Award of FDA orphan office grant for AAV clinical work
- New 'PRIME' designation from the EMA for accelerated assessment of avacopan in AAV
- Positive proof of concept data from dosing avacopan in aHUS patients
- Successful treatment throughout 2016 of a patient with C3G. This individual has been treated with avacopan continuously since 2015, under the U.K. equivalent of a compassionate use protocol. At the time his avacopan treatment started, his transplanted kidney was failing to C3G. At time of this writing, his kidney function had been stable for the year and a half he has been dosed, and the disease has shown marked clearance in successive renal biopsies with avacopan treatment.
- Conducting a clinical trial (still ongoing) in inoperable pancreatic cancer with CCX872
- Signing of Vifor—CCXI regional marketing deal for CCX140 in orphan renal disease and CKD
- Budget and expenditures in 2016 ran favorable to forecast

I think that it is fair to say that we fulfilled many promises in 2016.

Our recent deal-making provides some very important strategic pieces for our business.

I'd like to discuss briefly the important alliance between CCXI and renal care leader Vifor Pharma (and their associated partners Fresenius Medical Care). In May of 2016 we initiated a series of agreements with Vifor, and our alliance now involves both avacopan and CCX140. Importantly, CCXI retains U.S. rights entirely to these assets (also we retain rights in China), and Vifor has marketing rights outside the U.S.

The Vifor relationship has been key to creating value for shareholders:

- \$135 million in upfront cash payments and commitments to CCXI, strengthening our balance sheet so that we can move forward with confidence on our late stage clinical studies, with CCXI retaining rights to U.S. entirely
- Pre-royalties potential of the combined deals is approximately \$1.2 billion. Downstream royalties are substantial—teens to mid-twenties percent, tiered on sales
- CCXI also controls the clinical development of avacopan and CCX140 in rare disease

In addition to substantially adding to our cash reserves, investors will note that the Vifor transaction provides thorough validation of avacopan and CCX140 in renal disease. Vifor's diligence process, which resulted in their substantial investment in both the avacopan and CCX140 programs, provided them a deep understanding of these two rare renal assets and their potential.

Our agreements with Vifor, which have been described by some as elegant, brought in cash, but more importantly filled a fundamental strategic requirement for CCXI. We now have two assets in our rare renal portfolio, reducing our development risk and creating efficiencies for CCXI's forward integration strategy. We and others believe approval of these agents in the indications we are pursuing comprises market opportunities in the billions of dollars. The licensing of the international rights for our two late stage drugs allows us to focus on commercializing in what is widely considered to be the world's biggest market, the U.S. In short, we at CCXI will avoid the expensive cost structure and complexity of operating in multiple international markets, instead looking forward to a healthy stream of royalties from abroad, while we concentrate on forward integration to marketing these new therapeutics at home.

As we move into the future, we have promises to keep and intend to do so

As we move through 2017, we intend to continue to create value as we build the next part of our enterprise. We have fulfilled the promise of last year by becoming a late stage Phase III development company. The fact that this late stage development is occurring with unique assets of our own discovery, which we own entirely in the U.S., and for which we, the innovator company, are controlling development, is remarkable in itself. This is no small achievement for a pre-revenue biopharma enterprise.

We are now focused on the future. The list of goals for this year is ambitious: we will advance the Phase III ADVOCATE trial of avacopan in AAV, and launch other clinical endpoint studies for avacopan in diseases such as aHUS and, very importantly, C3G. We plan to launch a controlled trial for CCX140 in FSGS as well. We believe that any of those four trials in orphan renal disease, if successful, could lead ultimately to registration of avacopan or CCX140 in the respective kidney indication tested. Also in 2017, we will publish additional data from our clinical studies, including the Phase II results of avacopan for the treatment of AAV, and hope to achieve additional regulatory advancements, including Orphan Drug designations for avacopan and CCX140 in additional clinical indications.

All of these goals are in the service of the larger mission of the enterprise: to now build assertively toward forward integration such that we will have the capacity to market these innovations at home and abroad upon approval, improving and saving lives as a consequence. The creation of that kind of value – from concept to commercialization – is typically reserved for the stuff of breezy and wistful speculation. At ChemoCentryx, we are ever more close to experiencing it as reality.



CCXI LATE STAGE: KIDNEY DISEASE FRANCHISE

THERAPEUTIC AREA	DRUG	INDICATION (TARGET)	Preclinical	Phase I	Phase II	Phase III
Complement Inhibition in Orphan and Rare Diseases	Avacopan (formerly CCX168)	ANCA ASSOCIATED VASCULITIS (C5aR)				
		ATYPICAL HEMOLYTIC UREMIC SYNDROME (C5aR)				
		C3 GLOMERULOPATHY (C5aR)				
Chronic and Other Rare Kidney Diseases	CCX140	FOCAL SEGMENTAL GLOMERULOSCLEROSIS (CCR2)				
		DIABETIC NEPHROPATHY (CCR2)				

CCXI EARLY STAGE DRUG CANDIDATES

THERAPEUTIC AREA	DRUG	INDICATION (TARGET)	Preclinical	Phase I	Phase II	Phase III
Immuno-Oncology	CCX872	ADVANCED PANCREATIC CANCER (CCR2)				
		OTHER ONCOLOGY TARGETS (CCR1, CXCR2)				
Other Inflammatory and Autoimmune Diseases	VERCIRNON*	INFLAMMATORY BOWEL DISEASE (IBD): CROHN'S DISEASE (CCR9)				
	CCX507	IBD: ULCERATIVE COLITIS (CCR9)				
	CCX991	PSORIASIS AND TH17 DISEASES (CCR4)				

*Also known as CCX282 or Traficet-EN

**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, 2016

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:

Title of Each Class

Name of Each Exchange on Which Registered

Common Stock, par value \$0.001 per share

The NASDAQ Stock Market LLC

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

None

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

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

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

Indicate by check mark whether the registrant has submitted electronically 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 or a smaller reporting company. See definitions of "large accelerated filer", "accelerated filer" and "smaller reporting company" in Rule 12b-2 of the Exchange Act. (Check one):

Large accelerated filer

Accelerated filer

Non-accelerated filer

Smaller reporting company

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 \$68.0 million, based on the closing price of the registrant's common stock on the NASDAQ Global Select Market of \$4.49 per share.

The number of outstanding shares of the registrant's common stock, par value \$0.001 per share, as of March 3, 2017 was 48,153,311.

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 2017 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, 2016.

[THIS PAGE INTENTIONALLY LEFT BLANK]

CHEMOCENTRYX, INC.
FORM 10-K — ANNUAL REPORT
For the Fiscal Year Ended December 31, 2016

Table of Contents

	<u>Page</u>
PART I	
Item 1 Business	3
Item 1A Risk Factors	32
Item 1B Unresolved Staff Comments	62
Item 2 Properties	63
Item 3 Legal Proceedings	63
Item 4 Mine Safety Disclosures	63
PART II	
Item 5 Market for Registrant’s Common Equity, Related Stockholder Matters and Issuer Purchases of Equity Securities	64
Item 6 Selected Financial Data	67
Item 7 Management’s Discussion and Analysis of Financial Condition and Results of Operations	68
Item 7A Quantitative and Qualitative Disclosures About Market Risk	78
Item 8 Financial Statements and Supplementary Data	78
Item 9 Changes in and Disagreements with Accountants on Accounting and Financial Disclosure	78
Item 9A Controls and Procedures	78
Item 9B Other Information	79
PART III	
Item 10 Directors, Executive Officers and Corporate Governance	80
Item 11 Executive Compensation	80
Item 12 Security Ownership of Certain Beneficial Owners and Management and Related Stockholder Matters	80
Item 13 Certain Relationships and Related Transactions, and Director Independence	80
Item 14 Principal Accounting Fees and Services	80
PART IV	
Item 15 Exhibits, Financial Statement Schedules	81
Signatures	

[THIS PAGE INTENTIONALLY LEFT BLANK]

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 focuses on a specific chemoattractant receptor that selectively blocks its negative inflammatory or suppressive response, 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 drugs 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 us manage the wide array of opportunities, we have segmented our pipeline into early stage and late stage compounds.

Late Stage Compounds

We have chosen to focus initially on kidney disease, particularly on rare indications, where orphan 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 and Rare Diseases

Avacopan (CCX168) is an orally-administered complement inhibitor targeting the C5a receptor, or C5aR, and is being developed for orphan and rare diseases, including 1) anti-neutrophil cytoplasmic auto-antibody associated vasculitis, or AAV, a devastating autoimmune disease that destroys blood vessels and can lead to kidney failure; 2) atypical hemolytic uremic syndrome, or aHUS, a rare, life threatening disease; and 3) complement 3 glomerulopathy, or C3G, a debilitating kidney disease.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV and aHUS and by the European Medicines Agency, or EMA for the treatment of 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 accelerate its marketing authorization.

Following completion of two Phase II clinical trials in patients with AAV, the results of which demonstrated that avacopan was safe, 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 300 patients in up to 200 sites in the United States and Europe. We also plan to initiate clinical endpoint trials, ones that could potentially serve as registration trials, of avacopan for the treatment of patients with C3G and aHUS in 2017.

CCX140 — Chronic and Rare Kidney Diseases

CCX140 is 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 Phase II clinical trial of CCX140 in patients with DN met its primary endpoint by demonstrating that CCX140 given orally once daily added to an SOC renin-angiotensin-aldosterone system inhibitor treatment resulted in a statistically significant reduction in proteinuria, beyond that achieved with SOC alone. Based on the safety and efficacy data related to reduction in proteinuria and improvement of renal function observed in the Phase II trial in DN, we plan to initiate in 2017 a clinical endpoint trial of CCX140 for the treatment of patients with FSGS, for which there are currently no FDA-approved treatments.

Global Kidney Health Alliance with Vifor

In May 2016 we announced a partnership with Vifor (International) Ltd., or Vifor, a European-based world leader specializing in kidney disease, for the commercial rights to avacopan in Europe and certain other international markets, the Avacopan Agreement. We expanded our partnership with Vifor in December 2016 with an additional deal for our other late stage drug candidate, CCX140, whereby we granted Vifor worldwide rights outside of the United States and China; and in February 2017, we announced a further deal with Vifor that harmonized the geographic commercialization rights underlying the agreements for both drug candidates.

We have secured \$155 million in upfront cash payments and commitments, plus substantial potential milestone payments pursuant to our agreements with Vifor. Through our alliance, we maintain the commercial rights of avacopan and CCX140 in the United States and China, and also retain control of the clinical development programs for rare renal disease. Vifor gains the commercial rights for all other international markets, and will pay us double-digit tiered royalties 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 Compounds

While the science has led us to focus initially on kidney disease, our targeted blocking system 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 from other areas of inflammatory and autoimmune disorders, such as rheumatoid arthritis, inflammatory bowel disease, and psoriasis, as well as cancer, where our drug candidate CCX872 has shown promise in a Phase I trial for non-operable pancreatic cancer. 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	INDICATION (TARGET)	PRECLINICAL	PHASE I	PHASE II	PHASE III
Complement Inhibition in Orphan and Rare Diseases	Avacopan (formerly CCX168)	ANCA ASSOCIATED VASCULITIS (C5aR)	[Progress bar]			
		ATYPICAL HEMOLYTIC UREMIC SYNDROME (C5aR)	[Progress bar]			
		C3 GLOMERULOPATHY (C5aR)	[Progress bar]			
Chronic and Other Rare Kidney Diseases	CCX140	FOCAL SEGMENTAL GLOMERULOSCLEROSIS (CCR2)	[Progress bar]			
		DIABETIC NEPHROPATHY (CCR2)	[Progress bar]			

CCXI Early Stage Drug Candidates

THERAPEUTIC AREA	DRUG	INDICATION (TARGET)	PRECLINICAL	PHASE I	PHASE II	PHASE III
Immuno-Oncology	CCX872	ADVANCED PANCREATIC CANCER (CCR2)				
		OTHER ONCOLOGY TARGETS (CCR1, CXCR2)				
Other Inflammatory and Autoimmune Diseases	Vercirnon*	INFLAMMATORY BOWEL DISEASE (IBO): CROHN'S DISEASE (CCR9)				
	CCX507	IBD: ULCERATIVE COLITIS (CCR9)				
	CCX991	PSORIASIS AND TH17 DISEASES (CCR6)				

Late Stage Compounds

Avacopan (Formerly CCX168) — Complement Inhibition in Orphan and Rare Diseases

In our complement inhibition orphan and rare diseases program, our lead drug candidate is avacopan. Avacopan is a small molecule that targets the chemoattractant receptor known as C5aR, and is being developed for inflammatory and autoimmune diseases. Avacopan blocks 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 destroy foreign invaders (such as 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 the United States, under the Orphan Drug Act, the FDA has granted orphan drug designation for avacopan for the treatment of AAV, including granulomatosis with polyangiitis or Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome, and also for aHUS. 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. The European Commission granted orphan drug designation for avacopan for the treatment of granulomatosis with polyangiitis or Wegener's granulomatosis and microscopic polyangiitis.

Our most advanced clinical program in complement inhibition of orphan and rare diseases is in Phase III development in patients with AAV. We also have conducted a pilot clinical trial in patients with aHUS. Additionally, through a Special Needs protocol in the United Kingdom, we have treated one patient with C3G. In May 2016, pursuant to the Avacopan Agreement, Vifor licensed the rights to commercialize avacopan for orphan and rare renal diseases in Europe, certain other markets outside the United States and most of Asia. In February 2017, we agreed to amend the Avacopan Agreement 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.

AAV, C3G and aHUS are all rare autoimmune diseases that are characterized by inflammation that often affects the kidneys.

ANCA-Associated Vasculitis (AAV)

AAV is a rare, severe, and often fatal autoimmune disease that is caused by autoantibodies called anti-neutrophil cytoplasmic antibodies 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, 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.

Avacopan — A Novel C5a Receptor Inhibitor

Avacopan is a potent and highly specific inhibitor of the human C5a receptor. Avacopan 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 membrane attack complex), unlike the anti-C5-antibody, eculizumab. 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

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 CYC or RTX treatment. The primary efficacy objective was to evaluate the efficacy 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 which 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:

- 1) 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 MCP-1 (a marker of kidney inflammation) found in the urine;
- 2) 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
- 3) 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 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 30 mg 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 estimated glomerular filtration rate appeared to improve most in the 30 mg avacopan group, and renal response (based on improvement in hematuria, albuminuria, and eGFR) appeared to be highest in the 30 mg avacopan group.

Taken together, these results suggest that avacopan, a target-specific complement inhibitor, may be able to provide effective control of the disease while eliminating chronic steroids in the treatment of AAV. Avacopan also appeared safe and well-tolerated in the trial. 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.

In May 2016, we received PRIME designation for avacopan for AAV in Europe. This was based on the assessment by the EMA that (i) AAV is a highly severe disease with high mortality; (ii) current 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 held End-of-Phase II meetings with the FDA, and Protocol Assistance/Scientific Advice meetings with the EMA in 2016, and we reached agreement on the design and scope of the Phase III registration clinical trial in AAV, which was initiated in December 2016.

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 include approximately 300 patients with newly diagnosed or relapsing AAV. 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 believe that ADVOCATE, if successful, could initiate avacopan's commercial opportunity by achieving a marketing authorization in Europe and in the United States.

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

Under the Special Needs program in the United Kingdom (similar to compassionate use protocols in the United States), in September 2015 a C3G renal transplant recipient with deteriorating kidney function initiated treatment with and has responded well to treatment with avacopan. After only one month of initial treatment with avacopan, renal function (based on estimated glomerular filtration rate, or eGFR) stabilized. Moreover, sequential kidney biopsies taken after the patient had been on avacopan for two and seven months showed continued improvement in kidney histology based on a decrease in glomerular endocapillary proliferation and a marked reduction in the number of glomerular inflammatory macrophages, both histologic markers of disease activity as compared to the pre-treatment biopsy.

Prior to receiving treatment with avacopan, the C3G patient had received treatment with a wide spectrum of immunosuppressant drugs including RTX, CYC, mycophenolate mofetil, tacrolimus and glucocorticosteroids. All of these previous treatments had failed to prevent disease recurrence and progression, including new disease in the patient's transplanted kidney. The patient has been successfully treated with avacopan since September 2015 and remains on treatment. These findings provide the first evidence that avacopan may be effective in treating patients with this rare and debilitating disorder for which there are no approved therapies. We plan to initiate a clinical trial to potentially support registration of avacopan for the treatment of patients with C3G in 2017.

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.

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 complement factor H, or 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 (Soliris®) 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 can cost approximately \$500,000 per year in the United States.

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 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 terminal complement complex (or membrane attack complex), which is important in fighting *Neisseria* infections. Since avacopan is a small molecule, manufacturing cost is anticipated to be lower than the 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.

Clinical Development

In collaboration with external scientists, we have demonstrated that avacopan, when added to the serum of patients with aHUS, reduces the size of thrombus (blood clot) formation on vascular endothelial cells that has been stimulated by the aHUS serum. The positive effect of avacopan was concentration dependent, and the magnitude of the effect was similar to that observed with eculizumab or soluble complement receptor 1.

Based on these encouraging in vitro findings, we have performed a Phase II pilot clinical trial with avacopan in six patients with aHUS who are on dialysis. The primary efficacy objective of the trial is 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 plan to initiate a clinical trial for the treatment of patients with aHUS in 2017 to potentially support registration of avacopan in aHUS.

Avacopan Commercialization Strategy

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

In May 2016, we entered into the Avacopan Agreement with Vifor to commercialize avacopan for orphan and rare 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 agreed to amend the Avacopan Agreement expand the licensed territory to include all markets outside the United States and China and we received an additional \$20 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.

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 Rare Kidney Diseases

Our second drug candidate in the orphan and rare disease space is CCX140, an inhibitor of the chemokine receptor known as CCR2.

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

Renin-angiotensin-aldosterone system, or RAAS, inhibition 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 MCP-1 and a biomarker of inflammation, and the infiltration of immune cells called macrophages into the kidney in patients with chronic kidney disease. 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 (CCL2, the ligand of CCR2) 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.

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 plan to initiate a clinical endpoint study 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 SAEs 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 have 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 SOC, a RAAS inhibitor, plus placebo (control group), 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 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.

In 2017, we plan to initiate a controlled clinical trial of CCX140 in patients with FSGS. Reduction in proteinuria is widely considered as a beneficial outcome in the treatment of CKD including FSGS, and many experts regard the reduction of proteinuria as the likely registration endpoint for a new therapeutic in FSGS.

CCX140 Commercialization Strategy

We plan on building a sales infrastructure in the United States to commercialize our rare and orphan disease product 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 of the United States our partner Vifor will be responsible for the commercialization of CCX140.

In December 2016, we entered into a second collaboration and license agreement with Vifor, the CCX140 Agreement, pursuant to which we granted Vifor exclusive rights to commercialize CCX140 in rare renal diseases in markets outside the United States and China. We 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 non-refundable upfront commitment totaling \$50 million and will 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 chronic kidney disease, or CKD. Should Vifor later exercise the CKD option, we would receive co-promotion rights in CKD in the United States.

Early Stage Compounds

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 2014, approximately 46,000 people are expected to develop pancreatic cancer, and 40,000 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, or KRAS, 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 the human 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 SAEs 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. We will continue to follow the patients to assess overall survival later in 2017.

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.

Other Therapeutic Areas

Also in our CCR2 program, in October 2016, we announced the presentation of data from 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, with CCX872, a selective orally administered inhibitor of the chemokine receptor known as CCR2, at the October 2016 American College of Gastroenterology Annual Meeting. CCX872 demonstrated significant reductions in liver fibrosis 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

Inflammatory Bowel Disease, or IBD/Crohn's Disease and Ulcerative Colitis

IBD refers to two diseases — Crohn's disease and ulcerative colitis — both characterized by inflammation of the gastrointestinal tract. Crohn's disease can cause inflammation in any part of the digestive tract but often affects the tail end of the small intestine. Ulcerative colitis is inflammation of the large intestine. Both Crohn's disease and ulcerative colitis are chronic and recurring inflammatory conditions. Researchers believe that these conditions occur when the body's inflammatory cells become over-reactive and mount a destructive inflammatory response. Current treatments for IBD include steroids, 5-aminosalicylic acids, immunosuppressive therapies, such as azathioprine or biologic agents such as TNF- α inhibitors and integrin inhibitors, such as the anti- $\alpha 4\beta 7$ antibody, vedolizumab, and when all else fails, surgery.

Vercirnon

Our drug candidate vercirnon is intended to control the inflammatory response underlying IBD by targeting the chemokine receptor known as CCR9. In adults, CCR9 is found primarily on a population of T cells, a subset of the body's inflammatory cells, which migrate selectively to the digestive tract. It is believed that when CCR9's ligand, CCL25 (also known as TECK), is over-expressed, the migration of T cells to the small and large intestine causes persistent inflammation that may result in Crohn's disease or ulcerative colitis.

We completed nine clinical trials with vercirnon in a total of 785 subjects, including five Phase I clinical trials (three in the United States and two in the United Kingdom), one Thorough QT study in the United States (an assessment of cardiovascular safety which is required for regulatory approval), and three Phase II clinical trials (one in the Netherlands, the United Kingdom, and the United States, one in Finland and one (PROTECT-1) in Australia, Austria, Belgium, Brazil, Bulgaria, Canada, the Czech Republic, Denmark, France, Germany, Hungary, Israel, the Netherlands, Poland, South Africa, Sweden and the United Kingdom). We completed our PROTECT-1 Phase II clinical trial in 436 patients with moderate-to-severe Crohn's disease in 2009. Results from this clinical trial indicated that vercirnon was effective in inducing a clinical response over a 12-week treatment

period. The results also indicated that vercirnon was effective in maintaining clinical remission over an additional 36-week treatment period. Vercirnon was safe and well-tolerated in all clinical trials completed to date. Data from the first of four Phase III clinical trials, the SHIELD-1 study, conducted by our former collaboration partner, GSK, investigating vercirnon in 608 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. Results from SHIELD-4, a second induction study showed higher Crohn's disease activity index, or CDAI, response and remission rates with 500mg vercirnon twice daily compared to 500mg once daily. This is in contrast to the lack of dose response observed in the SHIELD-1 trial, and the response observed in the population of patients who completed 12 week dosing in SHIELD-4 was similar to the positive results from the Phase IIb PROTECT-1 clinical trial conducted by us. We are evaluating a potential future development and funding strategy for vercirnon. The assets, including vercirnon, all clinical study data, and IND are wholly owned by us.

Under the prior development and commercialization agreement with GSK which ended in 2013, we are subject to reverse royalties to GSK of 3% on annual worldwide net sales of vercirnon only if a regulatory agency were to deem one of GSK's SHIELD trials to be a pivotal Phase III clinical trial.

Second Generation CCR9 Inhibitor CCX507

Also in the area of IBD, our drug candidate CCX507 builds on our expertise in the area of CCR9 inhibitors and IBD. CCX507 is a second generation CCR9 inhibitor. CCX507 is selective for CCR9 relative to all other chemokine receptors, orally bioavailable, and has an excellent preclinical safety profile. CCX507 has a greater potency towards CCR9 than vercirnon. We completed Phase I clinical development, which demonstrated that CCX507 was safe and well-tolerated, and blocked CCR9 on circulating leukocytes. Additionally, preclinical data of CCX507 in combination with an anti- $\alpha 4\beta 7$ antibody or anti-TNF showed that combined treatment reduced the severity of colitis better than monotherapy with either drug alone. We plan to move CCX507 forward to Phase II clinical trials, potentially in conjunction with a strategic partner.

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.

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.

Recent work by others in the field has also revealed potential roles for CCR6 inhibitors in the treatment of colorectal cancer, or CRC. Specifically, in a mouse CRC model, mice that are genetically deficient in CCR6, as well as wild-type mice treated with a CCR6 inhibitor develop fewer intestinal polyps.

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 710 issued or allowed patents and approximately 220 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. There are approximately 22 issued or allowed patents and 24 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 70 issued or allowed patents and 12 patents pending worldwide relating to their chemical composition or use thereof. With respect to our drug candidates in the CCR9 and CCR1 programs, we have approximately 372 issued or allowed patents and 120 patents pending worldwide relating to their chemical composition or use thereof. We have approximately 100 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs. We have approximately 37 issued patents 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, Taiwan, Mexico and Singapore. 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 Canada and Brazil which, if issued, are anticipated to expire in 2029 (not including patent term extensions or supplementary protection certificates that may be available). We have patent applications pending covering certain synthetic methods related to making avacopan, which, if issued, are anticipated to expire in 2035.

CCX140 is covered by two issued patents in the United States for the composition-of-matter of CCX140 and pharmaceutical compositions thereof that will expire in 2029 and 2026, respectively (not including patent term extension that may be available to extend the term of the patents). CCX140 is also covered by two additional issued patents in the United States (covering certain methods of use) that will expire in 2028 and 2026 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 2028 and 2026, 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 Japan, Australia, Canada, China, Israel, Mexico and Hong Kong, covering CCX140 composition-of-matter. These issued patents will expire in 2028 or 2026 (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 India, South Korea and Brazil, 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 two issued patents 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 patents. CCX872 is covered by some issued patent in various countries including Europe, Australia, Canada, India, Hong-Kong, Japan, and Mexico that cover the composition of matter of CCX872 and will expire in 2026 not including a supplementary protection certificate that may be available to extend the term of the 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.

Millennium Pharmaceuticals, Inc., or Millennium, has obtained certain United States patents which include claims to small molecules that modulate CCR9, compositions thereof, and methods of using them to treat conditions such as IBD. Millennium may contend that the claims of these patents cover our patented vercirnon drug candidate. We believe that our activities related to vercirnon are currently exempt from patent infringement liability because these activities are strictly limited to obtaining information for regulatory approval. However, if and when our vercirnon related activities extend beyond those related to seeking regulatory approval, such as, for example, if and when we commercialize vercirnon, Millennium might then commence an infringement action against us based on these patents and/or other related patents that it may be granted in the future. If Millennium elects to sue us, we believe that we may have viable defenses to any such infringement suit. However, we cannot assure you that the relevant court would find in our favor with respect to such defenses.

Competition

We compete in the pharmaceutical, biotechnology and other related markets that address AAV, aHUS, DN and other renal diseases, IBD, rheumatoid arthritis, 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®).

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.

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.

Our CCR9 small molecule inhibitors such as CCX507 and vercirnon for the treatment of IBD, might compete against existing IBD treatments such as Remicade, Humira, and other TNF- α inhibitors, anti- α 4 β 7 antibodies such as vedolizumab (Entyvio), immunomodulatory drugs and steroids and potentially against other novel IBD drug candidates that are currently in development. Remicade is a humanized monoclonal antibody targeted to TNF- α , indicated for the treatment of Crohn's disease, ulcerative colitis, rheumatoid arthritis, psoriasis, psoriatic arthritis and ankylosing spondylitis. Humira, a similar drug, is also a human monoclonal antibody that acts as a TNF- α inhibitor. Marketed by AbbVie in the United States and Europe, Humira is approved for the treatment of Crohn's disease, ulcerative colitis, rheumatoid arthritis, psoriatic arthritis, juvenile idiopathic arthritis, plaque psoriasis, and ankylosing spondylitis. Biosimilar drugs to Humira, for example, may also become available to treat patients with IBD.

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 AbbVie, Alexion, Amgen, AstraZeneca, Biogen Idec, Bayer, Elan, GlaxoSmithKline, Johnson & Johnson, Merck, Merck Serono, Roche/Genentech, Takeda, Novartis, Pfizer, Sanofi 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 SOC. 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 Pfizer, GlaxoSmithKline, Bristol-Myers Squibb, Merck, Takeda, Sanofi, Incyte, Alexion, Allergan, Omeros, Dimerix, X4 Pharmaceuticals, 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 common chemical engineering and synthetic processes from readily available 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, regulations for use in our clinical trials. However, we currently rely on a single source supplier for our requirements of the API of each of these other drug candidates. The manufacture of pharmaceuticals 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 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 from time to time. If we were unable to obtain sufficient quantities of drug candidates or receive raw materials in a timely manner, we could be required to delay our ongoing clinical trials and seek alternative manufacturers, which would be costly and time-consuming. We believe we have multiple potential sources for our contract manufacturing.

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 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, and the data were submitted from the PROTECT-1 clinical trial in support of future U.S. marketing application for vercirnon. We are pursuing a similar development strategy for CCX140 which has recently 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 was conducted in North America. We have an open IND 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, 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.

Once an NDA has been accepted for filing, by law the FDA has 180 days to review and examine the application and respond to the applicant. However, the review process is often significantly extended by FDA requests for additional information or clarification. Under the Prescription Drug User Fee Act, the FDA has a

goal of responding to NDAs within ten months of the filing date for standard review, but this timeframe is also often extended. 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. 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.

A sponsor may also seek approval of its drug candidates under programs designed to accelerate 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, a fast track product 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 new 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 may also be eligible for "priority review," or review within a six month timeframe from the date a complete NDA is accepted for filing, if a sponsor shows that its drug candidate provides a significant improvement compared to marketed drugs. 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. We cannot predict whether any of our drug candidates will obtain such designations or approvals, or the ultimate impact, if any, of such designations or approvals on the timing or likelihood of FDA approval of any of our proposed drugs.

Drugs may be marketed only for the FDA approved indications and in accordance with the provisions of the approved labeling. Further, if there are any modifications to the drug, including changes in indications, labeling, or manufacturing processes or facilities, the applicant may be required to submit and obtain FDA approval of a new NDA or NDA supplement, which may require us to develop additional data or conduct additional preclinical studies and clinical trials.

Before approving an application, the FDA will inspect the facility or the facilities at which the finished drug product, and sometimes the active drug ingredient, is manufactured, and will not approve the drug unless cGMP

compliance is satisfactory. The FDA may also inspect the sites at which the clinical trials were conducted to assess their compliance, and will not approve the drug unless compliance with GCP requirements is satisfactory.

The testing and approval processes require substantial time, effort and financial resources, and each may take several years to complete. The FDA may not grant approval on a timely basis, or at all. Even if we believe a clinical trial has demonstrated safety and efficacy of one of our drug candidates for the treatment of a disease, the results may not be satisfactory to the FDA. Preclinical and clinical data may be interpreted by the FDA in different ways, which could delay, limit or prevent regulatory approval. We may encounter difficulties or unanticipated costs in our efforts to secure necessary governmental approvals which could delay or preclude us from marketing drugs. The FDA may limit the indications for use or place other conditions on any approvals that could restrict the commercial application of the drugs.

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 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 (1) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (2) 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 (3) 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.

Other Regulatory Requirements

Any drugs manufactured or distributed by us or our collaborators pursuant to FDA approvals would be subject to continuing regulation by the FDA, including recordkeeping requirements and reporting of adverse experiences associated with the drug. Drug manufacturers and their subcontractors are required to register their establishments with the FDA and certain state agencies, and are subject to periodic unannounced inspections by the FDA and certain state agencies for compliance with ongoing regulatory requirements, including cGMPs, which impose certain procedural and documentation requirements upon us and our third party manufacturers. Failure to comply with the statutory and regulatory requirements can subject a manufacturer to possible legal or regulatory action, such as warning letters, suspension of manufacturing, seizure of product, injunctive action or possible civil penalties. We cannot be certain that we or our present or future third party manufacturers or suppliers will be able to comply with the cGMP regulations and other ongoing FDA regulatory requirements. If we or our present or future third party manufacturers or suppliers are not able to comply with these requirements, the FDA may halt our clinical trials, require us to recall a drug from distribution or withdraw approval of the NDA for that drug.

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; and
- 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 CMS to test innovative payment and service delivery models to lower Medicare and Medicaid spending.

We expect that the new presidential administration and U.S. Congress will seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Since taking office, President Trump has continued to support the repeal of all or portions of the Affordable Care Act. In January 2017, the House and Senate passed a budget resolution that authorizes congressional committees to draft legislation to repeal all or portions of the Affordable Care Act and permits such legislation to pass with a majority vote in the Senate. President Trump has also recently issued an executive order in which he stated that it is his administration's policy to seek the prompt repeal of the Affordable Care Act and directed executive departments and federal agencies to waive, defer, grant exemptions from or delay the implementation of the provisions of the Affordable Care Act to the maximum extent permitted by law. 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;
- 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 third-party payors that are false or fraudulent;
- 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 Payment Sunshine Act, which requires manufacturers of drugs, devices, biologics and medical supplies for which payment is available under Medicare, Medicaid or the Children's Health Insurance Program (with certain exceptions) to report annually to the Centers for Medicare & Medicaid Services, or CMS, information related to payments or other "transfers of value" made to physicians (defined to include doctors, dentists, optometrists, podiatrists and chiropractors) and teaching hospitals, and applicable manufacturers and group purchasing organizations to report annually to CMS ownership and investment interests held by physicians (as defined above) and their immediate family members and payments or other "transfer of value" to such physician owners;
- the federal Health Insurance Portability and Accountability Act of 1996, as amended by the Health Information Technology for Economic and Clinical Health Act, which governs the conduct of certain electronic healthcare transactions and protects the security and privacy of protected health information;
- 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, 2016, we had 62 full-time employees, 27 of whom hold Ph.D.s, M.D.s or both. Of our total workforce, 47 employees are engaged in research and development, and 15 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 \$38.0 million, \$33.2 million and \$33.8 million in research and development in the years 2016, 2015, and 2014, 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 loss for the years ended December 31, 2016 and 2015 was \$40.0 million and \$47.3 million, respectively. As of December 31, 2016, we had an accumulated deficit of \$307.1 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. (“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, CCX140, avacopan, CCX872, vercirnon, CCX507 and CCX354 and 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 vercirnon 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, or EC, 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, avacopan, for example. 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 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 regulations and standards, commonly referred to as good clinical practices, or GCPs, 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, 2016, we had approximately \$123.8 million in cash, cash equivalents and investments, excluding the \$70.0 million upfront commitment in connection with the December 2016 CCX140 Agreement and 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 14, 2017. 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, 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.

We may not be able to obtain orphan drug exclusivity for avacopan for the treatment of AAV.

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 (1) it is intended for the diagnosis, prevention or treatment of a life-threatening or chronically debilitating condition; (2) 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 (3) 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 aHUS and AAV, including granulomatosis with polyangiitis or Wegener’s granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome. The European Commission granted orphan drug designation for avacopan for the treatment of granulomatosis with polyangiitis or Wegener’s granulomatosis and microscopic polyangiitis. 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 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, and we may seek to find a partner or alternative financing arrangements with respect to the completion of clinical development and commercialization of vercirnon. 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 new 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 (see "Item 1. Business — Our Proprietary Drug Discovery Platform, EnabaLink") 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 current good manufacturing practices, or 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, 2016, we had 62 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 will be 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 not limited to, AbbVie, Alexion, Amgen, AstraZeneca, Biogen Idec, Bayer, Elan, GlaxoSmithKline, Johnson & Johnson, Merck, Merck Serono, Roche/Genentech, Takeda, Novartis, Pfizer, Sanofi 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 SOC. 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 not limited to, Pfizer, GlaxoSmithKline, Bristol-Myers Squibb, Merck, Takeda, Sanofi, Incyte, Alexion, Allergan, Omeros, Dimerix, X4 Pharmaceuticals, 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, aHUS, FSGS, DN and other renal disease, IBD, 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 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, 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 an “emerging growth company” and our election to delay adoption of new or revised accounting standards applicable to public companies may result in our financial statements not being comparable to those of other public companies and we also are entitled to utilize other reduced disclosure and governance requirements applicable to emerging growth companies.

We are an “emerging growth company,” as defined in the Jumpstart Our Business Startups Act of 2012, or the JOBS Act, and we utilize certain exemptions from various reporting requirements that are applicable to other public companies that are not emerging growth companies including, but not limited to, not being required to provide the auditor attestation report otherwise required by Section 404 of the Sarbanes-Oxley Act, reduced disclosure obligations regarding executive compensation in our periodic reports and proxy statements, and exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved. In addition, Section 107 of the JOBS Act also provides that an emerging growth company can utilize the extended transition period provided in Section 7(a)(2)(B) of the Securities Act of 1933, as amended, or the Securities Act, for implementing new or revised accounting standards. In other words, an emerging growth company can delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have elected to delay such adoption of new or revised accounting standards, and as a result, we may not implement new or revised accounting standards on the relevant dates on which adoption of such standards is required for non-emerging growth companies. As a result of such election, our financial statements may not be comparable to the financial statements of other public companies. We cannot predict if investors will find our common stock less attractive because we will rely on these exemptions. If some investors find our common stock less attractive as a result, there may be a less active trading market for our common stock and our stock price may be more volatile. We may utilize these reporting exemptions until we are no longer an emerging growth company. We will remain an emerging growth company for up to five years following the year in which we completed our initial public offering, although if the market value of our common stock that is held by nonaffiliates exceeds \$700 million as of any June 30 before that time, we would cease to be an emerging growth company as of the following December 31.

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. Once we are no longer an emerging growth company as defined in the JOBS Act, we will also need 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 in a timely manner or with adequate compliance, we may be subject to sanctions or investigation by regulatory authorities, such as the Securities and

Exchange Commission, or 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 that occurred in July 1999 and June 2004, 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.

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

We may have to license rights from Millennium Pharmaceuticals, Inc. or engage in patent litigation in order to secure the rights necessary to commercialize vercirnon. Patent litigation could absorb significant management time and financial resources, and, if we do not prevail, could have a material adverse effect on our ability to derive revenues from vercirnon.

Millennium Pharmaceuticals, Inc., or Millennium, has obtained certain U.S. patents which include claims to small molecules that modulate CCR9, compositions thereof, and methods of using them to treat conditions such as IBD. We became aware of Millennium’s CCR9-related patent applications during our own routine patent and

patent literature review. Millennium, which was acquired by Takeda Pharmaceutical Company Limited, or Takeda, in May 2008 and is currently a wholly owned subsidiary of Takeda, may contend that the claims of these patents cover our patented vercirnon drug candidate. We believe that our activities related to vercirnon are currently exempt from patent infringement liability because these activities are strictly limited to obtaining information for regulatory approval. However, if and when our vercirnon related activities extend beyond those related to seeking regulatory approval, such as, for example, if and when we commercialize vercirnon, Millennium might then commence an infringement action against us based on these patents and/or other related patents that it may be granted in the future. If Millennium elects to sue us, we believe that we may have viable defenses to any such infringement suit. However, we cannot assure you that the relevant court would find in our favor with respect to such defenses. 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. During 2005, we did engage in preliminary discussions with Millennium regarding potentially collaborating with respect to CCR9, given that both we and Millennium have patents relating to CCR9. However, these discussions were general in nature and did not progress beyond the preliminary stage. In addition, in April 2012, an opposition was filed with the European Patent Office by Millennium with respect to one of our patents relating to broad genus claims describing small molecules that target CCR9, the scope of which also relates to vercirnon. The opposition filed by Millennium alleged that the subject matter of such patent is not novel; such patent does not involve an inventive step; such patent does not sufficiently disclose the invention and the subject matter of such patent extends beyond the content of its patent application. In October 2013, the Opposition Division of the European Patent Office verbally announced a decision against the opposition filed by Millennium, with a written decision following in January 2014. In this decision, the patent was maintained in amended form. Millennium did not appeal this decision. Thus, the decision to maintain the patent became final in April 2014. The patent has meanwhile been revalidated in all designated states. It can now only be invalidated nationally by filing a nullity action before a national court.

We may also be subject to negative publicity due to litigation. Pending or future patent litigation against us or any strategic partners by Millennium or anyone else 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. If Millennium is able to obtain an injunction and neither we nor our strategic partners are able to obtain a license, both we and our strategic partners would be precluded from the manufacture and sale of vercirnon. U.S. patents are entitled to a presumption of validity and the burden of proving invalidity would be heavily weighted against us. Specifically, we would be required to show by clear and convincing evidence that Millennium's patents are invalid. Such decisions on patent validity often favor the patent owner because of the presumption of validity. If we or our strategic partners are unable to show that Millennium's patent is invalid and neither we nor our strategic partners are able to obtain a license from Millennium for the use of their intellectual property at all or on commercially acceptable terms, this would preclude both us and our strategic partners from the manufacture and sale of vercirnon or related candidate compounds found to be covered by Millennium's patent claims.

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 710 issued or allowed patents and approximately 220 pending patent applications, with claims relating to all of our current clinical-stage drug candidates. There are approximately 22 issued or allowed patents and 24 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 70 issued or allowed patents and 12 patents pending worldwide relating to their chemical composition or use thereof. With respect to our drug candidates in the CCR9 and CCR1 programs, we have approximately 372 issued or allowed patents and 120 patents pending worldwide relating to their chemical composition or use thereof. We have approximately 100 patents issued or pending for our other preclinical-stage compounds in the C5aR, CCR2, CXCR7, CCR4, CXCR2 and CCR6 programs. We have approximately 37 issued patents 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 and 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. We have not submitted an application for or received marketing approval for any of our drug candidates. Obtaining approval of an NDA can be a lengthy, expensive and uncertain process. In addition, failure to comply with FDA and other applicable U.S. 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.

Prior to receiving approval to commercialize any of our drug candidates in the United States or abroad, we must demonstrate with substantial evidence from well controlled clinical trials, and to the satisfaction of the FDA 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 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 or other regulatory authorities denying approval of our drug candidates for any or all targeted indications.

Regulatory approval of an NDA or NDA supplement is not guaranteed, and the approval process is expensive and may take several years. The FDA also has 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 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 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 officials may not find the data from preclinical studies and clinical trials sufficient;
- the FDA might not approve our or our third party manufacturer's processes or facilities; or
- the FDA 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 and/or non-U.S. 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 also has authority to require a risk evaluation and mitigation strategy, or REMS, as part of an NDA 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 and/or non-U.S. regulatory authorities approve any of our drug candidates, we will be subject to extensive and ongoing regulatory requirements by the FDA and other regulatory authorities with regard to the labeling, packaging, adverse event reporting, storage, advertising, promotion and recordkeeping for our products. The FDA strictly regulates 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 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 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 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 and/or other non-U.S. 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, President Obama signed into law the 21st Century Cures Act, which 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 or in other countries. 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. Notably, on January 23, 2017, President Trump ordered a hiring freeze for all executive departments and agencies, including the FDA, which prohibits the FDA from filling employee vacancies or creating new positions. Under the terms of the order, the freeze will remain in effect until implementation of a plan to be recommended by the Director for the Office of Management and Budget, or OMB, in consultation with the Director of the Office of Personnel Management, to reduce the size of the federal workforce through attrition. Although certain positions at the FDA may ultimately be exempted from the freeze, an under-staffed FDA could result in delays in the FDA's responsiveness or in its ability to review submissions or applications, issue regulations or guidance or implement or enforce regulatory requirements in a timely fashion or at all. Moreover, on January 30, 2017, President Trump issued an Executive Order, applicable to all executive agencies, including the FDA, that requires that for each notice of proposed rulemaking or final regulation to be issued in fiscal year 2017, the agency shall identify at least two existing regulations to be repealed, unless prohibited by law. These requirements are referred to as the "two-for-one" provisions. This Executive Order includes a budget neutrality provision that requires the total incremental cost of all new regulations in the 2017 fiscal year, including repealed regulations, to be no greater than zero, except in limited circumstances. For fiscal years 2018 and beyond, the Executive Order requires agencies to identify regulations to offset any incremental cost of a new regulation and approximate the total costs or savings associated with each new regulation or repealed regulation. In interim guidance issued by the Office of Information and Regulatory Affairs within OMB on February 2, 2017, the administration indicates that the "two-for-one" provisions may apply not only to agency regulations, but also to significant agency guidance documents. It is difficult to predict how these requirements 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 of the European Medicines Agency, or 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.

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, President Obama signed into law the Affordable Care Act. 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 new presidential administration and U.S. Congress will seek to modify, repeal or otherwise invalidate all, or certain provisions of, the Affordable Care Act. Since taking office, President Trump has continued to support the repeal of all or portions of the Affordable Care Act. In January 2017, the House and Senate passed a budget resolution that authorizes congressional committees to draft legislation to repeal all or portions of the Affordable Care Act and permits such legislation to pass with a majority vote in the Senate. President Trump has also recently issued an executive order in which he stated that it is his administration’s policy to seek the prompt repeal of the Affordable Care Act and directed executive departments and federal agencies to waive, defer, grant exemptions from or delay the implementation of the provisions of the Affordable Care Act to the maximum extent permitted by law. 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 new 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.

If we fail to comply with healthcare regulations, we could face substantial penalties and our business, operations and financial condition could be adversely affected.

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;
- 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 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 Health Insurance Portability and Accountability Act of 1996, as amended by the Health Information Technology for Economic and Clinical Health Act, which governs the conduct of certain electronic healthcare transactions and protects the security and privacy of protected health information; 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.

Further, there has been a trend in the increase of federal and state laws and regulations regarding payments and other transfers of value paid to physicians and other healthcare providers and entities. The Open Payments Sunshine Act and state law equivalents impose requirements to report certain financial arrangements with physicians and others, including reporting any transfers of value made or distributed to prescribers and other healthcare providers and reporting any investment interests held by physicians and certain during each calendar year. The laws apply to certain applicable manufacturers, which are required to submit reports to the government by the 90th day of each calendar year. 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, 2016, the price per share for our common stock on the Nasdaq Global Select Market ranged from a low sale price of \$1.92 to a high sale price of \$9.10. 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, CCX507, 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 including, if necessary, those relating to vercirnon and other CCR9 drug candidates;
- 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 March 3, 2017, our officers and directors, together with holders of 5% or more of our outstanding common stock and their respective affiliates, beneficially owned approximately 60% 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, 2016, we had 48,057,920 shares of common stock outstanding. Of these shares, approximately 28,540,100 are freely tradeable, without restriction, in the public market. In addition, approximately 17,707,763 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 holders of shares of our common stock, warrants to purchase our capital stock and the shares of common stock issuable upon exercise of those warrants are entitled to rights with respect to the registration of their shares under the Securities Act. Registration of these shares under the Securities Act would result in the shares becoming freely tradable without restriction under the Securities Act, except for shares purchased by affiliates. In addition, 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 \$4.7 million for severance and other benefits and acceleration of vesting of stock options with an intrinsic value of \$10.9 million as of December 31, 2016 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. We currently intend to retain all available funds and any future earnings to fund the development and growth of our business. In addition, any future debt financing arrangement may contain 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 2017, 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.

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 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
<i>Fiscal 2015</i>		
First Quarter	\$9.20	\$6.51
Second Quarter	9.46	6.50
Third Quarter	9.19	5.40
Fourth Quarter	8.30	5.46

Holders of Common Stock

As of March 3, 2017, there were approximately 51 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.

Equity Compensation Plan Information

The following table summarizes securities available under our equity compensation plans as of December 31, 2016.

<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⁾	9,785,859	\$7.72	1,882,080
Equity compensation plans not approved by security holders:	—	—	—
Total	<u>9,785,859</u>	<u>\$7.72</u>	<u>1,882,080</u>

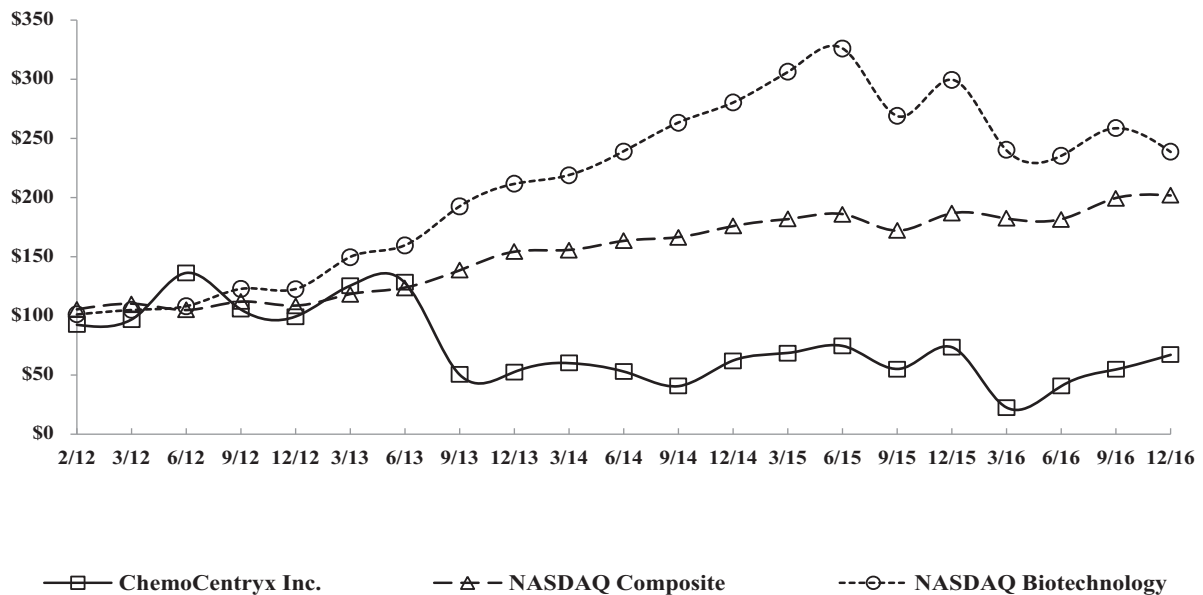
- (1) Consists of our Amended and Restated 1997 Stock Option/Stock Issuance Plan, our Amended and Restated 2002 Equity Incentive Plan and our 2012 Equity Incentive Award Plan, our Non-Employee Director Compensation Policy and our Employee Stock Purchase Plan, or ESPP.
- (2) Includes 9,345,515 shares subject to outstanding stock option and 440,344 shares subject to outstanding restricted stock unit as of December 31, 2016.
- (3) Calculated exclusive of outstanding restricted stock unit awards.
- (4) Of these shares, 1,655,524 shares were available for stock option awards and restricted stock units and restricted stock awards, and 226,556 were available for the ESPP, in each case as of December 31, 2016.

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 February 8, 2012 (the date our common stock commenced trading on the NASDAQ Global Select Market) through December 31, 2016 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 59 MONTH CUMULATIVE TOTAL RETURN*
Among ChemoCentryx Inc., the NASDAQ Composite Index and the NASDAQ Biotechnology Index



*\$100 invested on 2/8/12 in stock or 1/31/12 in index, including reinvestment of dividends.
Fiscal year ending December 31.

	2/12	3/12	6/12	9/12	12/12	3/13	6/13	9/13	12/13	3/14	6/14	9/14	12/14	3/15	6/15	9/15	12/15	3/16	6/16	9/16	12/16
ChemoCentryx Inc.	92.73	96.91	136.36	105.73	99.45	125.64	128.55	50.55	52.64	60.27	53.18	40.91	62.09	68.64	74.82	55.00	73.64	22.64	40.82	54.91	67.27
NASDAQ Composite	105.54	110.21	105.19	112.19	108.63	118.37	123.84	138.77	154.40	155.60	163.56	166.44	176.07	182.07	185.92	172.24	186.92	182.53	181.74	199.49	202.05
NASDAQ Biotechnology	101.49	105.09	108.24	122.92	122.74	149.68	159.63	192.65	211.78	218.97	239.08	263.44	280.42	306.46	326.19	269.19	299.63	240.48	235.37	258.77	238.86

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.

	Years Ended December 31,				
	2016	2015	2014	2013	2012
(in thousands except share and per share data)					
Consolidated Statement of Operations Data:					
Revenues:					
Collaboration and license revenue	\$ 11,435	\$ —	\$ —	\$ 6,060	\$ 5,419
Grant revenue	500	—	—	—	—
Total revenue	11,935	—	—	6,060	5,419
Operating expenses:					
Research and development	37,945	33,183	33,815	33,541	34,569
General and administrative	14,710	14,506	13,584	11,634	10,480
Total operating expenses	52,655	47,689	47,399	45,175	45,049
Loss from operations	(40,720)	(47,689)	(47,399)	(39,115)	(39,630)
Interest income	757	384	494	501	533
Interest expense	—	—	(24)	(59)	(794)
Net loss	\$ (39,963)	\$ (47,305)	\$ (46,929)	\$ (38,673)	\$ (39,891)
Basic and diluted net loss per share ⁽¹⁾	\$ (0.86)	\$ (1.08)	\$ (1.08)	\$ (0.95)	\$ (1.13)
Shares used to compute basic and diluted net loss per share	46,431,501	43,889,677	43,275,276	40,916,138	35,406,922

(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 loss per share.

	As of December 31,				
	2016	2015	2014	2013	2012
(in thousands)					
Consolidated Balance Sheet Data:					
Cash, cash equivalents and investments	\$ 123,761	\$ 76,289	\$ 114,620	\$ 149,874	\$ 118,956
Working capital	110,356	66,541	66,139	127,430	93,180
Total assets	155,872	78,155	116,981	152,422	122,323
Non-current equipment financing obligations	—	—	—	16	379
Accumulated deficit	(307,059)	(267,096)	(219,791)	(172,862)	(134,189)
Total stockholders’ equity	49,889	72,507	108,606	145,308	110,346

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 focuses on a specific chemoattractant receptor that selectively blocks its negative inflammatory or suppressive response, 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 drugs 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 us manage the wide array of opportunities, we have segmented our pipeline into early stage and late stage compounds.

Late Stage Compounds

We have chosen to focus initially on kidney disease, particularly on rare indications, where orphan 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 and Rare Diseases

Avacopan (CCX168) is an orally-administered complement inhibitor targeting the C5a receptor, or C5aR, and is being developed for orphan and rare diseases, including 1) anti-neutrophil cytoplasmic auto-antibody associated vasculitis, or AAV, a devastating autoimmune disease that destroys blood vessels and can lead to kidney failure; 2) atypical hemolytic uremic syndrome, or aHUS, a rare, life threatening disease; and 3) complement 3 glomerulopathy, or C3G, a debilitating kidney disease.

Avacopan has been granted orphan drug designation by the U.S. Food and Drug Administration, or FDA, for the treatment of AAV and aHUS and by the European Medicines Agency, or EMA for the treatment of 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 accelerate its marketing authorization.

Following completion of two Phase II clinical trials in patients with AAV, the results of which demonstrated that avacopan was safe, 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 300 patients in up

to 200 sites in the United States and Europe. We also plan to initiate clinical endpoint trials, ones that could potentially serve as registration trials, of avacopan for the treatment of patients with C3G and aHUS in 2017.

CCX140 — Chronic and Rare Kidney Diseases

CCX140 is 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 Phase II clinical trial of CCX140 in patients with DN met its primary endpoint by demonstrating that CCX140 given orally once daily added to an SOC renin-angiotensin-aldosterone system inhibitor treatment resulted in a statistically significant reduction in proteinuria, beyond that achieved with SOC alone. Based on the safety and efficacy data related to reduction in proteinuria and improvement of renal function observed in the Phase II trial in DN, we plan to initiate in 2017 a clinical endpoint trial of CCX140 for the treatment of patients with FSGS, for which there are currently no FDA-approved treatments.

Global Kidney Health Alliance with Vifor

In May 2016 we announced a partnership with Vifor (International) Ltd., or Vifor, a European-based world leader specializing in kidney disease, for the commercial rights to avacopan in Europe and certain other international markets, the Avacopan Agreement. We expanded our partnership with Vifor in December 2016 with an additional deal for our other late stage drug candidate, CCX140, whereby we granted Vifor worldwide rights outside of the United States and China; and in February 2017, we announced a further deal with Vifor that harmonized the geographic commercialization rights underlying the agreements for both drug candidates.

We have secured \$155 million in upfront cash payments and commitments, plus substantial potential milestone payments pursuant to our agreements with Vifor. Through our alliance, we maintain the commercial rights of avacopan and CCX140 in the United States and China, and also retain control of the clinical development programs for rare renal disease. Vifor gains the commercial rights for all other international markets, and will pay us double-digit tiered royalties 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 Compounds

While the science has led us to focus initially on kidney disease, our targeted blocking system 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 from other areas of inflammatory and autoimmune disorders, such as rheumatoid arthritis, inflammatory bowel disease, and psoriasis, as well as cancer, where our drug candidate CCX872 has shown promise in a Phase I trial for non-operable pancreatic cancer. 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, 2016, we had an accumulated deficit of \$307.1 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.

JOBS Act

In April 2012, the JOBS Act was enacted. Section 107 of the JOBS Act provides that an emerging growth company can utilize the extended transition period provided in Section 7(a)(2)(B) of the Securities Act for implementing new or revised accounting standards. In other words, an emerging growth company can delay the adoption of certain accounting standards until those standards would otherwise apply to private companies. We have elected to delay such adoption of new or revised accounting standards, and as a result, we may not implement new or revised accounting standards on the relevant dates on which adoption of such standards is required for other companies.

Subject to certain conditions set forth in the JOBS Act, as an emerging growth company, we intend to rely on certain of these exemptions, including without limitation, providing an auditor's attestation report on our system of internal controls over financial reporting pursuant to Section 404 and implementing any requirement that may be adopted regarding mandatory audit firm rotation or a supplement to the auditor's report providing additional information about the audit and the financial statements (auditor discussion and analysis). These exemptions will apply for a period of five years following the completion of our IPO although if the market value of our common stock that is held by nonaffiliates exceeds \$700 million as of any June 30 before that time, we would cease to be an emerging growth company as of the following December 31.

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 at the end of 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. Upfront fees 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) relates 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.

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 study 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 \$8.5 million, \$9.0 million and \$8.2 million for the years ended December 31, 2016, 2015, and 2014, respectively. At December 31, 2016 and 2015, we had \$9.6 million and \$12.6 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 2.37 years and 2.36 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, 2016, our revenue was 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 2014 and 2015.

Total revenue were as follows (in thousands):

	<u>Year Ended December 31,</u>		
	<u>2016</u>	<u>2015</u>	<u>2014</u>
Collaboration and license revenue	\$11,435	\$—	\$—
Grant revenue	500	—	—
Total revenue	<u>\$11,935</u>	<u>\$—</u>	<u>\$—</u>
Dollar increase	\$11,935	\$—	\$
Percentage increase	100%	—	

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):

	<u>Year Ended December 31,</u>		
	<u>2016</u>	<u>2015</u>	<u>2014</u>
Research and development expenses	\$37,945	\$33,183	\$33,815
Dollar (decrease) increase	\$ 4,762	\$ (632)	
Percentage (decrease) increase	14%	-2%	

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 Phase II clinical trials for avacopan for the treatment of AAV in 2016.

The decrease in research and development expense from 2014 to 2015 was primarily attributable lower Phase I clinical development expenses, partially offset by increases in research and drug discovery expenses and Phase II clinical expenses. The decrease in Phase I clinical development expenses was primarily due to the completion of Phase I clinical development of CCX507, our second generation CCR9 inhibitor, in 2014, which was partially offset by an increase in 2015 in expenses associated with the continued patient enrollment in the Phase I clinical study in CCX872, our second generation CCR2 inhibitor, for advanced pancreatic cancer. An increase in expenses in 2015 due to the continued patient enrollment of Phase II CLEAR and CLASSIC trials for avacopan and Phase II pilot clinical studies for avacopan in aHUS and IgAN, was nearly offset by a decrease in Phase II clinical expense due to the completion of our Phase II clinical trial in CCX140 for DN.

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

	<u>Year Ended December 31,</u>		
	<u>2016</u>	<u>2015</u>	<u>2014</u>
Phase I	\$ 5,959	\$ 3,686	\$ 4,783
Phase II	10,866	16,151	16,063
Phase III	8,019	—	—
Research and drug discovery	13,101	13,346	12,969
Total R&D	<u>\$37,945</u>	<u>\$33,183</u>	<u>\$33,815</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, CCX872 and vercirnon.

General and administrative expenses

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

	Year Ended December 31,		
	2016	2015	2014
General and administrative expenses	\$14,710	\$14,506	\$13,584
Dollar increase	\$ 204	\$ 922	
Percentage increase	1%	7%	

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

The increase from 2014 to 2015 was primarily due to increases in stock based compensation expense for stock option grants and restricted stock unit awards, intellectual property related expenses, and professional fees. Further, travel expenses and professional fees relating to our business development efforts also contributed to the increase.

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, legal and accounting related fees, and expenses associated with preparing to meet the requirements pursuant to the Sarbanes-Oxley Act of 2002, including in connection with the expiration of our status as an emerging growth company, expected to occur in 2017.

Other income (expense)

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

	Year Ended December 31,		
	2016	2015	2014
Interest income	\$757	\$384	\$494
Interest expense	—	—	(24)
Total other income (expense), net	<u>\$757</u>	<u>\$384</u>	<u>\$470</u>
Dollar increase (decrease)	\$373	\$ (86)	
Percentage increase (decrease)	97%	-18%	

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

The decrease in total other income, net from 2014 to 2015 was primarily due to a decrease in interest income earned on lower cash balances, which was partially offset by a decrease in interest expense as a result of full repayment of our equipment financing debt in the fourth quarter of 2014.

Liquidity and Capital Resources

As of December 31, 2016, we had approximately \$123.8 million in cash, cash equivalents and investments. Such amounts exclude the \$70.0 million upfront commitment in connection with the December 2016 CCX140 Agreement and the February 2017 amendment to the Avacopan Agreement. The following table shows a summary of our cash flows for each of the three years ended December 31, 2016, 2015, and 2014 (in thousands):

	Year Ended December 31,		
	2016	2015	2014
Cash provided by (used in)			
Operating activities	\$ 39,145	\$(39,327)	\$(34,315)
Investing activities	(48,764)	33,884	38,339
Financing activities	8,820	2,191	1,793

Operating activities. 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, which included \$66.6 million of deferred revenue in connection with the Avacopan Agreement, as well as a lower net loss. Net cash used by operating activities increased to \$39.3 million for the year ended December 31, 2015, from \$34.3 million for the same period in 2014 due primarily to changes in working capital items and a higher 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 \$8.8 million for the year ended December 31, 2016, which was primarily due to the receipt of \$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 compared to \$1.8 million for the same period in 2014. Net cash provided by financing activities for the years presented also included proceeds from the exercise of stock options and purchases from contributions to our 2012 Employee Stock Purchase Plan.

As of December 31, 2016, we had approximately \$123.8 million in cash, cash equivalents and investments, excluding the \$70.0 million upfront commitment in connection with the December 2016 CCX140 Agreement and 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 14, 2017. 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, 2016 (in thousands).

	Payments Due by Period				
	Total	Less than One Year	1-3 Years	3-5 Years	More than 5 Years
Operating lease ⁽¹⁾	\$2,167	\$915	\$1,252	\$—	\$—
Total contractual obligations	\$2,167	\$915	\$1,252	\$—	\$—

(1) We lease our facility in Mountain View, California. The lease expires in 2019.

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

In August 2014, the Financial Accounting Standards Boards, or FASB, issued Accounting Standards Updates, or ASU, No. 2014-15 (Subtopic 205-40)—Presentation of Financial Statements—Going Concern: Disclosure of Uncertainties about an Entity’s Ability to Continue as a Going Concern, or ASU 2014-15, which provides guidance about management’s responsibility to evaluate whether or not there is substantial doubt about our ability to continue as a going concern and to provide related footnote disclosure. ASU 2014-15 is effective for us for the year ending December 31, 2016. Early application is permitted. The adoption of this standard did not to have an impact on our financial statements.

In May 2015, the FASB issued a comprehensive new standard on revenue from contracts with customers. The 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 us 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.

We currently plan to adopt the accounting standard update on January 1, 2018, using the modified retrospective approach. The cumulative effect of adopting the accounting standard update will be recorded to retained earnings on January 1, 2018. We are currently at the early stages of analyzing our collaboration agreements to determine the differences in the accounting treatment under ASU 2014-09 compared to the current accounting treatment. During 2016, We entered into two license and collaboration agreements. We have primarily derived our revenues from license and collaboration agreements. The consideration we are eligible to receive under these agreements includes of upfront payments, research and development funding, milestone payments, and royalties. Each license and collaboration agreement is unique and will need to be assessed separately under the five-step process under the new standard. The new revenue recognition standard differs from the current accounting standard in many respects, such as in the accounting for variable considerations and the measurement of progress toward completion of performance obligations. While we have not completed an assessment of the impact of adoption, the adoption of ASU 2014-09 may have a material effect on our financial statements.

In February 2016, the FASB issued a new standard that 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 us on January 1, 2019. We are currently evaluating the impact of the adoption of this standard on its financial statements. However, we expect the adoption of our accounting guidance to result in an increase in lease assets and a corresponding increase in lease liabilities on our balance sheets.

In March 2016, the FASB issued guidance that changes the accounting for certain aspects of share-based payments to employees. The guidance requires the recognition of the income tax effects of awards in the income statement when the awards vest or are settled, thus eliminating additional paid in capital pools. The guidance also allows for the employer to repurchase more of an employee’s shares for tax withholding purposes without triggering liability accounting. In addition, the guidance allows for a policy election to account for forfeitures as they occur rather than on an estimated basis. The guidance is effective in 2017 with early adoption permitted. We adopted this new standard on January 1, 2017 and do not expect a material impact on our financial statements given the full valuation allowance position on its deferred tax assets.

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-25.

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, 2016, 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, 2016, 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, 2016 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, 2016.

Attestation Report of the Registered Public Accounting Firm

This Annual Report on Form 10-K does not include an attestation report of our registered public accounting firm due to an exemption established by the JOBS Act for “emerging growth companies”.

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, 2016, 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, or the Definitive Proxy Statement, to be filed with the Securities and Exchange Commission in connection with our 2017 Annual Meeting of Stockholders, which is expected to be filed not later than 120 days after the end of our fiscal year ended December 31, 2016, 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

Information required by this item will be contained in our Definitive Proxy Statement under the headings “Security Ownership of Certain Beneficial Owners and Management,” and is incorporated herein by reference.

Item 13. Certain Relationships, 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 report thereon of Ernst & Young LLP, an independent registered public accounting firm, are included in this Annual Report on Form 10-K:

	<u>Page</u>
Report of Independent Registered Public Accounting Firm	F-2
Audited Consolidated Financial Statements	
Consolidated Balance Sheets	F-3
Consolidated Statements of Operations	F-4
Consolidated Statements of Comprehensive Loss	F-5
Consolidated Statements of Stockholders' Equity	F-6
Consolidated Statements of Cash Flows	F-7
Notes to Consolidated Financial Statements	F-8

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 following the signature page of this Annual Report on Form 10-K, and is incorporated herein by reference.

[THIS PAGE INTENTIONALLY LEFT BLANK]

ChemoCentryx, Inc.

**Consolidated Financial Statements
As of December 31, 2016 and 2015
and for each of the three years in the period ended December 31, 2016**

Contents

	<u>Page</u>
Report of Independent Registered Public Accounting Firm	F-2
Audited Consolidated Financial Statements	
Consolidated Balance Sheets	F-3
Consolidated Statements of Operations	F-4
Consolidated Statements of Comprehensive Loss	F-5
Consolidated Statements of Stockholders' Equity	F-6
Consolidated Statements of Cash Flows	F-7
Notes to Consolidated Financial Statements	F-8

Report of Independent Registered Public Accounting Firm

The Board of Directors and Stockholders of ChemoCentryx, Inc.

We have audited the accompanying consolidated balance sheets of ChemoCentryx, Inc. as of December 31, 2016 and 2015, and the related consolidated statements of operations, comprehensive loss, stockholders' equity, and cash flows for each of the three years in the period ended December 31, 2016. 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 conducted our audits in accordance with the standards of the Public Company Accounting Oversight Board (United States). Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement. We were not engaged to perform an audit of the Company's internal control over financial reporting. Our audits included consideration of internal control over financial reporting as a basis for designing audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control over financial reporting. Accordingly, we express no such opinion. An audit also includes examining, on a test basis, evidence supporting the amounts and disclosures in the financial statements, assessing the accounting principles used and significant estimates made by management, and evaluating the overall financial statement presentation. We believe that our audits provide a reasonable basis for our opinion.

In our opinion, the financial statements referred to above present fairly, in all material respects, the consolidated financial position of ChemoCentryx, Inc. at December 31, 2016 and 2015, and the consolidated results of its operations and its cash flows for each of the three years in the period ended December 31, 2016, in conformity with U.S. generally accepted accounting principles.

/s/ Ernst & Young LLP

Redwood City, California
March 14, 2017

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

	December 31, 2016	December 31, 2015
Assets		
Current assets:		
Cash and cash equivalents	\$ 12,024	\$ 12,823
Short-term investments	105,740	58,455
Accounts Receivable	30,205	—
Prepaid expenses and other current assets	722	757
Total current assets	148,691	72,035
Property and equipment, net	905	949
Long-term investments	5,997	5,011
Other assets	279	160
Total assets	\$ 155,872	\$ 78,155
Liabilities and Stockholders' Equity		
Current liabilities:		
Accounts payable	\$ 671	\$ 675
Accrued liabilities	8,645	4,819
Deferred revenue	29,019	—
Total current liabilities	38,335	5,494
Deferred revenue	67,547	—
Other non-current liabilities	101	154
Total liabilities	105,983	5,648
Stockholders' equity:		
Preferred stock:		
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 at December 31, 2016 and December 31, 2015; 48,057,920 shares and 44,185,506 shares issued and outstanding at December 31, 2016 and December 31, 2015, respectively.	48	44
Additional paid-in capital	356,966	339,615
Note receivable	(16)	(16)
Accumulated other comprehensive loss	(50)	(40)
Accumulated deficit	(307,059)	(267,096)
Total stockholders' equity	49,889	72,507
Total liabilities and stockholders' equity	\$ 155,872	\$ 78,155

See accompanying notes.

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

	Year Ended December 31,		
	2016	2015	2014
Revenue:			
Collaboration and license revenue	\$ 11,435	\$ —	\$ —
Grant revenue	500	—	—
Total revenue	11,935	—	—
Operating expenses:			
Research and development	37,945	33,183	33,815
General and administrative	14,710	14,506	13,584
Total operating expenses	52,655	47,689	47,399
Loss from operations	(40,720)	(47,689)	(47,399)
Other income (expense):			
Interest income	757	384	494
Interest expense	—	—	(24)
Total other income, net	757	384	470
Net loss	<u><u>\$(39,963)</u></u>	<u><u>\$(47,305)</u></u>	<u><u>\$(46,929)</u></u>
Basic and diluted net loss per common share	<u><u>\$ (0.86)</u></u>	<u><u>\$ (1.08)</u></u>	<u><u>\$ (1.08)</u></u>
Shares used to compute basic and diluted net loss per common share	<u><u>46,432</u></u>	<u><u>43,890</u></u>	<u><u>43,275</u></u>

See accompanying notes.

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

	Year Ended December 31,		
	2016	2015	2014
Net loss	\$(39,963)	\$(47,305)	\$(46,929)
Unrealized (loss) gain on available-for-sale securities	(10)	30	(110)
Comprehensive loss	\$(39,973)	\$(47,275)	\$(47,039)

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 Income (Loss)	Accumulated Deficit	Total Stockholders' Equity
	Shares	Amount					
	(In thousands, except share and per share data)						
Balance as of December 31, 2013	42,888,168	\$ 43	\$318,103	\$ (16)	\$ 40	\$(172,862)	\$145,308
Net loss	—	—	—	—	—	(46,929)	(46,929)
Unrealized gain / (loss) on investments	—	—	—	—	(110)	—	(110)
Issuance of common stock under equity incentive and employee stock purchase plans	557,928	—	2,123	—	—	—	2,123
Employee stock-based compensation	—	—	7,960	—	—	—	7,960
Compensation expense related to options granted to consultants	—	—	254	—	—	—	254
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	<u>48,057,920</u>	<u>\$ 48</u>	<u>\$356,966</u>	<u>\$ (16)</u>	<u>\$ (50)</u>	<u>\$(307,059)</u>	<u>\$ 49,889</u>

See accompanying notes.

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

	<u>Year Ended December 31,</u>		
	<u>2016</u>	<u>2015</u>	<u>2014</u>
Operating activities			
Net loss	\$ (39,963)	\$(47,305)	\$ (46,929)
Adjustments to reconcile net loss to net cash provided by (used in) operating activities:			
Depreciation of property and equipment	348	477	543
Stock-based compensation	8,535	8,985	8,214
Noncash interest expense, net	179	1,007	2,157
Changes in assets and liabilities:			
Accounts receivable	(30,205)	—	393
Prepays and other current assets	35	236	(376)
Other assets	(119)	—	(21)
Accounts payable	(4)	(73)	(161)
Other liabilities	3,773	(2,654)	1,865
Deferred revenue	96,566	—	—
Net cash provided by (used in) operating activities	<u>39,145</u>	<u>(39,327)</u>	<u>(34,315)</u>
Investing activities			
Purchases of property and equipment, net	(304)	(218)	(352)
Purchases of investments	(136,234)	(24,372)	(100,837)
Sales of investments	—	4,051	—
Maturities of investments	87,774	54,423	139,528
Net cash provided by (used in) investing activities	<u>(48,764)</u>	<u>33,884</u>	<u>38,339</u>
Financing activities			
Proceeds from issuance of common stock	7,000	—	—
Proceeds from exercise of stock options and employee stock purchase plan	1,820	2,191	2,123
Payments on equipment financing obligations	—	—	(330)
Net cash provided by financing activities	<u>8,820</u>	<u>2,191</u>	<u>1,793</u>
Net increase (decrease) in cash and cash equivalents	(799)	(3,252)	5,817
Cash and cash equivalents at beginning of period	<u>12,823</u>	<u>16,075</u>	<u>10,258</u>
Cash and cash equivalents at end of period	<u>\$ 12,024</u>	<u>\$ 12,823</u>	<u>\$ 16,075</u>
Supplemental disclosures of cash flow information			
Cash paid for interest	\$ —	\$ —	\$ 137

See accompanying notes.

CHEMOCENTRYX, INC.
Notes to Consolidated Financial Statements
December 31, 2016

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 year ended December 31, 2016, 96% of the Company's total revenue were derived from the Company's collaboration with Vifor (International) Ltd. (Vifor). The Company did not generate any revenue in 2014 and 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,	
	2016	2015
Vifor (International) Ltd. ⁽¹⁾	\$30,000	\$—
U.S. Food and Drug Administration	205	—
	<u>\$30,205</u>	<u>\$—</u>

- (1) Accounts receivable excludes the additional \$20 million cash commitment due from Vifor in December 2017 in connection with the CCX140 Agreement. See Note 9, "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 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

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

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.

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) relates 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 Condensed 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.

Research and Development Expenses

All research and development expenses are recognized as incurred. Research and development expenses include, but 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, allocated facility costs.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

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 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 as 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 Loss

Comprehensive loss comprises net 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, 2014 and 2016, there were no sales of investments, and therefore there were no reclassifications.

Stock-Based Compensation

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

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

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 Loss Per Share

Basic net loss per common share is computed by dividing net 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 loss per share is computed by dividing net 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 loss per share when their effect is dilutive.

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

	Year Ended December 31,		
	2016	2015	2014
Options to purchase common stock, including purchases from contributions to ESPP	9,358,389	7,861,953	6,849,607
Restricted stock units	440,344	67,481	135,135
Restricted stock awards	31,306	—	—
Warrants to purchase common stock	150,000	150,000	150,000
	<u>9,980,039</u>	<u>8,079,434</u>	<u>7,134,742</u>

Recent Accounting Pronouncements

In August 2014, the Financial Accounting Standards Boards (FASB) issued Accounting Standards Updates (ASU) No. 2014-15 (Subtopic 205-40) - Presentation of Financial Statements—Going Concern: Disclosure of Uncertainties about an Entity's Ability to Continue as a Going Concern (ASU 2014-15) which provides guidance about management's responsibility to evaluate whether or not there is substantial doubt about the Company's

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

2. Summary of Significant Accounting Policies (continued)

ability to continue as a going concern and to provide related footnote disclosure. ASU 2014-15 is effective for the Company the year ending December 31, 2016. Early application is permitted. The adoption of this standard did not have an impact on its financial statements.

In May 2015, the FASB issued a comprehensive new standard on revenue from contracts with customers. The 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.

The Company currently plans to adopt the accounting standard update on January 1, 2018, using the modified retrospective approach. The cumulative effect of adopting the accounting standard update will be recorded to retained earnings on January 1, 2018. The Company is currently at the early stages of analyzing its collaboration agreements to determine the differences in the accounting treatment under ASU 2014-09 compared to the current accounting treatment. During 2016, the Company entered into two license and collaboration agreements. The Company has primarily derived its revenues from license and collaboration agreements. The consideration the Company is eligible to receive under these agreements includes of upfront payments, research and development funding, milestone payments, and royalties. Each license and collaboration agreement is unique and will need to be assessed separately under the five-step process under the new standard. The new revenue recognition standard differs from the current accounting standard in many respects, such as in the accounting for variable considerations and the measurement of progress toward completion of performance obligations. While the Company has not completed an assessment of the impact of adoption, the adoption of ASU 2014-09 may have a material effect on its financial statements.

In February 2016, the FASB issued a new standard that 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 guidance that changes the accounting for certain aspects of share-based payments to employees. The guidance requires the recognition of the income tax effects of awards in the income statement when the awards vest or are settled, thus eliminating additional paid in capital pools. The guidance also allows for the employer to repurchase more of an employee's shares for tax withholding purposes without triggering liability accounting. In addition, the guidance allows for a policy election to account for forfeitures as they occur rather than on an estimated basis. The guidance is effective in 2017 with early adoption permitted. The Company adopted this new standard on January 1, 2017 and does not expect a material impact on its financial statements given the full valuation allowance position on its deferred tax assets.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

3. Cash Equivalents and Investments

The amortized cost and fair value of cash equivalents and investments at December 31, 2016 and 2015 were as follows (in thousands):

	December 31, 2016			
	Amortized Cost	Gross Unrealized		Fair Value
		Gains	Losses	
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

	December 31, 2015			
	Amortized Cost	Gross Unrealized		Fair Value
		Gains	Losses	
Money market fund	\$11,340	\$—	\$—	\$11,340
U.S. treasury securities	14,027	1	(2)	14,026
Government-sponsored agencies	30,959	—	(25)	30,934
Commercial paper	3,992	—	—	3,992
Corporate debt securities	14,528	—	(14)	14,514
Total available-for-sale securities	\$74,846	\$ 1	\$(41)	\$74,806
Classified as:				
Cash equivalents				\$11,340
Short-term investments				58,455
Long-term investments				5,011
Total available-for-sale securities				\$74,806

Cash equivalents in the tables above exclude cash of \$2.3 million and \$1.5 million as of December 31, 2016 and 2015, respectively. All available-for-sale securities held as of December 31, 2016, 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, 2016, have been in a continuous unrealized loss position for more than 12 months. As of December 31, 2016, 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.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

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, 2016 and 2015 (in thousands):

Description	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	\$ 9,746	\$111,737	\$—	\$121,483

Description	December 31, 2015			
	Level 1	Level 2	Level 3	Total
Money market fund	\$11,340	\$ —	\$—	\$ 11,340
U.S. treasury securities	—	14,026	—	14,026
Government-sponsored agencies	—	30,934	—	30,934
Commercial paper	—	3,992	—	3,992
Corporate debt securities	—	14,514	—	14,514
Total assets	\$11,340	\$ 63,466	\$—	\$ 74,806

During the year ended December 31, 2016 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.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

5. Property and Equipment

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

	December 31,	
	2016	2015
Lab equipment	\$ 5,950	\$ 5,740
Computer equipment and software	1,511	1,458
Furniture and fixtures	528	530
Tenant improvements	866	832
	8,855	8,560
Less: accumulated depreciation	(7,950)	(7,611)
	\$ 905	\$ 949

6. Accrued Liabilities

Accrued liabilities consist of the following (in thousands):

	December 31,	
	2016	2015
Research and development related	\$5,482	\$2,223
Compensation related	2,460	1,908
Consulting and professional services	421	454
Other	282	234
	\$8,645	\$4,819

7. 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. The total rent obligation is being expensed ratably over the term of the agreement, as amended. Rental expenses for the years ended December 31, 2016, 2015, and 2014 were \$1.2 million, \$1.1 million and \$1.0 million, respectively.

Future minimum lease payments under all noncancelable operating leases as of December 31, 2016, are as follows (in thousands):

Year ending December 31:	
2017	\$ 915
2018	937
2019	315
Total minimum lease payments	\$2,167

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

8. 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, 2016. 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, which was outstanding as of December 31, 2016 and December 31, 2015.

For the years ended December 31, 2016, 2015, and 2014, the Company paid Bio-Techne \$114,000, \$62,000 and \$93,000, respectively, for research materials. As of December 31, 2016 and 2015, the Company had an accounts payable balance due to Bio-Techne for the purchase of research materials of \$24,885 and zero, respectively.

9. Collaboration and License 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 other countries not licensed to Vifor. 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. As of December 31, 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 approximates 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 to 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.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Collaboration and License Agreements (continued)

As of December 31, 2016, the \$12.5 million potentially creditable towards a CCX140 license agreement expired and was reclassified to the amortizable portion of deferred revenue, which continues to be recognized over the estimated period of performance under the Avacopan Agreement ending in June 2020. For the year ended December 31, 2016, the Company recognized \$11.4 million of collaboration and license revenue under the Avacopan Agreement.

Upon achievement of certain regulatory and commercial milestones with avacopan, the Company will receive additional payments of up to \$510.0 million under the Avacopan Agreement. In addition, the Company will receive royalties, with rates ranging between the teens and mid-twenties, on future potential net sales of avacopan by Vifor in the licensed territories.

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 U.S. and China, in rare renal diseases. CCX140 is an orally-administered inhibitor of the chemokine receptor known as CCR2. ChemoCentryx retains marketing rights for rare renal disease in the U.S. and China, while Vifor has commercialization rights in the rest of the world.

Pursuant to the CCX140 Agreement, ChemoCentryx will be responsible for the clinical development of CCX140 in rare renal diseases, while sharing the cost of such development with Vifor. 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, ChemoCentryx would receive co-promotion rights in CKD in the U.S.

Under the terms of the CCX140 Agreement, the Company received a non-refundable upfront commitment totaling \$50.0 million, \$30.0 million of which was reflected in accounts receivable as of December 31, 2016. The remaining \$20.0 million due on the first anniversary of the CCX140 Agreement was not reflected in accounts receivable as of December 31, 2016.

The upfront commitment of \$50 million will be recognized over the estimated period of performance under the CCX140 Agreement, which approximates 5.0 years, ending in December 2021. 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 to 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 upfront consideration of \$50.0 million into a single unit of accounting.

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 double-digit royalties 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

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

9. Collaboration and License Agreements (continued)

the allocation of arrangement consideration if and when achieved, resulting in an accounting treatment similar to the upfront payment. As of December 31, 2016, the Company has not received any milestone payments under the Avacopan Agreement or the CCX140 Agreement. 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. The avacopan territory expansion signed in February 2017 and the original Avacopan Agreement signed in May 2016 will be accounted for as a combined agreement.

10. Government Grant

In April 2016, the Company was awarded an Orphan Products Development grant by the U.S. Food and Drug Administration in the amount of \$500,000 to support the clinical development of avacopan. The term of the grant expires in May 2017. During the year ended December 31, 2016, the Company recognized \$500,000 of grant revenue. As of December 31, 2016, \$205,000 was recorded as accounts receivable.

11. 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, 2016, a total of 9,600,000 shares of the Company's common stock were reserved for issuance under the 2012 Plan. In addition, the number of shares available for issuance under the 2012 Plan will be annually increased by an amount equal to the lesser of: 2,000,000 shares; 4% of the outstanding shares of the Company's common stock as of the last day of the Company's immediately preceding fiscal year; or an amount determined by the Company's Board of Directors. In November 2016, the Board of Directors approved an increase to the number of shares reserved for issuance under the 2012 Plan by 1,900,000 shares effective January 1, 2017. Collectively, the 1997 Plan, the 2002 Plan and the 2012 Plan are known as the Stock Plans.

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, 2015 and 2014, the weighted average grant date fair value for restricted stock granted was \$7.88 and \$4.81, respectively.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Equity Incentive Plans (continued)

The activity for restricted stock is summarized as follows:

	Shares	Weighted Average Grant-Date Fair Value
Balance at December 31, 2015	67,481	\$7.93
Granted	458,116	4.48
Vested	(53,947)	7.75
Canceled	—	—
Unvested at December 31, 2016	471,650	\$4.60

As of December 31, 2016, there was \$1.4 million of unrecognized compensation expense associated with unvested restricted stock, which is expected to be recognized over a weighted-average period of 1.9 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.

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

	Available for Grant	Shares	Weighted Average Exercise Price	Weighted Average Remaining Contractual Term	Aggregate Intrinsic Value
Balance at December 31, 2015	2,157,641	7,847,449	\$8.52		\$ 8,103,675
Shares authorized	1,750,000				
Granted ⁽¹⁾	(2,522,216)	2,064,100	4.04		
Exercised		(295,935)	4.37		
Forfeited and expired	270,099	(270,099)	6.41		
Outstanding at December 31, 2016	1,655,524	9,345,515	\$7.72	6.58	10,789,001
Vested at December 31, 2016		5,938,347	8.89	5.39	3,608,574
Exercisable at December 31, 2016		5,938,972	8.89	5.39	3,610,136
Vested and expected to vest, net of estimated forfeiture at December 31, 2016		9,182,327	\$7.77	6.53	\$10,372,111

(1) The difference between the number of shares available for grant and shares outstanding represents the RSUs and RSAs granted for the period.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Equity Incentive Plans (continued)

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 \$0.7 million, \$2.1 million and \$1.1 million during 2016, 2015, and 2014, respectively. As of December 31, 2016, there was \$9.6 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 2.37 years.

Early Exercise of Stock Options

Certain equity incentive plans allow for the granting of options that may be exercised before the options have vested. The difference between the number of shares vested and exercisable as of December 31, 2016 in the table above represents such shares that are exercisable before they are vested. Shares issued as a result of early exercise that have not vested are subject to repurchase by the Company upon termination of the purchaser's employment or services, at the price paid by the purchaser.

For the years ended December 31, 2016 and 2015, there were no shares of common stock issued related to the early exercise of stock options subject to repurchase by the Company.

As of December 31, 2016, stock options outstanding were as follows:

Exercise Price Range	Options Outstanding	
	Shares	Weighted-Average Contractual Life
\$2.10 - \$3.29	268,000	9.29
\$3.57	1,323,100	9.18
\$3.72 - \$5.94	373,241	7.04
\$6.00	1,089,837	2.16
\$6.05 - \$6.30	1,280,704	5.61
\$6.60 - \$6.90	226,154	4.40
\$7.10	988,500	7.13
\$7.12 - \$7.85	290,200	9.63
\$8.19	1,391,966	8.10
\$8.22 - \$15.90	2,113,813	5.93
	9,345,515	6.58

Employee Stock Purchase Plan

In February 2012, the stockholders approved the ESPP. As of December 31, 2016, a total of 750,000 shares of the Company's common stock were reserved for issuance under the ESPP. In addition, the number of shares available for issuance under the ESPP may be annually increased on the first day of each fiscal year during the term of the ESPP, beginning with the 2012 fiscal year, by an amount equal to the lesser of: 300,000 shares; 1% of outstanding shares of the Company's common stock; or an amount determined by the Company's Board of Directors. The ESPP provides for an aggregate limit of 3,000,000 shares of common stock that may be issued under the ESPP during the term of the ESPP. In November 2016, the Board of Directors approved an increase to the number of shares reserved for issuance under the ESPP by 200,000 shares effective January 1, 2017.

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Equity Incentive Plans (continued)

The Company issued 157,893 share, 134,579 shares and 149,788 shares under the ESPP in 2016, 2015 and 2014, respectively. As of December 31, 2016, 226,556 shares were available for issuance under the ESPP. As of December 31, 2016, 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>2016</u>	<u>2015</u>	<u>2014</u>
Research and development	\$3,245	\$3,240	\$2,756
General and administrative	4,977	5,620	5,204
Total	<u>\$8,222</u>	<u>\$8,860</u>	<u>\$7,960</u>

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 2016, 2015, and 2014 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>2016</u>	<u>2015</u>	<u>2014</u>	<u>2016</u>	<u>2015</u>	<u>2014</u>
Dividend yield	0%	0%	0%	0%	0%	0%
Volatility	65.6%	67.6%	74.1%	99.7%	60.1%	55.5%
Weighted-average expected life (in years)	6.0	6.0	6.0	0.5	0.5	0.5
Risk-free interest rate	1.58%	1.70%	1.96%	0.47%	0.21%	0.06%
Weighted average grant date fair value	\$2.43	\$5.00	\$4.29	\$2.29	\$2.22	\$1.49

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

11. Equity Incentive Plans (continued)

Stock Options Granted to Nonemployees

During 2016, 2015 and 2014, the Company granted to consultants options to purchase 15,000, 90,300, and 150,000 shares of common stock, respectively. The stock-based compensation expense related to nonemployees will fluctuate as the fair value of the Company's common stock fluctuates. In connection with grants of stock options to nonemployees, the Company recorded stock-based compensation expense as follows (in thousands):

	Year Ended December 31,		
	2016	2015	2014
Research and development	\$313	\$105	\$254
General and administrative	—	20	—
Total	\$313	\$125	\$254

Valuation Assumptions

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

	Year Ended December 31,		
	2016	2015	2014
Dividend yield	0%	0%	0%
Volatility	65-68%	65-66%	68-76%
Weighted-average expected life (in years)	6.1-9.9	5.6-9.9	6.6-9.8
Risk-free interest rate	1.3-2.4%	1.7-2.4%	2.0-2.7%

12. 401(k) Plan

In October 1997, the Company established the ChemoCentryx 401(k) Plan and Trust (the 401(k) Plan). Employees may contribute, up to the percentage limit imposed by the Internal Revenue Code of 1986, as amended, an amount of their salary each calendar year until termination of their employment with the Company. The Company may elect to make matching contributions, as per the Plan; however, no matching contributions were made in the years ended December 31, 2016, 2015, and 2014.

13. Income Taxes

The Company's loss before tax is only attributable to U.S. operations. A reconciliation of the federal statutory income tax rate to the Company's effective income tax rate is as follows:

	Year Ended December 31,		
	2016	2015	2014
Federal statutory income tax rate	(34.0%)	(34.0%)	(34.0%)
State income taxes, net of federal benefit	(5.80)	(5.80)	(5.80)
Permanent items	2.10	1.90	2.60
Research and development credits	(2.80)	(2.10)	(1.80)
Change in valuation allowance	38.1	40.0	39.0
Other	2.40	—	—
(Benefit from) provisions for income taxes	— %	— %	— %

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

13. Income Taxes (continued)

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

	2016	2015
Net operating loss carryforwards	\$ 102,269	\$ 90,813
Research and development credit	8,719	7,583
Amortization of deferred stock compensation - non-qualified	10,567	8,603
Reserves and accruals	1,747	1,125
Depreciation and amortization	445	420
Net deferred tax asset	123,747	108,544
Less: valuation allowance	(123,747)	(108,544)
Net deferred tax assets	\$ —	\$ —

The Company has concluded that its deferred tax assets are not more likely than not to be realized. Accordingly, the total deferred tax assets have been fully offset by a valuation allowance. The Company's valuation allowance increased by approximately \$15.2 million, \$19.3 million, and \$18.3 million during 2016, 2015, and 2014 respectively.

At December 31, 2016, the Company had federal and state net operating loss carryforwards of approximately \$264.5 million and \$247.9 million, respectively. The federal net operating loss carryforwards begin to expire in 2025 and the state net operating loss carryforwards begin to expire in 2019, if not utilized.

The Company has federal and state research and development credit carryforwards of \$10.0 million and \$5.3 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, as amended, and similar state provisions. The annual limitation may result in the expiration of certain net operating loss and credit carryforwards before their utilization.

The deferred tax asset balances as of December 31, 2016 and 2015 did not include excess tax benefits from stock option exercises. The amount excluded at December 31, 2016 and 2015 was \$5.3 million and \$5.5 million respectively. The use of tax benefits associated with the stock option related deductions will be credited directly to stockholders' equity upon ultimate realization.

A reconciliation of the Company's unrecognized tax benefits for the years ended December 31, 2016, 2015, and 2014, is as follows (in thousands):

	Unrecognized Income Tax Benefits
Balance as of December 31, 2014	\$ 5,390
Additions for current tax positions	495
Releases	(1,013)
Balance as of December 31, 2015	\$ 4,872
Additions for current tax positions	558
Balance as of December 31, 2016	\$ 5,430

CHEMOCENTRYX, INC.

Notes to Consolidated Financial Statements (continued)

13. Income Taxes (continued)

As of December 31, 2016 and 2015, the Company had approximately \$5.4 million and \$4.9 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 2013 and 2012, 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, 2016, there has been no interest expense or penalties related to unrecognized tax benefits.

The IRS has completed its audit for the years ended December 31, 2007 and 2008, and there have been no adjustments to the Company's attributes carryforwards; however, the research and development credit may be subject to re-examination.

The Company has completed the examination by the California Franchise Tax Board for the years ended December 31, 2008 and 2009. The examination resulted in an update to the Company's California net operating loss carryforwards and R&D credits. The Company is not currently under examination in any other jurisdictions.

14. Selected Quarterly Financial Data (unaudited)

Selected quarterly results from operations for the years ended December 31, 2016 and 2015 are as follows (in thousands except per share amounts):

	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 and diluted net loss per share	(0.34)	(0.22)	(0.15)	(0.16)
	2015 Quarter Ended			
	March 31	June 30	September 30	December 31
Revenue	\$ —	\$ —	\$ —	\$ —
Net loss	(12,006)	(12,078)	(11,647)	(11,574)
Basic and diluted net loss per share	(0.28)	(0.28)	(0.26)	(0.26)

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

15. Subsequent Event

In 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 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 million upfront cash commitment for the new rights, plus tiered double-digit royalties on potential net sales.

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 14, 2017

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 14, 2017
<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 14, 2017
<u>/s/ Thomas A. Edwards</u> Thomas A. Edwards	Director	March 14, 2017
<u>/s/ Joseph M. Feczko, M.D.</u> Joseph M. Feczko, M.D.	Director	March 14, 2017
<u>/s/ Roger C. Lucas, Ph.D.</u> Roger C. Lucas, Ph.D.	Director	March 14, 2017
<u>/s/ Henry A. McKinnell, Jr., Ph.D.</u> Henry A. McKinnell, Jr., Ph.D.	Director	March 14, 2017
<u>/s/ Geoffrey M. Parker</u> Geoffrey M. Parker	Director	March 14, 2017
<u>/s/ James L. Tyree</u> James L. Tyree	Director	March 14, 2017

CORPORATE INFORMATION

ChemoCentryx, Inc.
850 Maude Avenue
Mountain View, CA 94043
650.210.2900 tel
650.210.2910 fax
<http://www.chemocentryx.com>

CORPORATE COUNSEL

Latham & Watkins
San Diego, CA

TRANSFER AGENT

American Stock Transfer & Trust Company, LLC
6201 15th Avenue
Brooklyn, NY 11219
888-444-0057
customerservice@astinvestor.com
<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
Vice Chairman, Bio-Techne Corporation
Lead Independent Director, ChemoCentryx, Inc.

Thomas A. Edwards
Retired Partner, Latham & Watkins, LLP

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
Former Executive Vice President and Chief Financial Officer,
Anacor Pharmaceuticals, Inc,
Chairman of the Audit Committee, ChemoCentryx, Inc.

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

MANAGEMENT

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

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

Pirow Bekker, MD, PhD
Chief Medical Officer

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

CHEMOCENTRYX, INC.

850 Maude Avenue | Mountain View, CA 94043 | 650.210.2900 tel | 650.210.2910 fax

<http://www.chemocentryx.com>