Xenon Pharmaceuticals Reports Third Quarter 2019 Financial Results and Provides Corporate Update

November 5, 2019

In Addition to Ongoing XEN1101 Phase 2b Clinical Trial in Adult Focal Epilepsy, Company Advances Epilepsy Programs and Anticipates Initiating Pediatric Clinical Trials in KCNQ2 (XEN496) and SCN8A (XEN901) Developmental and Epileptic Encephalopathies in 2020

XEN007 Phase 2 Proof-Of-Concept, Physician-Led Study Initiated in Pediatric Patients with Treatment-Resistant Childhood Absence Epilepsy

Conference Call at 4:30 pm ET Today

BURNABY, British Columbia, Nov. 05, 2019 (GLOBE NEWSWIRE) -- Xenon Pharmaceuticals Inc. (Nasdaq:XENE), a clinical stage biopharmaceutical company, today reported its financial results for the third quarter ended September 30, 2019 and provided a corporate update.

Dr. Simon Pimstone, Xenon's Chief Executive Officer, said, "We continue to make strong progress in advancing our portfolio of neurology-focused candidates. Patient enrollment is ongoing across sites in the U.S., Canada, and Europe for our XEN1101 Phase 2b clinical trial in adult focal epilepsy. We have completed development of pediatric-specific formulations for both our XEN496 and XEN901 programs, and we are currently conducting pharmacokinetic studies in healthy adult volunteers to support the anticipated pediatric clinical trials in both KCNQ2- and SCN8A-related developmental and epileptic encephalopathies."

Dr. Pimstone continued, "Looking ahead, we anticipate a number of important milestone opportunities across our epilepsy pipeline, including XEN901, XEN496, and XEN1101. We have numerous activities and scientific posters planned at the upcoming American Epilepsy Society (AES) Annual Meeting in Baltimore, and look forward to connecting with our network of leading physicians, clinicians, and scientific advisory board members, as well as patients and advocacy groups. We anticipate having multiple mid to late stage epilepsy clinical trials underway in 2020 putting us in a position to generate important clinical data across our portfolio."

Dr. Pimstone added, "We are delighted to announce the initiation of a physician-led Phase 2 study examining the efficacy of XEN007 in pediatric patients diagnosed with treatment-resistant childhood absence epilepsy."

Dr. Pimstone continued, "This past quarter we also successfully completed a transaction when Flexion Therapeutics acquired XEN402 for development as FX301 for pain. We believe FX301's extended release formulation from a thermosensitive hydrogel could be well suited for control of post-operative pain based on the in vivo data generated with XEN402, and we look forward to its progression through clinical development."

Highlights and Anticipated Milestones

- XEN496 (active ingredient ezogabine) is a Kv7 potassium channel modulator being developed by Xenon. The FDA has granted orphan drug designation (ODD) for XEN496 as a treatment of KCNQ2 developmental and epileptic encephalopathy (KCNQ2-DEE). Xenon has developed XEN496 as a pediatric-specific, granule formulation to be packaged as single-dose sprinkle capsules. A planned pharmacokinetic (PK) study will test XEN496 in healthy adult volunteers, with all subjects expected to be dosed by year-end. Based on the anticipated timing of data from stability studies ongoing for XEN496 and based on the anticipated timing of the PK study data in adult volunteers, Xenon expects to file an Investigational New Drug (IND) application in the first quarter of 2020 in order to initiate a Phase 3 clinical trial in KCNQ2-DEE. The FDA has indicated that it is acceptable to study XEN496 in infants and children up to four years old, and that a single, small pivotal trial may be considered adequate in order to demonstrate XEN496's efficacy in KCNQ2-DEE, provided the study shows evidence of a clinically meaningful benefit in patients with the intended indication.

- XEN1101 is a differentiated Kv7 potassium channel modulator being developed for the treatment of epilepsy and potentially other neurological disorders. A Phase 2b double-blind, placebo-controlled, multicenter clinical trial is underway to evaluate the clinical efficacy, safety and tolerability of XEN1101 administered as adjunctive treatment in approximately 300 adult patients with focal epilepsy. The primary endpoint is the median percent change in monthly focal seizure frequency from baseline compared to treatment period of active versus placebo. Patient enrollment for this XEN1101 Phase 2b clinical trial is ongoing in the United States, Canada and Europe. Long term six-and nine-month toxicology studies have now been completed, providing support to the planned 12-month open label extension for patients enrolled in the Phase 2b clinical trial. Depending upon the rate of enrollment, top-line results are anticipated in the second half of 2020.

- XEN901 is a potent, highly selective Nav1.6 sodium channel inhibitor being developed for the treatment of epilepsy. A Phase 1 clinical trial was completed using a powder-in-capsule formulation of XEN901 in healthy adult subjects. Xenon

received feedback from the FDA regarding the requirements for advancing XEN901 directly into a pediatric clinical trial to study its efficacy in SCN8A developmental and epileptic encephalopathy (SCN8A-DEE). Xenon has developed a pediatric-specific, granule formulation of XEN901, and juvenile toxicology studies to support pediatric development activities have recently been completed. All healthy adult volunteers have now been dosed in a PK study testing the novel pediatric formulation of XEN901. Based on the anticipated timing of data from stability studies ongoing for XEN901 and based on the anticipated timing of the PK study data in adult volunteers, Xenon anticipates filing an IND, or IND-equivalent, submission in the first quarter of 2020 in order to start a proposed Phase 2 or 3 clinical trial in SCN8A-DEE patients, and continues to evaluate other potential indications for XEN901, including adult focal epilepsy. Xenon continues to evaluate selective Nav1.6 compounds and Nav1.2/1.6 dual acting compounds for development behind XEN901, and expects to highlight some of this pre-clinical work at the upcoming AES meeting in December.

- XEN007 (active ingredient flunarizine) is a CNS-acting calcium channel modulator that modulates Cav2.1 and T-type calcium channels. Other reported mechanisms include dopamine, histamine and serotonin inhibition. A physician-led, Phase 2 proof-of-concept study has recently been initiated to examine the potential clinical efficacy, safety, and tolerability of XEN007 as an adjunctive treatment in pediatric patients diagnosed with treatment-resistant childhood absence epilepsy. Absence seizures are characterized by an abrupt impairment of awareness with arrest in behaviour, staring, eye lid fluttering, and automatisms associated with generalized 3 Hz spike wave discharges (SWD) on electroencephalogram (EEG). A child may have one or many (up to 100) absence seizures a day and have problems with attention and learning. XEN007 has demonstrated efficacy in pre-clinical models of absence seizures, and flunarizine has been shown to be well tolerated clinically. XEN007 significantly reduced the number and duration of SWDs on EEG in these models as mono-therapy and, when combined with valproic acid or ethosuximide, significantly reduced the SWD EEGs more than any drug alone. Results from this Phase 2 investigator-led proof-of-concept study are expected in 2020. Depending on the results from the study, CAE may represent a potential orphan indication for future development of XEN007. Available in certain countries outside of the United States, flunarizine has been reported to have clinical benefit in treating migraine and other neurological disorders. The FDA granted a rare pediatric disease designation for XEN007 as a treatment of alternating hemiplegia of childhood (AHC), and previously granted ODD for XEN007 as a treatment of both AHC and hemiplegic migraine. Xenon has entered into key licensing and manufacturing agreements to support the advanced clinical development of XEN007, with a number of orphan pediatric neurological conditions and development pathways under consideration;

- In September 2019, Xenon entered into a definitive agreement with Flexion Therapeutics, Inc. that provides Flexion with the global rights to develop and commercialize XEN402, a Nav1.7 inhibitor. Flexion's new pre-clinical program, known as FX301, will consist of XEN402 formulated for extended release from a thermosensitive hydrogel. The initial development of FX301 is intended to support administration as a peripheral nerve block for control of post-operative pain. In addition to an upfront payment of $3 million, Xenon is eligible for up to $9 million in milestone payments through initiation of a Phase 2 proof-of-concept clinical trial. Xenon may be entitled to future clinical development and global regulatory approval milestone payments of up to $40.75 million, commercialization milestone payments of up to $75 million, as well as future sales royalties ranging from mid-single to low-double digit percentages. Flexion has indicated that it anticipates initiating FX301 clinical trials in 2021.

Third Quarter 2019 Financial Results

Cash and cash equivalents and marketable securities as of September 30, 2019 were $94.6 million, compared to $119.3 million as of December 31, 2018. There were 25,868,594 common shares and 1,016,000 Series 1 Preferred Shares, which are convertible into common shares on a one-for-one basis at the option of the holder, subject to certain limitations, outstanding as of September 30, 2019.

Based on current assumptions, which include fully supporting the planned clinical development of XEN496, XEN1101, XEN901 and XEN007, Xenon anticipates having sufficient cash to fund operations into 2021, excluding any revenue generated from existing partnerships or potential new partnering arrangements.

For the quarter ended September 30, 2019, Xenon reported total revenue of $3.5 million in connection with the agreement entered into in September 2019 with Flexion Therapeutics, Inc. for the global rights to develop and commercialize XEN402. No revenues were recognized in the comparative quarter.

Research and development expenses for the quarter ended September 30, 2019 were $9.8 million, compared to $6.2 million for the same period in 2018. The increase of $3.5 million was primarily attributable to increased spending on our clinical development product candidates XEN496, XEN901 and XEN1101.

General and administrative expenses for the quarter ended September 30, 2019 were $2.7 million, compared $1.9 million for the same period in 2018. The increase of $0.8 million was primarily attributable to increased legal expenses for intellectual property protection and increased costs for recruitment activities.

Other income for the quarter ended September 30, 2019 was $0.1 million, compared to other expenses of $0.2 million for the same period in 2018. The change of $0.3 million was primarily driven by a decrease in interest expense incurred on a term loan.

Net loss for the quarter ended September 30, 2019 was $8.9 million, compared to $14.4 million for the same period in 2018. The change was primarily attributable to a one-time payment made in September 2018 for the buy-out of all future milestone payments

and royalties owed with respect to the XEN1101 program and revenue recognized in the current quarter pursuant to the agreement with Flexion, partially offset by higher research and development and general and administrative expenses as compared to the same period in 2018.

At-the-Market Equity Offering

Xenon also announced today that it has entered into an at-the-market equity offering sales agreement with Jefferies LLC and Stifel, Nicolaus & Company, Incorporated, under which Xenon may sell its common shares, from time-to-time, for up to $50.0 million in aggregate sales proceeds in "at the market" transactions. Sales of the common shares, if any, will only be conducted in the United States through the Nasdaq or another exchange at market prices. No sales of common shares will be made in Canada.

Conference Call Information

Xenon will host a conference call and live audio webcast today at 4:30 p.m. Eastern Time (1:30 p.m. Pacific Time) to discuss its third quarter 2019 financial results and to provide a business update. To participate in the call, please dial (855) 779-9075, or (631) 485-4866 for international callers, and provide conference ID number 5087474. The webcast will be broadcast live on the "Investors" section of Xenon's website at www.xenon-pharma.com and will be available for replay following the call for 30 days.

About Xenon Pharmaceuticals Inc.

We are a clinical stage biopharmaceutical company committed to developing innovative therapeutics to improve the lives of patients with neurological disorders, including rare central nervous system (CNS) conditions. We are advancing a novel product pipeline of neurology therapies to address areas of high unmet medical need, with a focus on epilepsy. For more information, please visit www.xenon-pharma.com.

Safe Harbor Statement

This press release contains forward-looking statements within the meaning of Section 27A of the Securities Act of 1933 and Section 21E of the Securities Exchange Act of 1934 and the Private Securities Litigation Reform Act of 1995 and Canadian securities laws. These forward-looking statements and supporting assumptions are not based on historical fact, and include statements regarding the timing of and results from clinical trials and pre-clinical development activities, including those related to XEN496, XEN901, XEN1101, XEN007 and our other product candidates; the potential efficacy, safety profile, future development plans, addressable market, regulatory success and commercial potential of XEN496, XEN901, XEN1101, XEN007 and our other product candidates; the anticipated timing of IND, or IND equivalent, submissions and the initiation of future clinical trials for XEN496, XEN901, XEN1101, XEN007 and our other product candidates; the efficacy of our clinical trial designs; our ability to successfully develop and achieve milestones in the XEN496, XEN901, XEN1101, XEN007 and other development programs; the timing and results of our interactions with regulators; the potential to advance certain of our product candidates directly into Phase 2 or later stage clinical trials; anticipated enrollment in our clinical trials; the progress and potential of our other ongoing development programs; the sufficiency of our cash to fund operations into 2021; the potential receipt of milestone payments and royalties from our collaborators and partners; the timing of potential publication or presentation of future clinical data and the sale of any common shares pursuant to the at-the-market equity offering, including the price, volume and timing of any distributions. These forward-looking statements are based on current assumptions that involve risks, uncertainties and other factors that may cause the actual results, events or developments to be materially different from those expressed or implied by such forward-looking statements. These risks and uncertainties, many of which are beyond our control, include, but are not limited to: clinical trials may not demonstrate safety and efficacy of any of our or our collaborators' product candidates; our assumptions regarding our planned expenditures and sufficiency of our cash to fund operations may be incorrect; our ongoing discovery and pre-clinical efforts may not yield additional product candidates; any of our or our collaborators' product candidates may fail in development, may not receive required regulatory approvals, or may be delayed to a point where they are not commercially viable; we may not achieve additional milestones in our proprietary or partnered programs; regulatory agencies may not permit certain of our product candidates to advance directly into a Phase 2 or later clinical trials, may impose additional requirements or delay the initiation of clinical trials; the impact of competition; the impact of expanded product development and clinical activities on operating expenses; adverse conditions in the general domestic and global economic markets; as well as the other risks identified in our filings with the Securities and Exchange Commission and the securities commissions in British Columbia, Alberta and Ontario. These forward-looking statements speak only as of the date hereof and we assume no obligation to update these forward-looking statements, and readers are cautioned not to place undue reliance on such forward-looking statements.

XENON PHARMACEUTICALS INC.
Condensed Consolidated Balance Sheets
(Expressed in thousands of U.S. dollars)

	September 30, 2019	December 31, 2018
Assets		
Current assets:		

Cash and cash equivalents and marketable securities	$ 94,603	$ 119,306
Other current assets	1,875	2,026
Other assets	2,426	1,096
Total assets	$ 98,904	$ 122,428
Liabilities		
Current liabilities:		
Accounts payable and accrued expenses	7,923	4,119
Other current liabilities	3,774	—
Other liabilities	12,643	15,014
Total liabilities	$ 24,340	$ 19,133
Shareholders' equity	$ 74,564	$ 103,295
Total liabilities and shareholders' equity	$ 98,904	$ 122,428

XENON PHARMACEUTICALS INC.
Condensed Consolidated Statements of Operations
(Expressed in thousands of U.S. dollars except share and per share amounts)

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2019	**2018**	**2019**	**2018**
Revenue:	$ 3,500	$ —	$ 3,500	$ —
Operating expenses:				
Research and development	9,751	6,248	27,093	17,232
General and administrative	2,700	1,938	7,628	6,354
Buy-out of future milestones and royalties	—	6,000	—	6,000
Total operating expenses	12,451	14,186	34,721	29,586
Loss from operations	(8,951)	(14,186)	(31,221)	(29,586)
Other income (loss)	85	(205)	1,015	3,643
Loss before income taxes	(8,866)	(14,391)	(30,206)	(25,943)
Income tax (expense) recovery	(5)	8	(13)	4
Net loss and comprehensive loss	(8,871)	(14,383)	(30,219)	(25,939)
Net loss attributable to preferred shareholders	(336)	(1,621)	(1,146)	(2,506)
Net loss attributable to common shareholders	$ (8,535)	$ (12,762)	$ (29,073)	$ (23,433)
Net loss per common share:				
Basic and diluted	$ (0.33)	$ (0.63)	$ (1.13)	$ (1.34)
Weighted-average common shares outstanding:				
Basic and diluted	25,793,482	20,306,298	25,773,732	17,472,403

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