



Lexicon Pharmaceuticals Reports First Quarter 2019 Financial Results and Provides a Business Update

May 1, 2019

The Woodlands, Texas, May 1, 2019 - Lexicon Pharmaceuticals, Inc. (Nasdaq: LXRX), today reported financial results and provided a business update for the three months ended March 31, 2019.

"XERMELO net sales grew more than 20% in the first quarter of 2019 versus the same period in 2018. We advanced the telotristat ethyl Phase 2 program for biliary tract cancer, with the first patient dosed, and we continued making progress in advancing LX9211, a candidate for neuropathic pain," said Lonnel Coats, Lexicon's president and chief executive officer. "We are pleased with the recent approval of Zynquista in type 1 diabetes in Europe, which represents the second product originating from our own laboratories to be approved in a major region in a span of only 26 months. In the U.S., we and our collaborator, Sanofi, will be working with the FDA to better understand sotagliflozin's potential pathway to approval in type 1 diabetes. Lastly, we look forward to topline results from the first of several Phase 3 studies of sotagliflozin in type 2 diabetes. We will provide updates on our progress throughout the year."

First Quarter Product and Pipeline Highlights

XERMELO® (telotristat ethyl)

- XERMELO U.S. net sales were \$6.7 million in the first quarter of 2019.
- The first patient was dosed in the **Telotristat Ethyl for Advanced Biliary Tract Cancer**, or TELE-ABC, study, a Phase 2a clinical study of telotristat ethyl in patients with biliary tract cancer.
- Data from a patient-reported survey demonstrating improvement in carcinoid syndrome symptoms after initiation of XERMELO therapy in the real world were presented at the 16th Annual European Neuroendocrine Tumor Society Conference (ENETS).

Zynquista™ (sotagliflozin)

- The U.S. Food and Drug Administration (FDA) issued a complete response letter (CRL) for sotagliflozin for the treatment of adults with type 1 diabetes in combination with insulin in March. The CRL followed the Endocrinologic and Metabolic Drugs Advisory Committee's eight to eight vote in January on the question of whether the overall benefits of sotagliflozin outweighed the risks to support approval.
- In March, the European Medicines Agency's Committee for Medicinal Products for Human Use (CHMP) adopted a positive opinion on the Marketing Authorization of Zynquista in both a 200-mg and 400-mg dose for use as an adjunct to insulin therapy to improve blood sugar (glycemic) control in adults with type 1 diabetes (T1D) mellitus with a body mass index (BMI) ≥ 27 kg/m², who have failed to achieve adequate glycemic control despite optimal insulin therapy. Subsequent to quarter-end, Zynquista received formal approval in the European Union in accordance with the CHMP opinion.

First Quarter 2019 Financial Highlights

Revenues: Revenues for the three months ended March 31, 2019 decreased to \$9.2 million from \$25.4 million for the corresponding period in 2018, primarily due to lower revenues recognized from our collaboration and license agreement with Sanofi, partially offset by a milestone payment from Ipsen and an increase in net product revenues. Net product revenues for the three months ended March 31, 2019 included \$6.7 million from net sales of XERMELO in the U.S., up 24% from the corresponding period in 2018.

Cost of Sales: Cost of sales related to sales of XERMELO for the three months ended March 31, 2019 and 2018 was \$0.6 million and \$0.5 million, respectively.

Research and Development (R&D) Expenses: Research and development expenses for the three months ended March 31, 2019 decreased to \$12.0 million from \$47.7 million for the corresponding period in 2018, primarily due to lower external clinical development costs relating to sotagliflozin.

Selling, General and Administrative (SG&A) Expenses: Selling, general and administrative expenses for the three months ended March 31, 2019 decreased to \$14.1 million from \$14.9 million for the corresponding period in 2018, primarily due to decreased marketing costs, partially offset by higher salaries and benefits related to increased headcount.

Net Loss: Net loss for the three months ended March 31, 2019 was \$21.8 million, or \$0.21 per share, compared to a net loss of

\$41.8 million, or \$0.40 per share, in the corresponding period in 2018. For the three months ended March 31, 2019 and 2018, net loss included non-cash, stock-based compensation expense of \$3.4 million and \$3.1 million, respectively.

Cash and Investments: As of March 31, 2019, Lexicon had \$133.1 million in cash and investments, as compared to \$160.1 million as of December 31, 2018.

Anticipated Near-Term Milestones

- June, September 2019 – Presentation of new analyses from pivotal studies of sotagliflozin in type 1 diabetes at the annual ADA and EASD meetings
- 2H 2019 – Topline Phase 1b data for LX9211
- 2019 – Topline results from core Phase 3 studies for sotagliflozin in type 2 diabetes
- 2019 – Completion of patient enrollment of the initial safety cohort in the Phase 2 study for telotristat ethyl in biliary tract cancer

Conference Call and Webcast Information

Lexicon management will hold a live conference call and webcast today at 8:00 am EDT / 7:00 am CDT to review its financial and operating results and to provide a general business update. The dial-in number for the conference call is 888-645-5785 (U.S./Canada) or 970-300-1531 (international). The conference ID for all callers is 8663398. The live webcast and replay may be accessed by visiting Lexicon's website at www.lexipharma.com/investors. An archived version of the webcast will be available on the website for 14 days.

About XERMELO (telotristat ethyl)

Discovered using Lexicon's unique approach to gene science, XERMELO (telotristat ethyl) is the first and only approved oral therapy for carcinoid syndrome diarrhea in combination with somatostatin analog (SSA) therapy in adults inadequately controlled by SSAs. XERMELO targets tryptophan hydroxylase, an enzyme that mediates the excess serotonin production within metastatic neuroendocrine tumor (mNET) cells. Lexicon has built the in-house capability and infrastructure to launch and market XERMELO in the U.S., where it retains all commercialization rights. Lexicon also retains rights to market XERMELO in Japan. Lexicon has established a license and collaboration agreement with Ipsen to commercialize XERMELO in Europe and other countries outside of U.S. and Japan.

XERMELO was approved by the U.S. Food and Drug Administration on February 28, 2017 and by the European Commission on September 19, 2017 for the treatment of carcinoid syndrome diarrhea in combination with SSA therapy in adults inadequately controlled by SSA therapy. Carcinoid syndrome is a rare condition that occurs in patients living with metastatic NETs (mNETs) and is characterized by frequent and debilitating diarrhea. XERMELO targets the overproduction of serotonin inside mNET cells, providing an additional treatment option for patients suffering from carcinoid syndrome diarrhea.

XERMELO (telotristat ethyl) Important Safety Information

- **Warnings and Precautions:** XERMELO may cause constipation, which can be serious. Monitor for signs and symptoms of constipation and/or severe, persistent, or worsening abdominal pain in patients taking XERMELO. Discontinue XERMELO if severe constipation or severe, persistent, or worsening abdominal pain develops.
- **Adverse Reactions:** The most common adverse reactions ($\geq 5\%$) include nausea, headache, increased gamma-glutamyl-transferase, depression, flatulence, decreased appetite, peripheral edema, and pyrexia.
- **Drug Interactions:** If necessary, consider increasing the dose of concomitant CYP3A4 substrates, as XERMELO may decrease their systemic exposure. If combination treatment with XERMELO and short-acting octreotide is needed, administer short-acting octreotide at least 30 minutes after administering XERMELO.

For more information about XERMELO, see Full Prescribing Information at www.xermelo.com.

About Zynquista (sotagliflozin)

Discovered using Lexicon's unique approach to gene science, sotagliflozin is an investigational oral dual inhibitor of two proteins responsible for glucose regulation known as sodium-glucose co-transporter types 1 and 2 (SGLT1 and SGLT2). SGLT1 is responsible for glucose absorption in the gastrointestinal tract, and SGLT2 is responsible for glucose reabsorption by the kidney.

Lexicon entered into a collaboration and license agreement with Sanofi in November 2015 under which Lexicon granted Sanofi an exclusive, worldwide (excluding Japan), royalty-bearing right and license to develop, manufacture and commercialize sotagliflozin. Lexicon is responsible for all clinical development activities relating to type 1 diabetes and has exercised an exclusive option to co-promote and have a significant role, in collaboration with Sanofi, in the commercialization of sotagliflozin for the treatment of type 1 diabetes in the U.S. Sanofi is responsible for all clinical development and commercialization of sotagliflozin for the treatment of type 2 diabetes worldwide (excluding Japan) and is solely responsible for the commercialization of sotagliflozin for the treatment of type 1 diabetes outside the U.S. (excluding Japan). Zynquista has been approved in the European Union for use as an adjunct to insulin therapy to improve glycemic control in adults with type 1 diabetes and a body mass index ≥ 27 kg/m², who could not achieve adequate glycemic control despite optimal insulin therapy. Sotagliflozin has not yet been approved for use in any other jurisdiction.

About Lexicon Pharmaceuticals

Lexicon is a fully integrated biopharmaceutical company that is applying a unique approach to gene science based on Nobel Prize-winning technology to discover and develop precise medicines for patients with serious, chronic conditions. Through its Genome5000™ program, Lexicon scientists have studied the role and function of nearly 5,000 genes over the last 20 years and have identified more than 100 protein targets with significant therapeutic potential in a range of diseases. Through the precise targeting of these proteins, Lexicon is pioneering the discovery and development of innovative medicines to safely and effectively treat disease. In addition to its first commercial product, XERMELO for carcinoid syndrome diarrhea, Lexicon has a pipeline of promising drug candidates in clinical and pre-clinical development in diabetes and metabolism and neuropathic pain. For additional information please visit www.lexpharma.com.

Safe Harbor Statement

This press release contains “forward-looking statements,” including statements relating to Lexicon’s long-term outlook on its business, including the commercialization of XERMELO (telotristat ethyl), the clinical development of, the regulatory filings for, and the potential therapeutic and commercial potential of telotristat ethyl, sotagliflozin, LX2761 and LX9211. In addition, this press release also contains forward looking statements relating to Lexicon’s growth and future operating results, discovery, development and commercialization of products, strategic alliances and intellectual property, as well as other matters that are not historical facts or information. All forward-looking statements are based on management’s current assumptions and expectations and involve risks, uncertainties and other important factors, specifically including Lexicon’s ability to meet its capital requirements, successfully commercialize XERMELO, successfully conduct preclinical and clinical development and obtain necessary regulatory approvals of telotristat ethyl, sotagliflozin, LX2761, LX9211 and its other potential drug candidates on its anticipated timelines, achieve its operational objectives, obtain patent protection for its discoveries and establish strategic alliances, as well as additional factors relating to manufacturing, intellectual property rights, and the therapeutic or commercial value of its drug candidates. Any of these risks, uncertainties and other factors may cause Lexicon’s actual results to be materially different from any future results expressed or implied by such forward-looking statements. Information identifying such important factors is contained under “Risk Factors” in Lexicon’s annual report on Form 10-K for the year ended December 31, 2018, as filed with the Securities and Exchange Commission. Lexicon undertakes no obligation to update or revise any such forward-looking statements, whether as a result of new information, future events or otherwise.

For Investor Inquiries:

Kimberly Lee, D.O.
Head of Investor Relations and Corporate Strategy
Lexicon Pharmaceuticals
(281) 863-3383
klee@lexpharma.com

For Media Inquiries:

Chas Schultz
Executive Director, Corporate Communications and Patient Advocacy
Lexicon Pharmaceuticals
(281) 863-3421
cschultz@lexpharma.com