XENON

XENON

CORPORATE OVERVIEW

May 2020

NASDAQ: XENE | www.xenon-pharma.com

Forward Looking Statement/Safe Harbor

This presentation and the accompanying oral commentary contain forward-looking statements that involve risks, uncertainties and assumptions. If the risks or uncertainties ever materialize or the assumptions prove incorrect, our results may differ materially from those expressed or implied by such forward-looking statements. All statements other than statements of historical fact could be deemed forward-looking, including, but not limited to, statements regarding the anticipated impact and timing of the COVID-19 pandemic on our business, research and clinical development plans and timelines and results of operations; the timing of and results from clinical trials and pre-clinical development activities, including those related to XEN496, XEN1101, XEN007, and other proprietary products, and those related to NBI-921352, FX-301, and other partnered product candidates; the potential efficacy, safety profile, future development plans, addressable market, regulatory success and commercial potential of XEN496, XEN1101, XEN007 and other proprietary and partnered product candidates; the anticipated timing of IND, or IND equivalent, submissions and the initiation of future clinical trials for XEN496, XEN1101, XEN007, and other proprietary products, and those related to NBI-921352, FX-301, and other partnered candidates; the efficacy of our clinical trial designs; our ability to successfully develop and achieve milestones in the XEN496, XEN1101, XEN007 and other proprietary 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 and the timing thereof; the progress and potential of our other ongoing development programs; the potential receipt of milestone payments and royalties from our collaborators; our expectation of having sufficient cash to fund operations into 2022; and the timing of potential publication or presentation of future clinical data.

These statements are based on estimates and information available to us at the time of this presentation and are not guarantees of future performance. Actual results could differ materially from our current expectations as a result of many factors, including but not limited to: the impact of the COVID-19 pandemic on our business, research and clinical development plans and timelines and results of operations may be more severe and more prolonged than currently anticipated; 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; regulatory agencies may be delayed in reviewing, commenting on or approving any of our or our collaborators' clinical development plans as a result of the COVID-19 pandemic, which could further delay development timelines; 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. Except as required by law, we assume no obligation and do not intend to update these forward-looking statements or to conform these statements to actual results or to changes in our expectations.

NOTE: Comparisons of XEN1101 and ezogabine are based on results in published literature, not based on data resulting from head-to-head trials, and are not direct comparisons of safety or efficacy. Different protocol designs, trial designs, patient selection and populations, number of patients, trial endpoints, trial objectives and other parameters that are not the same between the relevant trials may cause any comparisons of results from different trials to be unreliable.

Xenon Overview

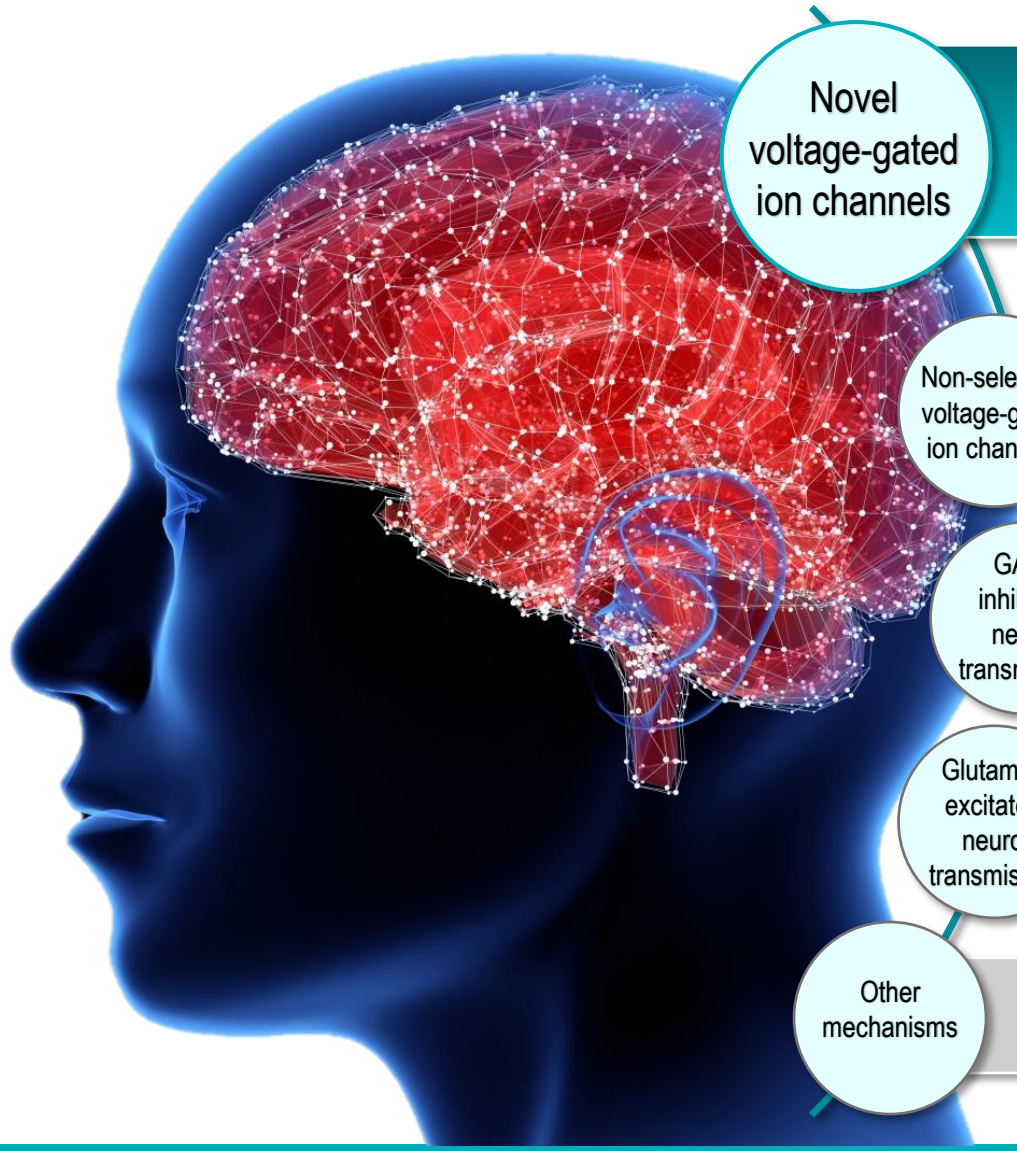
- Small molecule, ion channel neurology-focused biopharma company (NASDAQ: XENE)
- Mid-to-late stage clinical trials and important clinical data anticipated over next 12-18 months
- Strong partnerships with collaborators
- Solid financial position
  - $229.7M in cash, cash equivalents and marketable securities as of March 31, 2020
  - Up to $1.7B in potential milestone payments related to Neurocrine collaboration, including $25M milestone expected in 2020

Ion Channel, Neurology-Focused Pipeline

Therapeutic Program *Indication*	Pre-clinical	Phase 1	Phase 2	Phase 3
XEN496 (Potassium Channel Modulator) *Orphan Pediatric Epilepsy*				
XEN1101 (Potassium Channel Modulator) *Adult Focal Epilepsy*				
XEN007* (Calcium Channel Inhibitor) *Childhood Absence Epilepsy*				
Ion Channel Modulators *Orphan Channelopathies*				
NBI-921352 (XEN901) and $Na_V1.6/1.2$ Sodium Channel Inhibitors *Epilepsy (Orphan Pediatric and Adult Focal)* — Neurocrine Biosciences				
FX301 *Post-operative Pain* — flexion				
$Na_V1.7$ Inhibitors *Pain* — Genentech, A Member of the Roche Group				

*A physician-led, Phase 2 proof-of-concept study is ongoing to examine XEN007 as an adjunctive treatment in pediatric patients diagnosed with treatment-resistant childhood absence epilepsy (CAE).

Novel, Proprietary K_V7 Channel Modulators



XEN1101 (K_V7) | XEN496 (K_V7)

Major Classes Of Approved Anti-Seizure Medications (ASMs)

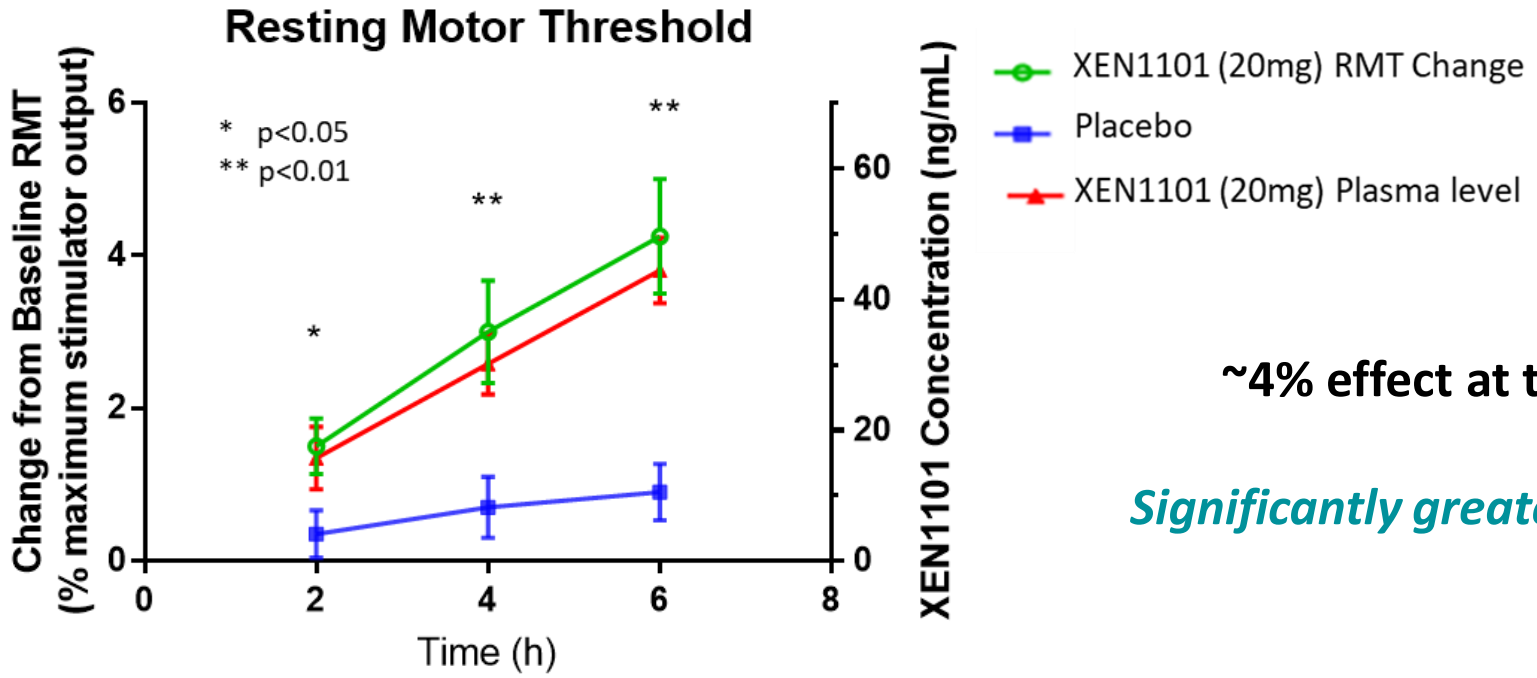
Non-selective voltage-gated ion channels	Phenytoin (Na+); Carbamazepine (Na+); Pregabalin (Ca^{2}+); Lacosamide (Na+)
GABA inhibitory neuro-transmission	Clobazam; Diazepam; Vigabatrin; Tiagabine
Glutamate excitatory neuro-transmission	Topiramate (KA); Perampanel (AMPA)
Other mechanisms	Levetiracetam; Brivaracetam; Cannabidiol (CBD), Cenobamate

XEN1101: Potential "Next-Gen" K_V7 Potassium Channel Modulator

- Same proven MOA in adult focal seizures as ezogabine, but with improvements
  - More potent *in vitro* and *in vivo*
  - 3- to 4-fold selective for KCNQ2/3 over other KCNQ channels
  - Once daily dosing with evening administration
  - No pigmented dimers and no predicted discoloration liability
- Phase 1 studies completed
  - PK supporting once-daily dosing
  - Mild, transient AE profile consistent with MOA (e.g. dizziness, sedation, blurred vision)
  - No safety signals in ECG or Safety Labs; no SAEs
  - Robust TMS signal in Phase 1b study
- 300-patient Phase 2b clinical trial underway in Adult Focal Epilepsy
- Xenon is exploring the development of XEN1101 in other promising neurological indications

Summary of XEN1101 Phase 1b TMS Results

- TMS was used to evaluate the corticospinal and cortical activity profile of XEN1101 compared to placebo in healthy male volunteers
  - Significant plasma concentration dependent reduction of corticospinal (RMT) and cortical (TEP) excitability



~4% effect at t=6 hours vs placebo

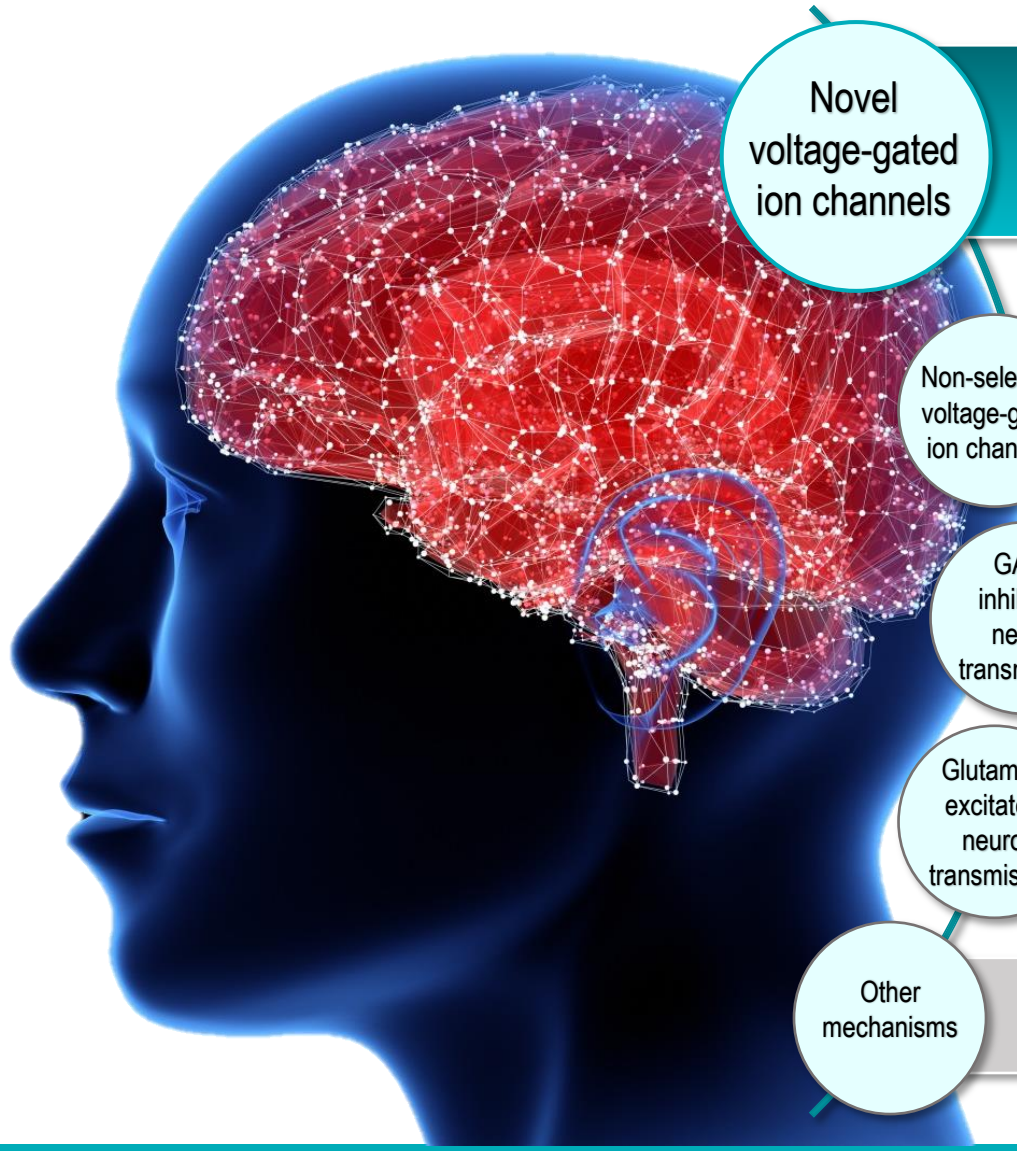
Significantly greater effect than ezogabine

XEN1101 Phase 2b Clinical Trial Underway

- Phase 2b clinical trial (called the X-TOLE study) underway in adult patients with focal epilepsy
  - ~300 patients randomized (blinded) to 1 of 3 active treatment groups or placebo in a 2:1:1:2 fashion (XEN1101 25 mg : 20 mg : 10 mg : Placebo)
  - Primary endpoint is the median percent change in monthly focal seizure frequency from baseline compared to treatment period of active versus placebo
  - Patient enrollment underway in U.S., Canada and Europe, with plans to expand with additional sites in both existing and new jurisdictions
- Blinded safety data shows XEN1101 is well-tolerated
  - The rate of discontinuations in study are lower than modeled
  - To date, ~90% of subjects from double-blind portion have rolled into open-label extension
  - Based on blinded safety data to date, interim analysis is not needed (as study remains consistent with original models)
- Topline data expected in 1H:2021*

*Guidance is dependent upon feedback from the clinical sites and patient enrollment rates given the ongoing COVID-19 pandemic.

Novel, Proprietary K_V7 Channel Modulators



XEN1101 (K_V7) | XEN496 (K_V7)

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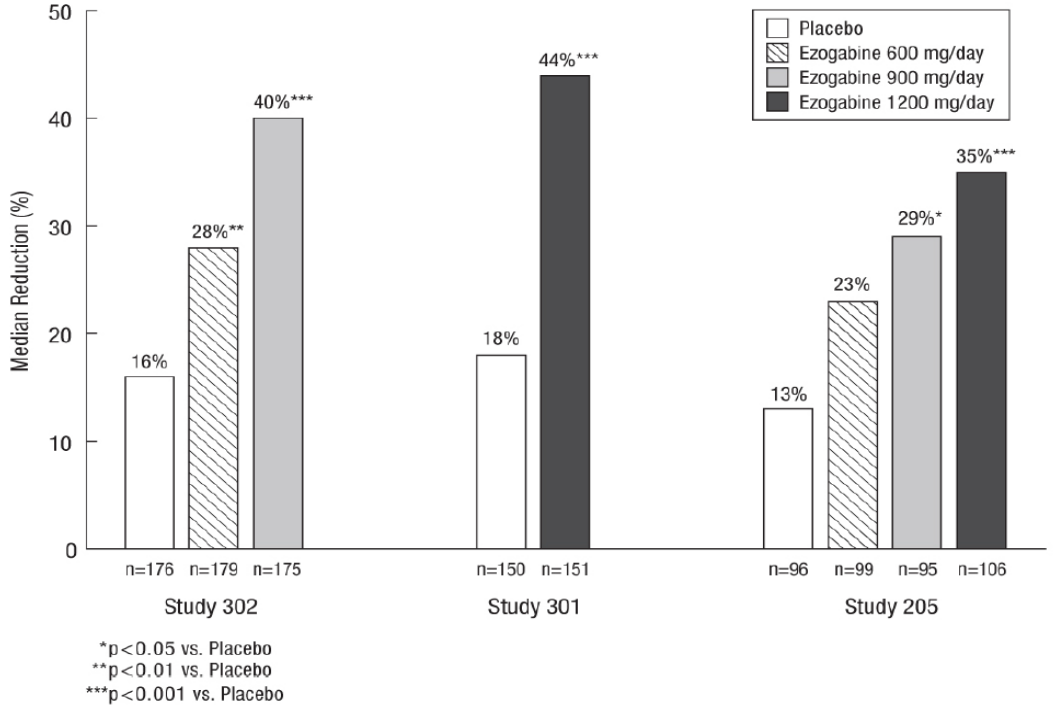
XEN496 Overview: Phase 3 Ready K_V7 Potassium Channel Modulator

- XEN496, active ingredient ezogabine (retigabine)
- Only anti-seizure medication previously approved by FDA with MOA that potentiates K_V7-mediated potassium current
- Proven mechanism in adult focal seizures
- Potential for precision medicine approach to treat KCNQ2-DEE pediatric epilepsy
- Phase 3 ready
  - Regulatory support to conduct small, pivotal trial
  - Novel, pediatric-friendly formulation developed

Proven Mechanism of Action in Adult Epilepsy



Precision Medicine Approach in Pediatric KCNQ2-DEE Patients

About KCNQ2-DEE

- Severe neurodevelopmental disorder caused by dominant negative missense mutations in the KCNQ2 that presents during first week of life
- Recent epidemiology study from Europe reported KCNQ2 birth rate of ~1 in 17,000[1]

[1]Symonds et al. *Brain*, August 2019.

Case Studies Suggest XEN496 is Active in this Often Refractory Disease

Case Study of 11 KCNQ2-DEE Patients *Millichap 2016*	**Medical Record Review/Parent Interviews** *Olson 2017* (8 Families)
Ezogabine use (assessed by the treating physicians and parents) was associated with : • improvement in seizures and/or development in 3 of the 4 patients treated before 6 months of age, and 2 of the 7 patients treated later • 3 of the 4 infants treated before 6 months old were seizure free or occasional seizures <1/week • No serious side effects were observed	Interviews/medical record review of KCNQ2-DEE patients prescribed ezogabine: • Sustained improvement in seizure frequency in 5 of the 6 children with at least weekly seizures • Improvements in development or cognition in all 8 children • Urinary retention/hesitation in 3 patients, but overall well tolerated

"Our hope is that XEN496 could represent a genetically targeted treatment that improves the lives of children living with this debilitating disease."

Jim Johnson, President, KCNQ2 Cure Alliance

New, Proprietary, Pediatric-Friendly Formulation of XEN496

- XEN496 is a granule formulation, packaged as single-dose sprinkle capsules
  - Sprinkle capsules to contain different weights of XEN496 based on patient's weight/targeted drug level
  - Parents/caregivers open the capsules and disperse the granules into the chosen semi-solid or liquid food "carrier"

XEN496 Granules

- PK study in 24 adult healthy volunteers is complete
  - 400 mg dose in fed or fasted states
  - XEN496's absorption and elimination curves comparable to historical PK data for immediate-release ezogabine tablets
  - Results support planned XEN496 Phase 3 trial in KCNQ2-DEE

Typical sprinkle capsule

Clinical Development of XEN496

- Clinical development in KCNQ2-DEE pediatric population
  - ✓ GSK provided right of reference to FDA
  - ✓ FDA granted *Orphan Drug Designation* and *Fast Track* designation
  - ✓ Steering committee of KCNQ2-DEE experts
  - ✓ Letters of support sent to FDA from KOLs, patients, and advocacy groups
  - ✓ Developed a pediatric friendly formulation with novel IP
  - ✓ Completed adult PK study with data supporting plans for Phase 3 trial
- Initiatives to support clinical trial include:
  - Improving access to diagnosis through the Behind the Seizures™ program and other partnerships
  - Patient/caregiver surveys to inform trial design and endpoints

Next Steps

- Initiate Phase 3 clinical trial in 2020*
- Randomized, double-blind, placebo-controlled study
  - ***Anticipated primary endpoint***: median % change in seizure frequency from baseline compared to treatment period of active versus placebo
- ~40 KCNQ2-DEE patients (infants up to six years old)

*Guidance is dependent upon the ability to initiate clinical sites and patient enrollment given the ongoing COVID-19 pandemic.

XEN007: Calcium Channel Modulator

- Active ingredient flunarizine
- CNS calcium channel modulator (Cav2.1 and T-type calcium channels)
- ~30 years' clinical use, including pediatrics; never developed in the U.S.
- Various development strategies and potential indications for XEN007 are under consideration

About Childhood Absence Epilepsy (CAE)

- CAE affects ~10% of children with epilepsy; onset generally between 3 to 13 years old, with a peak at 6 to 7 years old
- 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 (SWDs) on EEG
- Child may have one or many (up to 100) absence seizures per day and have problems with attention and learning

Orphanet; epilepsy.com; Killory et al., 2011; Shinnar et al., 2015; Masur et al., 2013

Physician-Led Phase 2 POC Study in CAE Underway

*Examining XEN007 as an adjunctive treatment in **pediatric patients** diagnosed with treatment-resistant **childhood absence epilepsy***

- Flunarizine shown to be well-tolerated clinically and significantly reduced the number and duration of SWDs on EEG in pre-clinical models of absence seizures
- Results expected in 2020*

*Guidance is dependent upon patient enrollment rates given the ongoing COVID-19 pandemic.

Selective $Na_V1.6$ Inhibitor for Rare Pediatric Epilepsy (NBI-921352)

NBI-921352 (formerly XEN901)

- Neurocrine has exclusive license to XEN901 (now called NBI-921352) and other pre-clinical, selective $Na_V1.6$ inhibitors and dual $Na_V1.2/1.6$ inhibitors for development
- Potent and selective inhibitor to precisely target the sodium channel affected by the genetic mutation of *SCN8A* – $Na_V1.6$
- Initiation of Phase 2 study in SCN8A-DEE anticipated in 2H:2020
  - Xenon eligible to receive up to $25M (cash + equity investment) upon the FDA acceptance of an IND for NBI-921352 expected in mid-2020

About SCN8A-DEE

- Rare form of early-onset epilepsy with occurrence of **seizures** beginning in the first 18 months of life
- **Physical and psychological symptoms** include recurrent seizures of all types, developmental delays, learning difficulties, muscle spasms, poor coordination, sleep problems, and autistic-like features
- **No approved treatments**

$Na_V1.7$ Inhibitor for Post-Operative Pain (FX301)

FX301 (formerly XEN402)

- Flexion Therapeutics, Inc. acquired global rights to develop and commercialize XEN402, a Nav1.7 inhibitor, also known as funapide
- FX301 consists of XEN402 formulated for extended release from a thermosensitive hydrogel intended to support administration as a peripheral nerve block for control of post-operative pain
- Flexion anticipates initiating human clinical trials in 2021
  - A GLP toxicology study with FX301 commenced in April 2020, triggering a $0.5 million milestone payment to Xenon

Supportive FX301 Pre-Clinical Data

- New pre-clinical FX301 data presented on the American Society of Regional Anesthesia and Acute Pain website:
  - FX301 provided sustained, post-operative analgesic effect with no impairment in motor function compared to liposomal bupivacaine and placebo
  - High local concentrations of funapide were measured at the site of administration for the duration of the study, which is consistent with the creation of a depot providing controlled drug release

Multiple Catalysts & Value-Creating Milestone Opportunities

XEN1101

- Phase 2b clinical trial (X-TOLE study) in adult focal seizures ongoing in Canada, U.S. and Europe
- Top-line results anticipated in 1H:2021*
- Evaluating additional potential indications for XEN1101 development

XEN496

- Adult PK study completed / FDA feedback on Phase 3 clinical trial design received in Q2:2020
- Initiation of Phase 3 clinical trial in pediatric KCNQ2-DEE anticipated in 2020*

XEN007

- Physician-led Phase 2 open label study in CAE; results expected in 2020*

Partnered Programs

NBI-921352 (XEN901) / Neurocrine Biosciences

- Neurocrine anticipates filing IND in mid-2020 for clinical trial in SCN8A-DEE, triggering $25M (cash + equity investment) upon FDA acceptance of SCN8A-DEE IND

FX301 / Flexion Therapeutics

- FX301 ($Na_V1.7$ inhibitor for the management of post-operative pain) expected to enter clinical trials in 2021

*Guidance given is dependent upon patient enrollment rates and/or the ability to initiate clinical sites given the ongoing COVID-19 pandemic.

For more information:

investors@xenon-pharma.com