CORPORATE OVERVIEW

SEPTEMBER 2021

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, FX301, 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, FX301, 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 expansion of the X-TOLE clinical trial and the anticipated timing of the topline data therefrom; the progress and potential of our other ongoing development programs; the potential receipt of milestone payments and royalties from our collaborators; 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, including impact on our clinical trial sites, collaborators, and contractors who act for or on our behalf, 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 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; impact of new or changing laws and regulations; 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

- Neurology-focused biopharma company
  - Leaders in small molecule, ion channel drug development
- Multiple mid- to late-stage clinical trials underway
- Strong financial position
  - $260.5 million in cash, cash equivalents and marketable securities (as of June 30, 2021)
  - 41,117,568 common shares, 1,081,081 pre-funded warrants and 1,016,000 preferred shares

Xenon's Ion Channel, Neurology-Focused Pipeline

Therapeutic Program *Indication*	Pre-clinical	Phase 1	Phase 2	Phase 3
XEN496 (Potassium Channel Modulator) *Orphan Pediatric Epilepsy (EPIK Clinical Trial)*				
XEN1101 (Potassium Channel Modulator) *Adult Focal Epilepsy (X-TOLE Clinical Trial)*				
XEN1101* (Potassium Channel Modulator) *Major Depressive Disorder (MDD) /Mount Sinai Collaboration*				
XEN007* (Calcium Channel Inhibitor) *Pediatric Absence Epilepsy*				
Ion Channel Modulators *Orphan Channelopathies*				
NBI-921352 (XEN901) and $Na_V1.6/1.2$ Sodium Channel Inhibitors *Epilepsy (Pediatric SCN8A and Adult Focal-Onset) / Neurocrine Biosciences*				
FX301 (Topical $Na_V1.7$ Inhibitor) *Post-operative Pain / Flexion Therapeutics*				

*Investigator Sponsored Phase 2 Proof-of-Concept Study

XEN1101 is a Novel "Next-Gen" K_V7 Channel Modulator

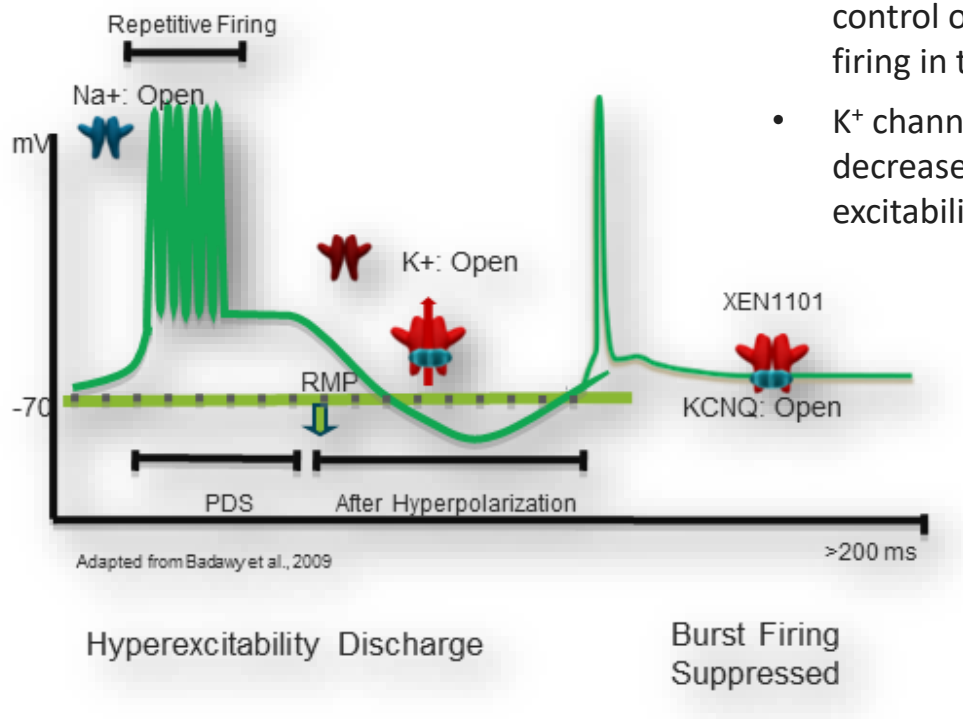
Potential **"only-in-class"** **K_V7 potassium** channel modulator to treat adult focal seizures

Addresses limitations of first-gen K_V7 modulator, ezogabine

Novel MOAs needed for rational **polypharmacy approach**

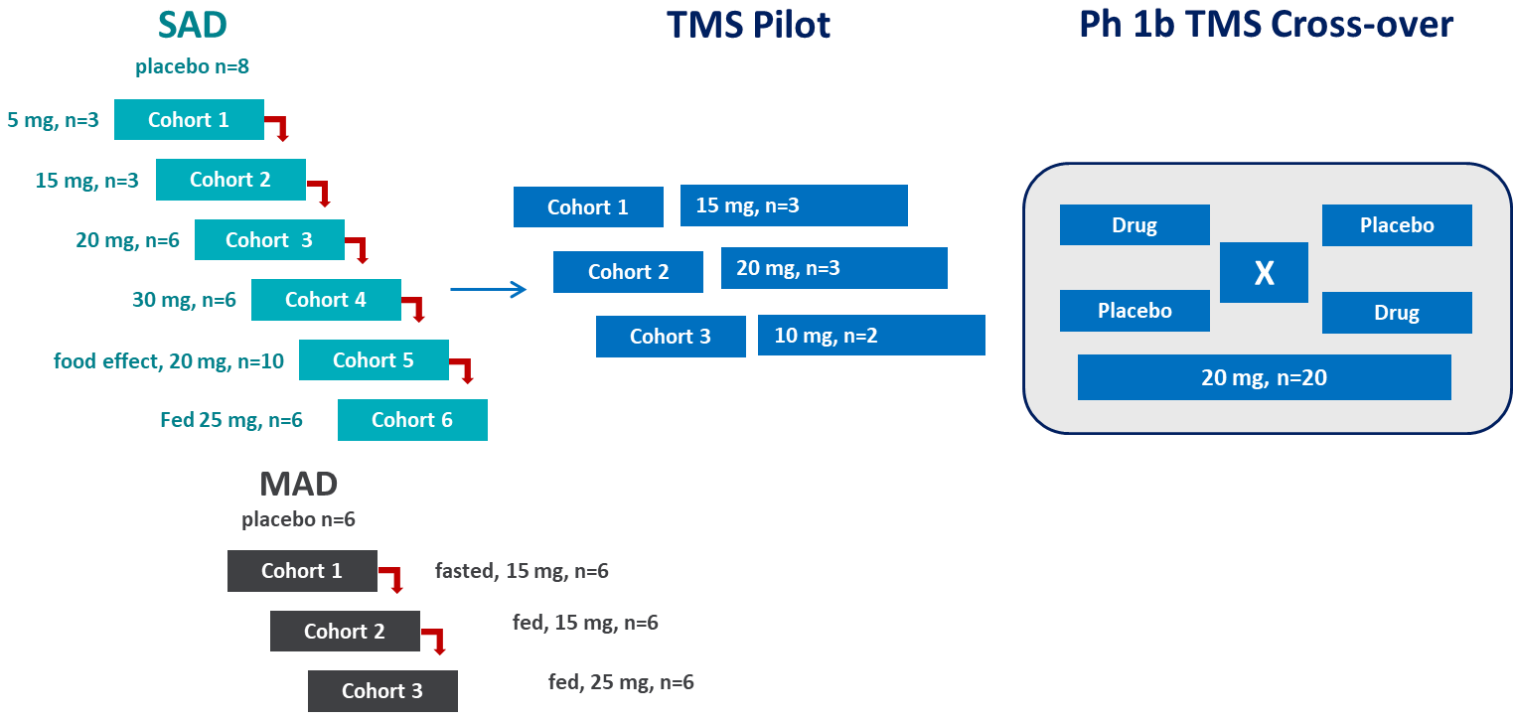
Potential efficacy for **common comorbidity** of depression

Mechanism of Action Previously Validated with Ezogabine



- K+ channels have important inhibitory control over neuronal firing in the CNS
- K^+ channel potentiator decreases hyper-excitability in the brain

XEN1101 Phase 1 Adaptive Integrated Design

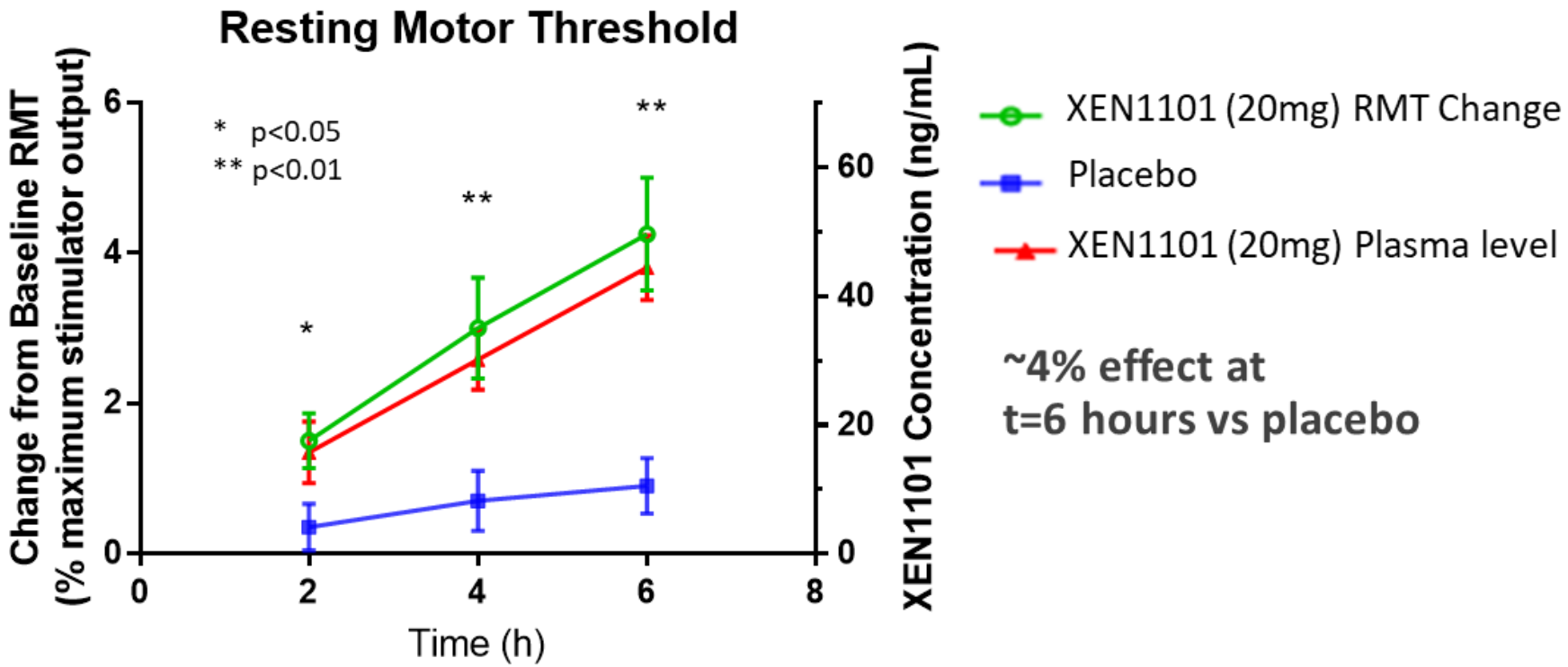


Summary of Single and Multiple Dose Findings

- XEN1101 has a PK profile consistent with QD
  - Long terminal elimination half life
- Near steady-state within 1 week, full steady-state within 3 weeks
- Absorption is enhanced by food
- Exposure increased dose proportionally (15 - 25 mg QD) in fed state
- Low inter-individual PK variability with repeat dose
- AE profile consistent with CNS MOA (e.g., dizziness, sedation, blurred vision)
  - No signal of urinary retention
    - Post-void residual volume normal (bladder ultrasound)
    - Minimal renal excretion of drug
  - No safety signals in ECG or safety labs; no SAEs

Summary of XEN1101 Phase 1b TMS Results

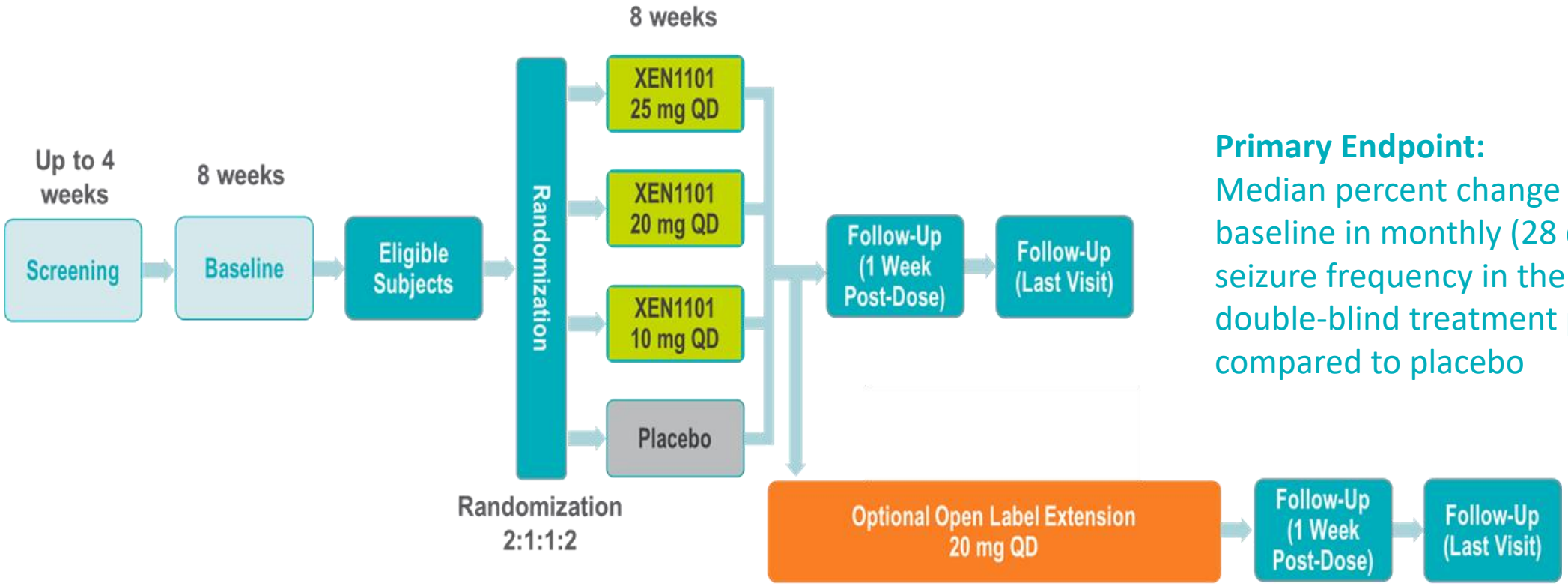
- TMS was used to evaluate the corticospinal and cortical activity profile of XEN1101 compared to placebo in healthy male volunteers



Significant plasma concentration dependent reduction of corticospinal (RMT) and cortical (TEP) excitability

X-TOLE Phase 2b Clinical Trial Underway

- Randomized, placebo-controlled Phase 2b clinical trial in ~300 adult patients with focal epilepsy



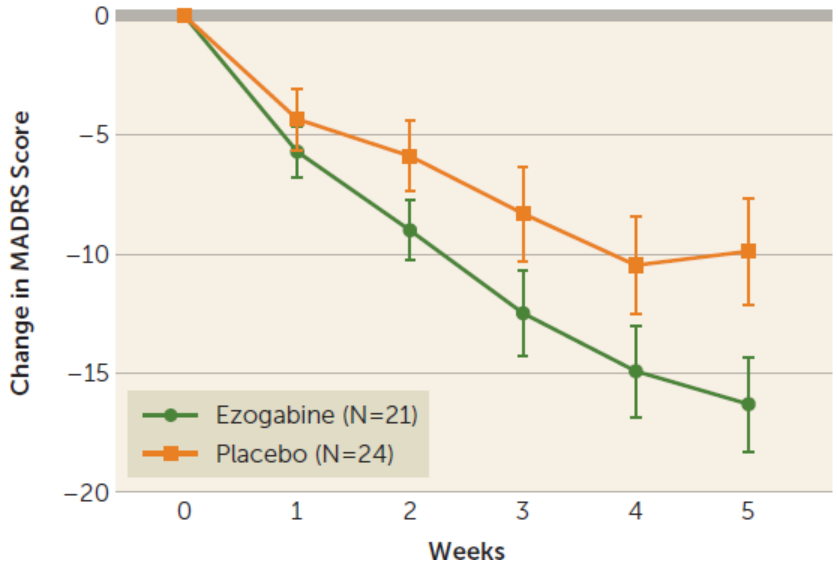
Primary Endpoint:
Median percent change (MPC) from baseline in monthly (28 days) focal seizure frequency in the 8-week double-blind treatment period compared to placebo

Randomization of 326 patients completed in late June; topline data expected in late September to mid-October 2021

XEN1101 Phase 2 POC Studies in Major Depressive Disorder

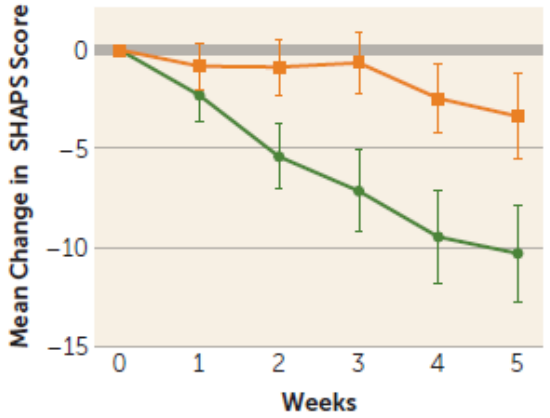
- Promising clinical results with ezogabine when targeting KCNQ channels as a treatment for Major Depressive Disorder (MDD)

Montgomery–Åsberg Depression Rating Scale



Ezogabine, compared with placebo, was associated with a large improvement in depression as measured by the Montgomery-Åsberg Depression Rating Scale (MADRS score change: -7.9±3, p<.001)

Snaith-Hamilton Pleasure Scale



Compared with placebo, ezogabine was associated with a large improvement in hedonic capacity as measured by the Snaith-Hamilton Pleasure Scale (SHAPS score change: -6.9±3.2, p<.001)

- Anticipate initiation of investigator-led (Mount Sinai) Phase 2 proof-of-concept clinical trial of XEN1101 for treatment of MDD and anhedonia in the coming months
- In parallel, Xenon is planning a company-sponsored clinical study in MDD based on encouraging pre-clinical data with XEN1101

Costi et al., "Impact of the KCNQ2/3 Channel Opener Ezogabine on Reward Circuit Activity and Clinical Symptoms in Depression: Results from a Randomized Controlled Trial." *Am J Psychiatry*. 2021.

Summary: XEN1101's Differentiated Profile in Adult Focal Epilepsy

One pill, once-daily

No titration required

Unique MoA and low DDI risk can be leveraged in rational polypharmacy

Forgiving PK provides coverage for missed doses

Proven anti-seizure MoA

Broad efficacy in multiple pre-clinical seizure models

May provide mood benefit beyond seizure control

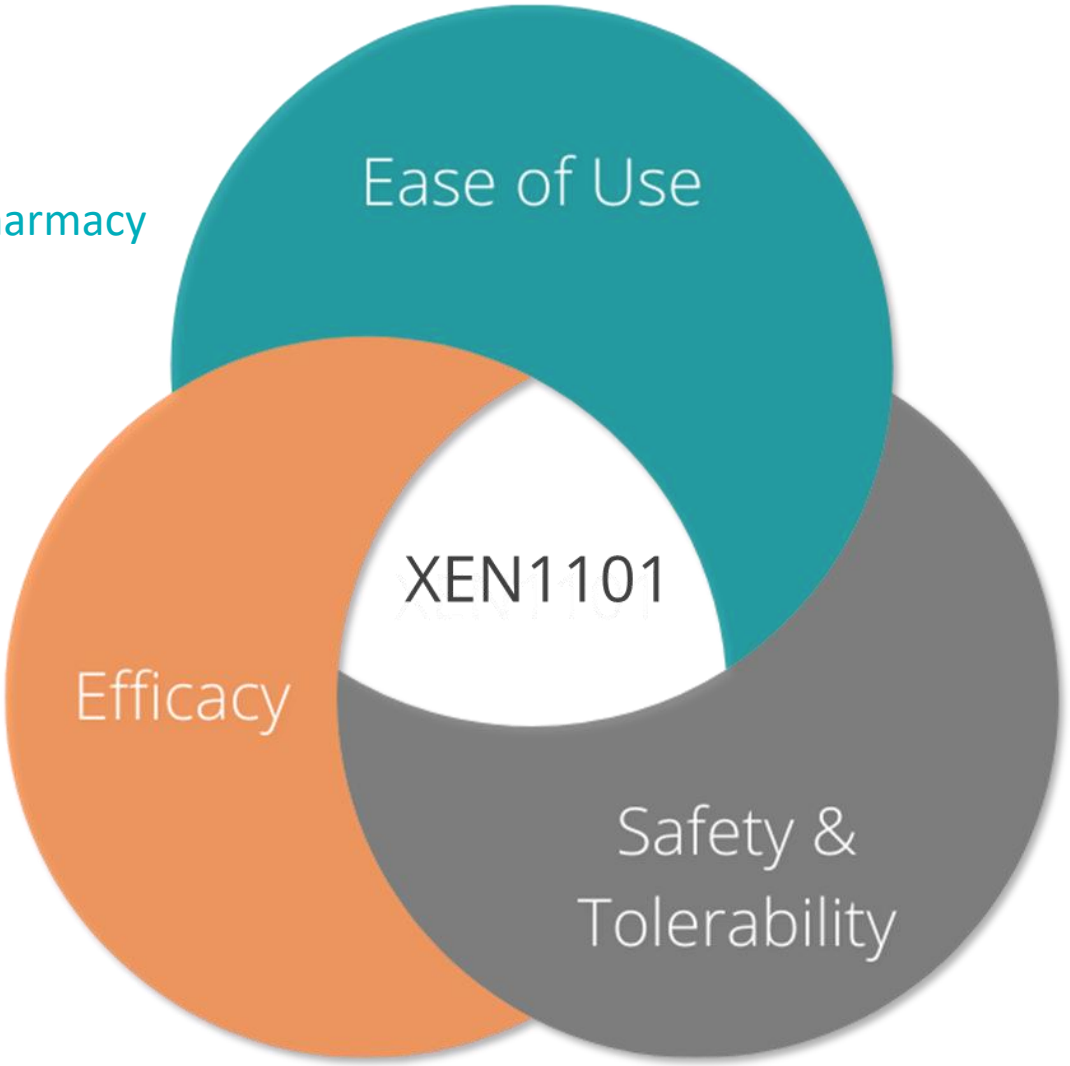
Strong TMS engagement

Well-tolerated with low drop-out rates in P2b

Not expected to exacerbate psych co-morbidities

Evening dose = C_{max} during sleep

No drug allergic reactions observed



About KCNQ2-DEE

- Rare, severe neurodevelopmental disorder caused by dominant negative missense mutations in the KCNQ2 that presents during first week of life with estimated KCNQ2 birth rate of ~1 in 17,000 (Symonds et al. 2019)

Summary of Published Case Reports of KCNQ2-DEE Patients Treated with Ezogabine

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

Case studies suggest ezogabine is active in this often-refractory disease

XEN496: Potential Precision Medicine Approach for KCNQ2-DEE

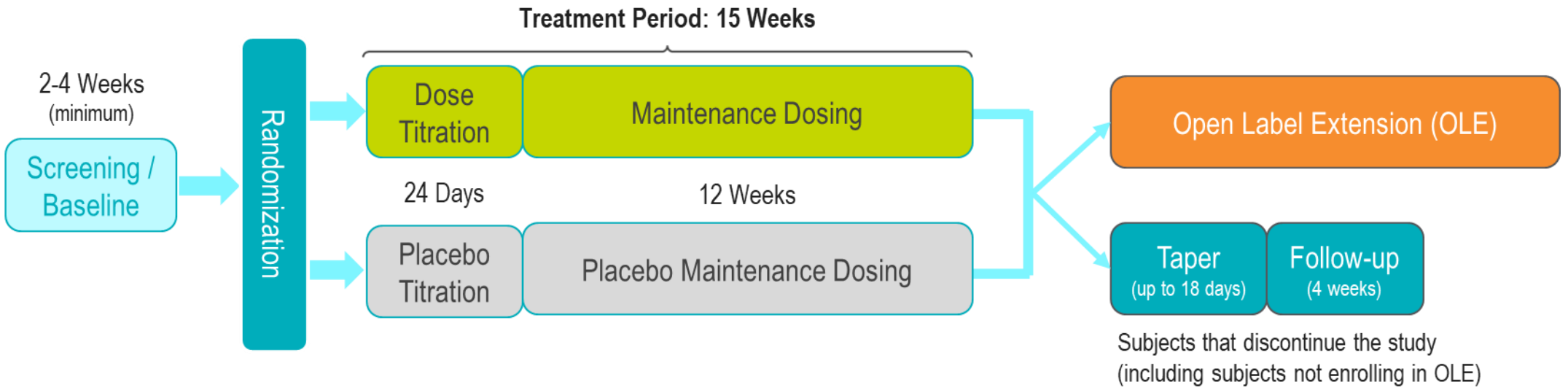
Development of Proprietary XEN496

- XEN496 is pediatric-specific, granule formulation of ezogabine to be presented as sprinkle capsules
- MOA that potentiates Kv7-mediated potassium current
- Ezogabine previously approved by FDA with proven mechanism in adult focal seizures
- Potential for precision medicine approach to treat rare KCNQ2-DEE pediatric epilepsy
- Fast Track designation and Orphan Drug Designation in U.S. and Orphan Medicinal Product Designation (Europe)

"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

EPIK Phase 3 Clinical Trial Underway



Primary Objective: To evaluate the efficacy of XEN496 as adjunctive therapy in reducing seizure frequency from baseline, compared to placebo in pediatric subjects with KCNQ2-DEE

XEN007: Calcium Channel Modulator

- XEN007: active ingredient flunarizine, a CNS calcium channel modulator (Cav2.1 and T-type calcium channels) with ~30 years of clinical use, including pediatrics, but never developed in the U.S.

Physician-Led Phase 2 POC Study Underway

Examining XEN007 as an adjunctive treatment in pediatric patients diagnosed with treatment-resistant absence seizures, including childhood absence epilepsy (CAE) and juvenile absence epilepsy (JAE)

- Promising interim data collected from a small number of patients was presented at AES2020 in December 2020
  - XEN007 resulted in > 50% reduction in diary recorded seizures in all 3 CAE patients, with 2 showing >80% reduction
  - EEGs for the three CAE subjects showed resolution of absences for 2 subjects and 94% reduction for one
- Additional results from a larger data set are anticipated by the end of this year, which will help inform the future development of XEN007

More about CAE

- Affects ~10-17% of children with epilepsy; onset between 4 -10 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

Partnered Programs

NBI-921352 (formerly XEN901)

- Clinical stage selective $Na_V1.6$ sodium channel inhibitor with potential in SCN8A-DEE and other forms of epilepsy
- Neurocrine Biosciences has exclusive license to NBI-921352 and other $Na_V1.6$ & dual $Na_V1.2/1.6$ inhibitors for development
- Anticipate initiation of Phase 2 trial in adolescent SCN8A-DEE patients in 2H:2021 followed by amendment to protocol to include younger patients
- Neurocrine also expects to initiate a Phase 2 trial later this year for the treatment of focal-onset seizures in adults
- Xenon received $10M regulatory milestone in September 2021; potential for additional $15M for next IND-related milestone, as well as other potential collaboration milestone payments and future sales royalties

FX301 (formerly XEN402)

- Flexion has the global rights to develop and commercialize XEN402, now known as FX301, a $Na_V1.7$ inhibitor
- Consists of XEN402 formulated for extended release from a thermosensitive hydrogel
- Initial development of FX301 is intended to support administration as a peripheral nerve block for control of post-operative pain
- Flexion anticipates topline data from Phase 1b proof-of-concept clinical trial of popliteal fossa block with FX301 in patients undergoing bunionectomy in late 2021
- Xenon is eligible to receive certain clinical, regulatory, and commercial milestone payments, as well as future sales royalties

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
- Randomization of 326 patients completed in June; topline results anticipated late Sept. to mid-Oct. 2021
- Mount Sinai investigator-sponsored Ph2 POC in MDD to be initiated in near-term, and planning underway for a company-sponsored clinical study in MDD

XEN496

- Phase 3 clinical trial (EPIK study) in pediatric KCNQ2-DEE underway

XEN007

- Investigator-led Phase 2 open label study in pediatric absence seizures; results from larger cohort by year-end

Partnered Programs

NBI-921352 (XEN901) / Neurocrine Biosciences

- Neurocrine anticipates initiation of Phase 2 clinical trials in 2021
- $10M regulatory milestone achieved in Sept. 2021 and potential $15M for next regulatory milestone

FX301 / Flexion Therapeutics

- Topline results for FX301 Phase 1b POC clinical trial anticipated in late 2021

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