XENON

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Corporate Overview

November 2019

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 our expectations regarding the timing of and results from clinical trials and pre-clinical development activities, including those related to XEN1101, XEN901, XEN496, XEN007 and our other product candidates, the potential efficacy, safety profile, future development plans, addressable market, regulatory success and commercial potential of XEN1101, XEN901, XEN496 ,XEN007 and our other product candidates, the anticipated timing of IND, or IND equivalent, submissions and the initiation of future clinical trials for XEN1101, XEN901, XEN496, XEN007 and our other product candidates, the efficacy of our clinical trial designs, our ability to successfully develop and achieve milestones in the XEN1101, XEN901, XEN496 , XEN007 and other development programs, the design of our clinical trials and anticipated enrollment, and the progress and potential of our other ongoing development programs.

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 promising results in early clinical trials may not be replicated in subsequent clinical trials; clinical trials may not demonstrate safety and efficacy of any of our or our collaborators' 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; the impact of competition; 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

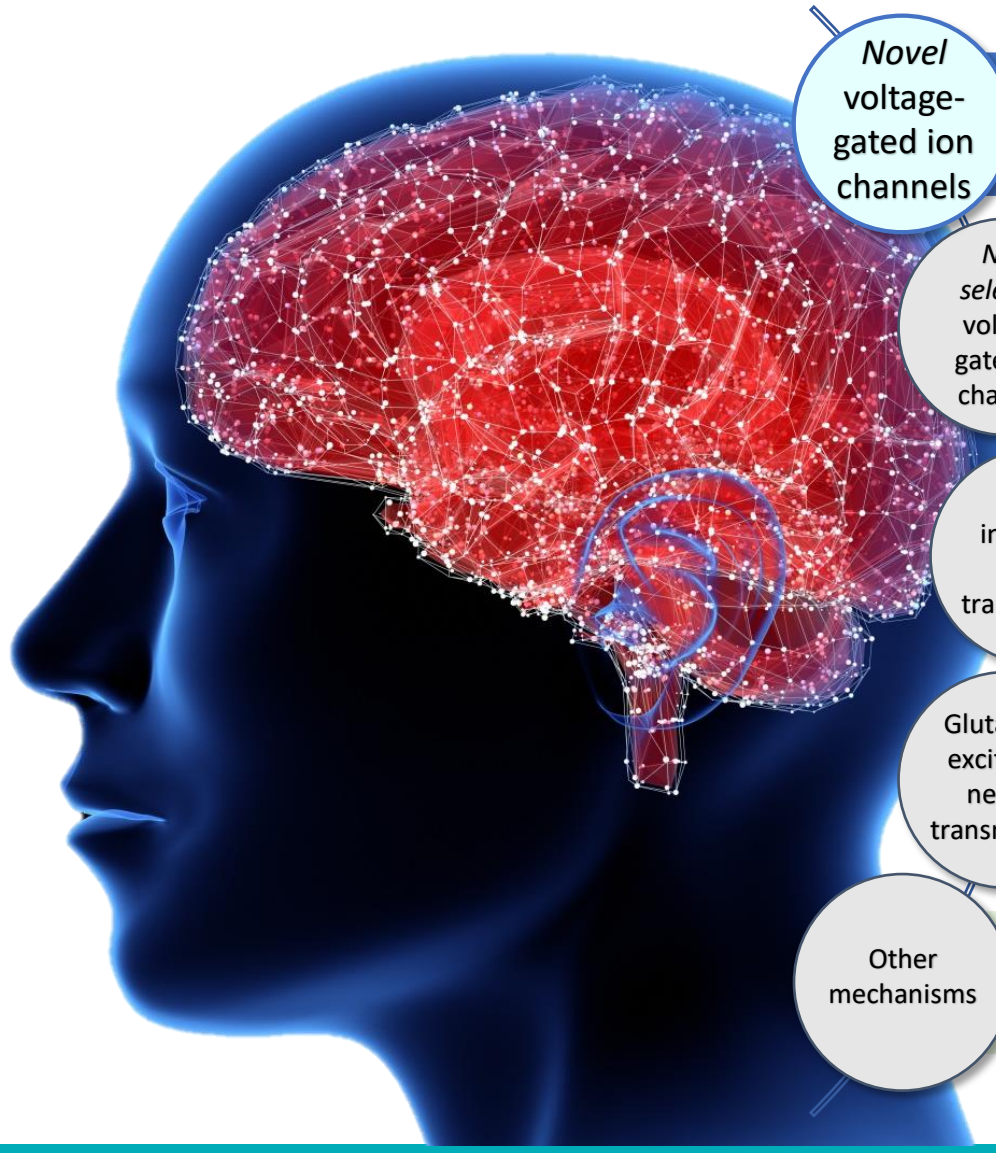
- Small molecule, ion channel neurology-focused biopharma company
  - NASDAQ: XENE
- Multiple mid-to-late stage clinical trials underway or being initiated in the near term
- Strong financial position
  - $94.6M in cash, cash equivalents and marketable securities (as of Sept. 30/19)
    - ~25.8M common shares and 1M preferred shares (as of Sept. 30/19)

Ion Channel, Neurology-Focused Pipeline

Therapeutic Program *Indication*	Pre-clinical	Phase 1	Phase 2	Phase 3
XEN496 (Kv7 Potassium Channel Modulator) *Orphan Pediatric Epilepsy*				
XEN1101 (Kv7 Potassium Channel Modulator) *Adult Focal Epilepsy*				
XEN901 (Nav1.6 Sodium Channel Inhibitor) *Orphan Pediatric Epilepsy or Adult Focal Epilepsy*				
XEN007* (Cav2.1 and T-Type Calcium Channel Inhibitor) *Orphan Neurological Indications*				
Ion Channel Modulators *Orphan Channelopathies*				
$Na_V1.7$ Inhibitors *Pain (Partnered Program)* Genentech A Member of the Roche Group				
FX301 *Post-operative Pain (Partnered Program)* flexion				

*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).

Major Classes of Anti-Epileptic Drugs (AEDs)



Novel voltage-gated ion channels	**XEN496** (Kv7) | **XEN1101** (Kv7) | **XEN901** (Nav1.6)
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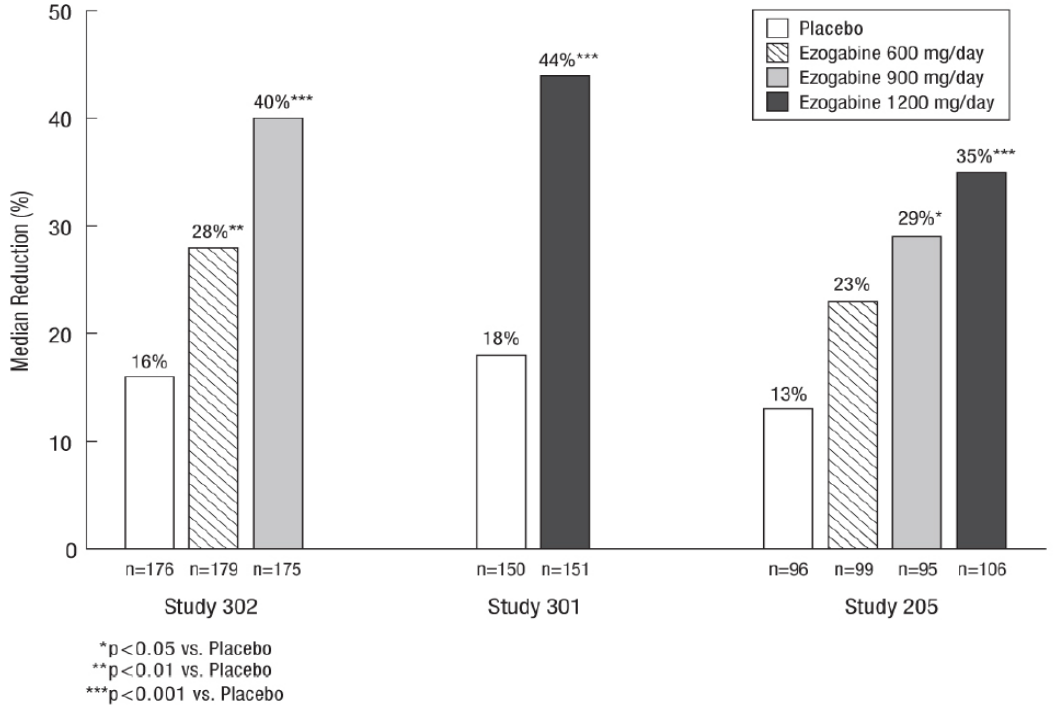
Other mechanisms	Levetiracetam; Brivaracetam; Cannabidiol (CBD)

XEN496: Phase 3 Ready Kv7 Potassium Channel Modulator

- XEN496, active ingredient ezogabine (retigabine)
- Only AED previously approved by FDA with MOA that potentiates Kv7-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
- Presents during first week of life
  - Frequent daily refractory tonic seizures, status common
  - Most often associated with severe developmental delay
  - Present with language/social impairment, outbursts, repetitive and self injurious behaviours
  - Motor disabilities
  - Seizures wane by 4 years of age, can occur in clusters thereafter
- Recent epidemiology study from Europe reported KCNQ-DEE birth rate of ~1 in 17,000 (more common than initially thought)
  - Compared to Dravet Syndrome estimated at ~1 in 12,000-15,000

> “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

Weckhuysen et al. “KCNQ2 encephalopathy: Emerging phenotype of a neonatal epileptic encephalopathy.” *Annals of Neurology*, January 2012.

Symonds et al. “Incidence and phenotypes of childhood-onset genetic epilepsies: a prospective population-based national cohort.” *Brain*, August 2019.

KCNQ2-DEE Patients Treated with Ezogabine - Published Case Reports

Case Study of 11 KCNQ2-DEE Patients *Millichap 2016*	**Medical Record Review/Parent Interviews** *Olson 2017* (8 Families)
Ezogabine associated with improvements in seizures and/or development in: • 3 of the 4 infants treated before 6 month old were seizure free or occasional seizures <1/week • 2 of the 7 treated later • No serious adverse effects	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

Case Studies Suggest XEN496 is Efficacious in this Often Refractory Disease

Pathway to Market for XEN496

- Prove efficacy and safety in KCNQ2-DEE pediatric population
  - ✓ GSK provided right of reference to FDA
  - ✓ Granted orphan drug designation (ODD)
  - ✓ Steering committee of KCNQ2 experts
  - ✓ Letters of support sent to FDA from KOLs, patients, and advocacy groups
  - ✓ Developed a pediatric friendly formulation with novel IP
- Initiatives to support clinical trial include:
  - Improving access to diagnosis through the Behind the Seizures™ program and other diagnostic partnerships
  - Conducting patient/caregiver surveys to inform trial design and FDA interactions
- Next steps:
  - Adult PK study with XEN496
  - Submit IND application in Q1:20 to initiate Phase 3 clinical trial in KCNQ2-DEE

New, Proprietary, Pediatric-Friendly Formulation of XEN496

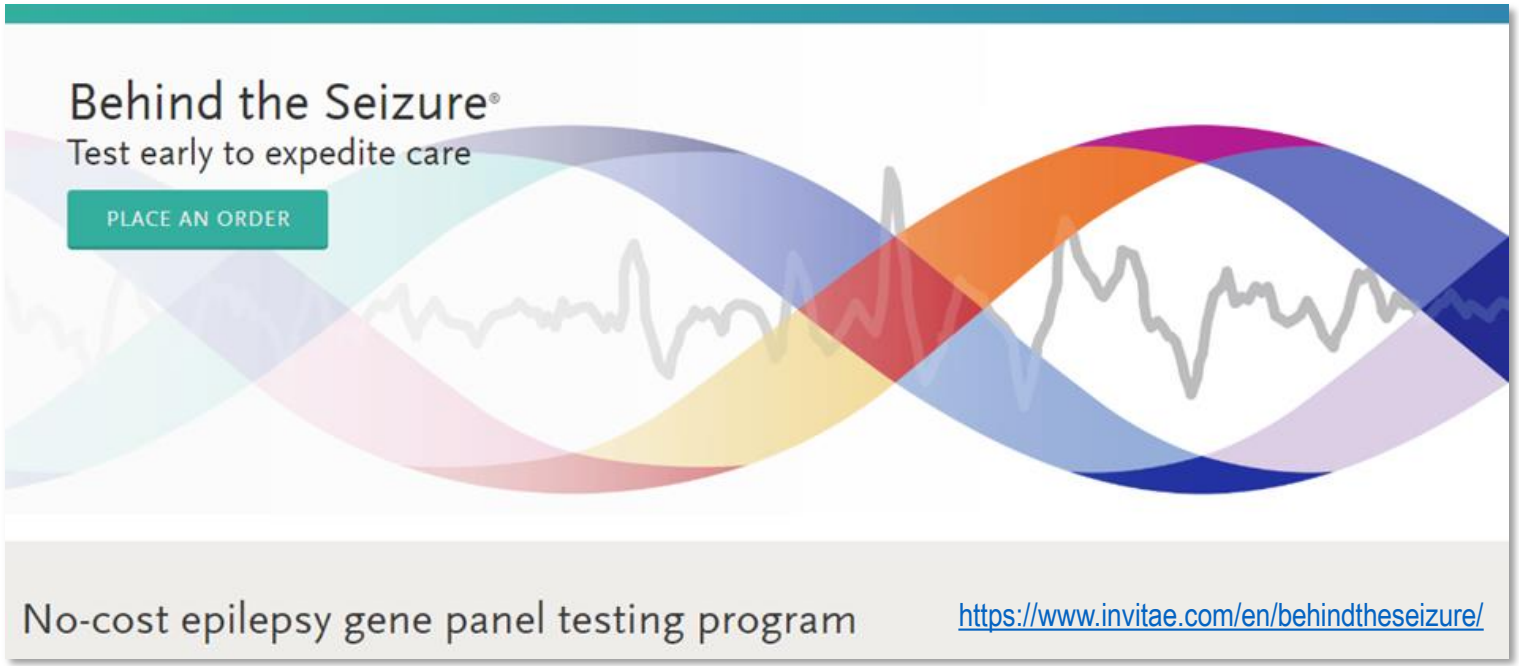
- XEN496 is a granule formulation, packaged as single-dose sprinkle capsules
  - Sprinkle capsules containing different weights of XEN496 will be manufactured based on patient's weight/targeted drug level
- Parents/caregivers open the capsules and disperse the granules into the chosen semi-solid or liquid food "carrier"
- Standard *in vitro* testing has shown that XEN496 acts as an "immediate-release" drug product
- Long term stability studies underway
- XEN496 does not have an objectionable taste or mouthfeel

XEN496 Granules

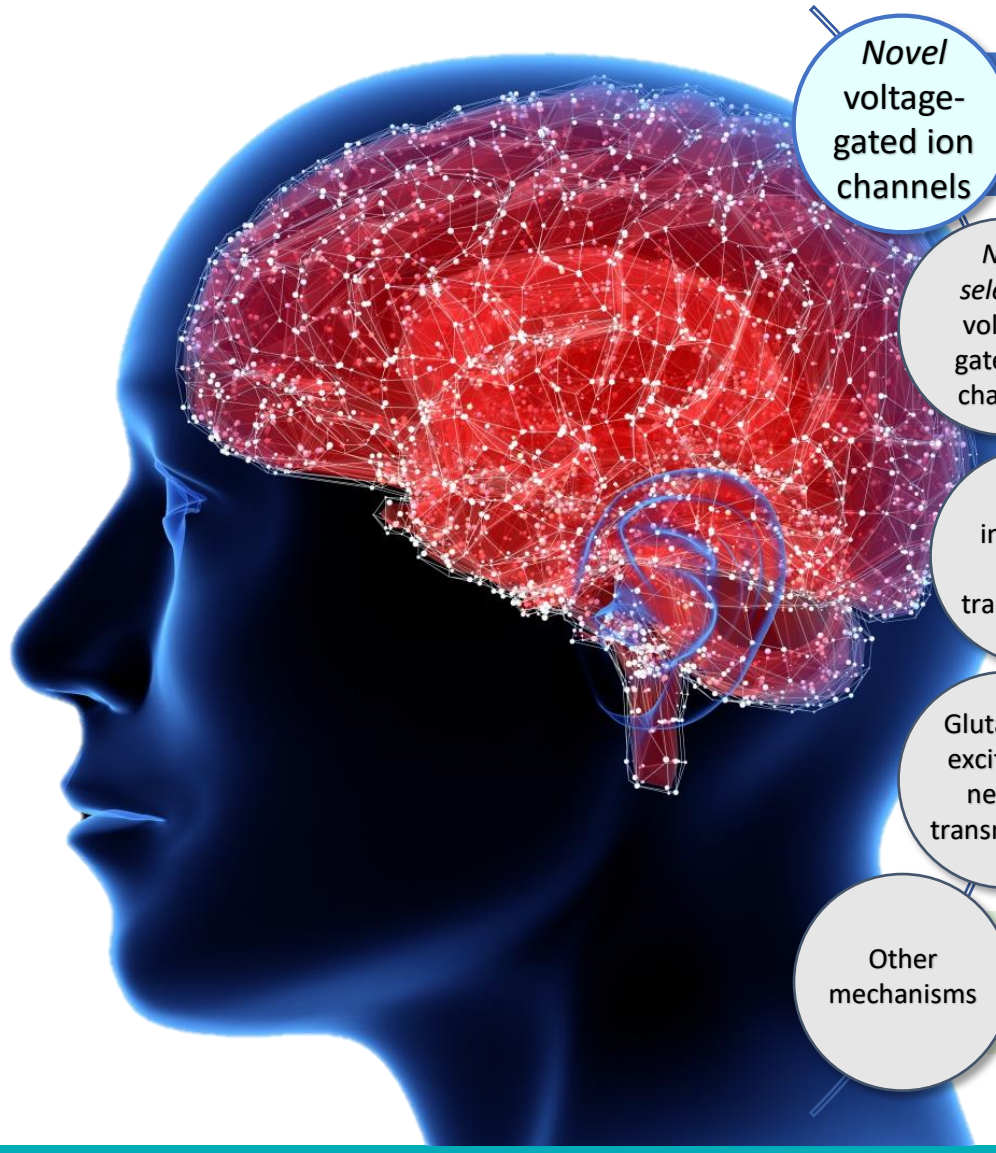
Typical sprinkle capsule

Behind the Seizure™ Program

- Collaboration with Invitae, BioMarin and Stoke
- 187 gene panel
- Offer no-cost testing to all children up to 60 months of age
  - 300+ institutions have participated
  - ~150 tests/month for children 0-2 years old



Major Classes of Anti-Epileptic Drugs (AEDs)



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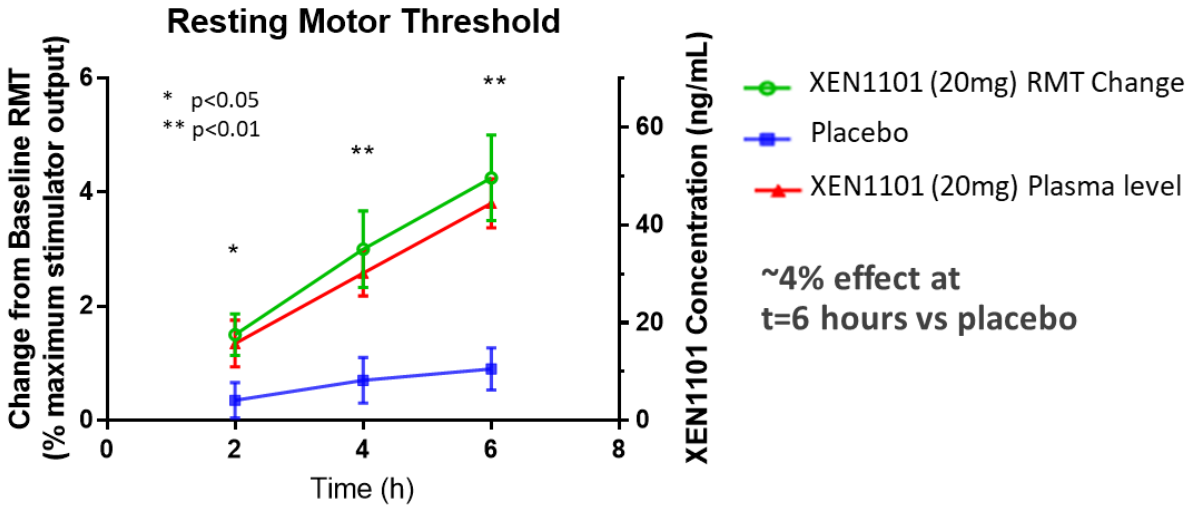
XEN1101: Potential “Next-Gen” Kv7 Potassium Channel Modulator

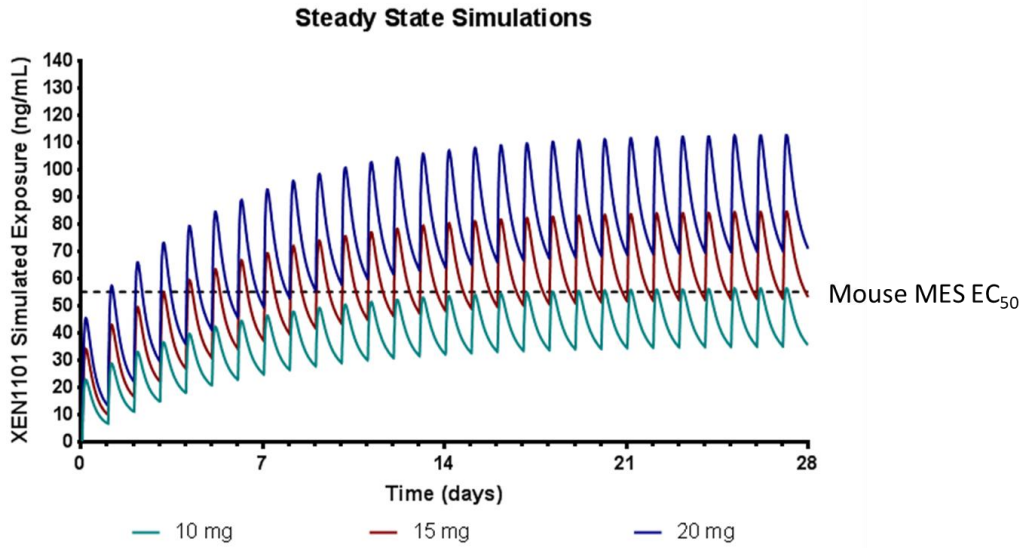
- Same proven MOA in adult focal seizures as ezogabine, but with improvements
  - More potent *in vitro* and *in vivo*
  - Once daily dosing with potential evening administration
  - 3- to 4-fold selective for KCNQ2/3 over other KCNQ channels
  - No pigmented dimers and no predicted discoloration liability
- 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

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
- RMT effect of XEN1101 (20 mg) is ~2x ezogabine at 1/20th the dose (400 mg)



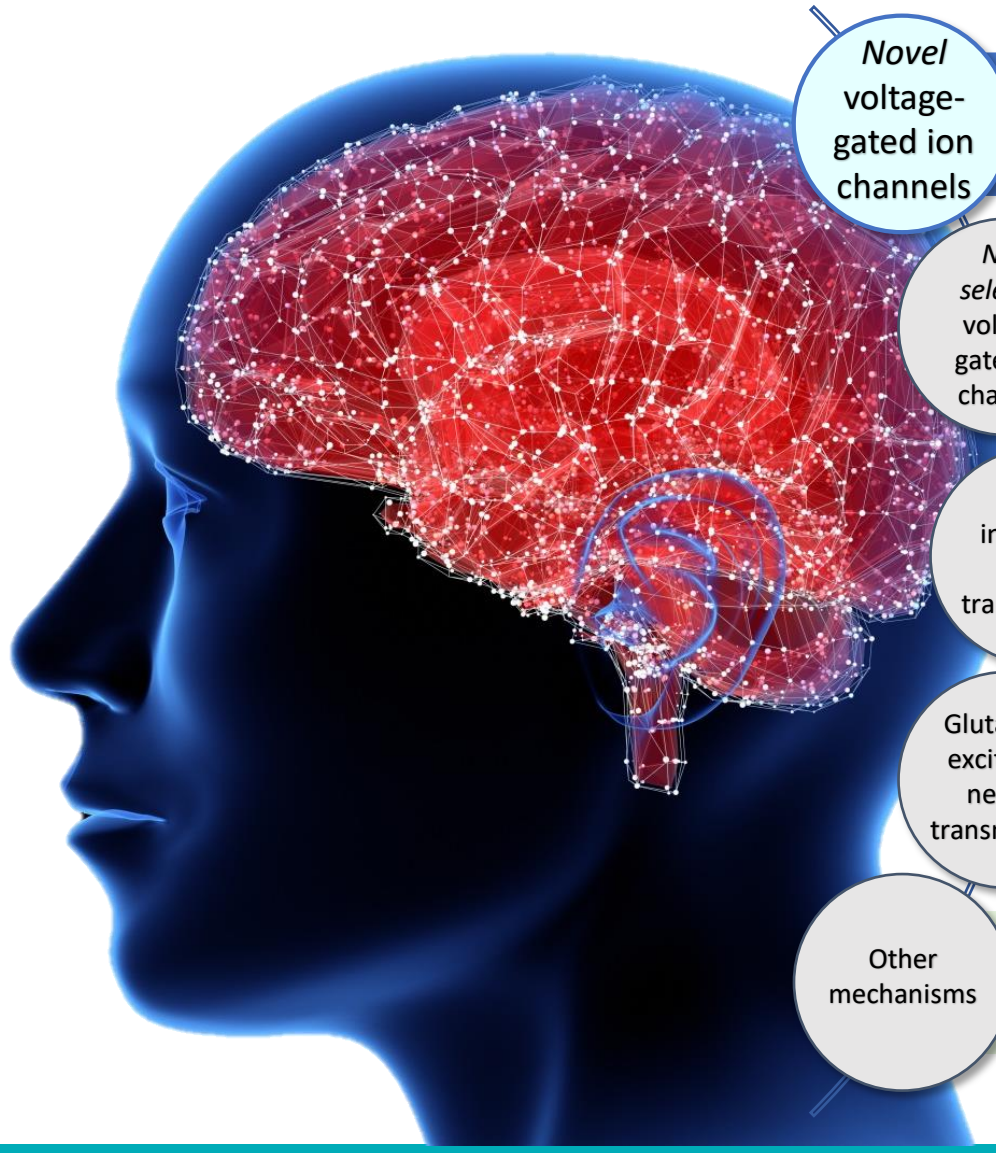


Chronic low daily dosing achieves exposures required for TMS and pre-clinical efficacy

XEN1101 Phase 2b Clinical Trial Underway

- Phase 2b clinical trial 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 now underway in U.S., Canada and Europe
- Long term 6- and 9-month tox studies completed to support planned 12-month OLE
- Topline data expected in 2H:2020

Major Classes of Anti-Epileptic Drugs (AEDs)



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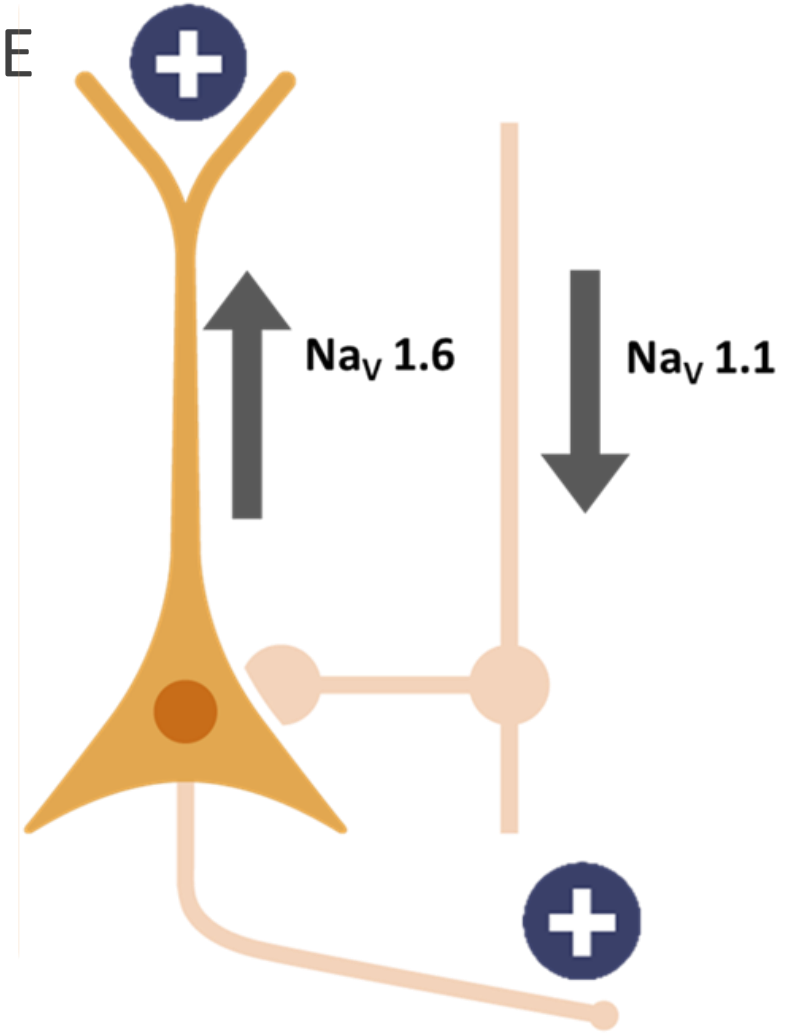
XEN901: A Novel, Unique Sodium Channel Inhibitor

- Precision medicine approach to address SCN8A-DEE and potentially other EIEEs
- Potential to achieve higher levels of seizure freedom with an improved side effect profile
- Favorable PK and safety profile in Phase 1
- Data from pilot TMS study supports cortical and corticospinal effect

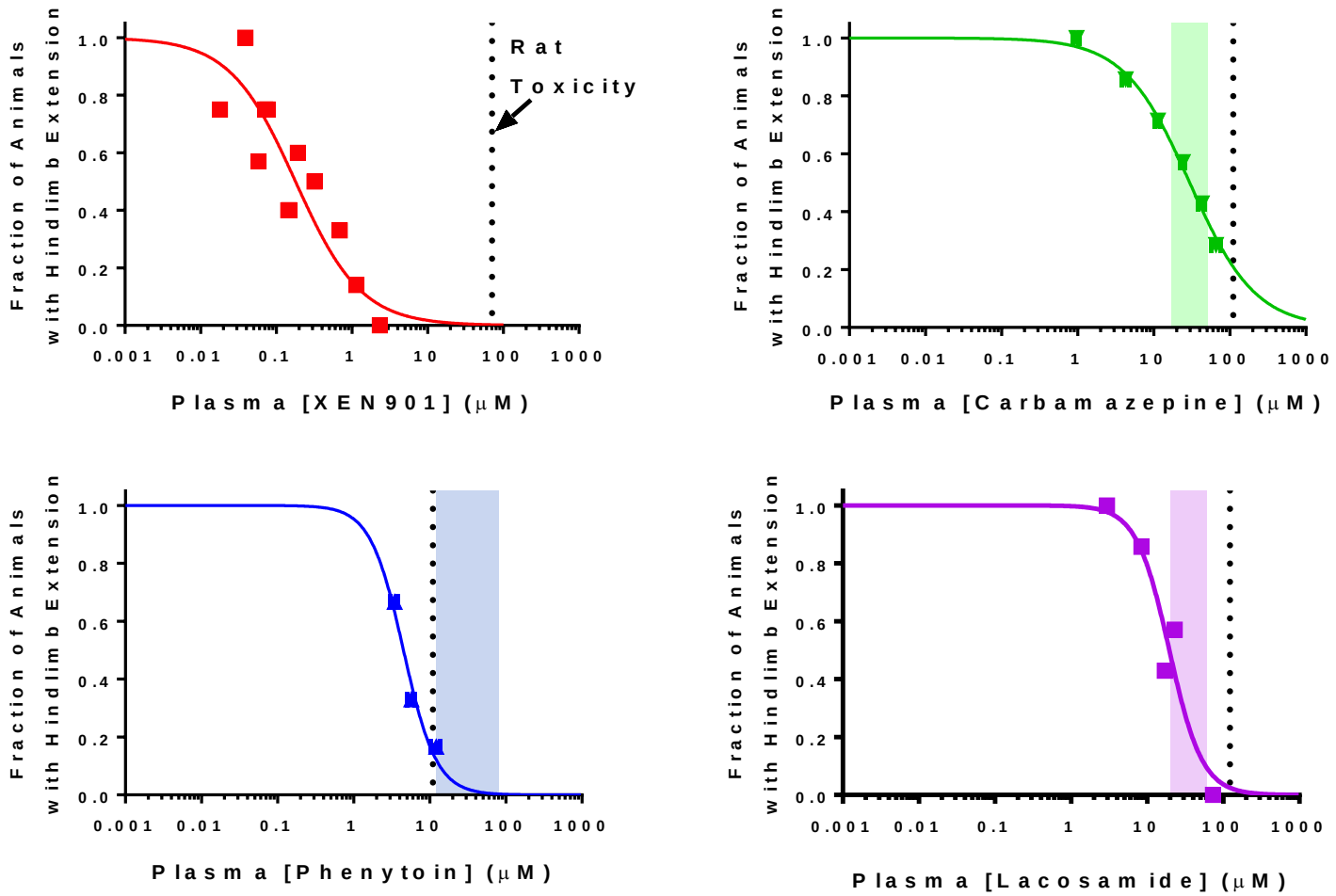


Potential "Best in Class" Sodium Channel Inhibitor

Striving for Seizure Freedom with XEN901

- Current agents lack therapeutic index needed to achieve seizure freedom for many patients
- High therapeutic index of XEN901 could enable high level of efficacy with minimal adverse events

Rat MES Assay



Shaded areas correspond to published clinical plasma concentrations

Summary of Clinical Development of XEN901

- **Phase 1 Results**:
  - Favorable pharmacokinetics (PK) data predicts BID dosing
  - XEN901 overall generally safe and well tolerated at the doses examined
    - Majority of adverse events (AEs) were deemed unrelated to XEN901, and were mild or moderate, transient and resolved spontaneously
    - All AEs considered possibly related to XEN901 were mild; only nausea and dizziness were reported in more than 1 subject
    - No SAEs, deaths, or clinically significant ECG, vital signs or laboratory findings
  - Pilot TMS study showed increases in RMT and AMT of ~2% suggesting activity in target CNS tissue
- **Phase 2 Clinical Planning**:
  - Completed development of a pediatric-specific granule formulation of XEN901
  - Completed juvenile tox studies to support pediatric development activities
  - Stability studies ongoing
  - PK study to test new pediatric formulation ongoing
  - Anticipate filing an IND, or IND equivalent, submission in Q1:20 to initiate Phase 2 or 3 clinical trial in SCN8A-DEE patients
  - Continue to evaluate other potential indications, including adult focal epilepsy

XEN007: Calcium Channel Modulator

- Active ingredient flunarizine
- Safe CNS calcium channel modulator (Cav2.1 and T-type calcium channels)
- ~30 years' clinical use, including pediatrics; never developed in the U.S.
- Xenon has obtained:
  - ODD for hemiplegic migraine (HM) and alternating hemiplegia of childhood (AHC) and a Rare Pediatric Disease Designation for AHC
  - Exclusive access to regulatory files and reports for U.S. territories
  - Commercial drug product supply
- Various development strategies and potential indications for XEN007 are under consideration

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 has demonstrated efficacy in pre-clinical models of absence seizures
- Results expected in 2020

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 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

Multiple Catalysts & Value-Creating Milestones

XEN496

- Adult PK study with pediatric-specific formulation
- Submit IND in Q1:20 to enable pivotal Phase 3 trial in pediatric KCNQ2-DEE

XEN1101

- Phase 2b clinical trial (n=300) in adult focal seizures underway in Canada, U.S. and Europe
- Top-line results anticipated in 2H:2020

XEN901

- Adult PK study with pediatric-specific formulation
- Submit IND, or IND equivalent, in Q1:20 to start proposed Phase 2 or 3 clinical trial in SCN8A-DEE

XEN007

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

For more information:

investors@xenon-pharma.com