Important Notices

Forward-Looking Statements

Zogenix cautions you that statements included in this presentation that are not a description of historical facts are forward-looking statements. Words such as “believes,” “anticipates,” “plans,” “expects,” “indicates,” “will,” “intends,” “potential,” “suggests,” “assuming,” “designed” and similar expressions are intended to identify forward-looking statements. These statements include statement regarding Fintepla providing a treatment option for patients with Dravet syndrome; Zogenix's plans to commercialize Fintepla; the timing of review by the European Medicines Agency and the Japanese regulatory authority; Fintepla's potential as a treatment for seizures associated with Lennox-Gastaut syndrome (LGS); Zogenix's plans to finalize the studies and data required to support an sNDA for Fintepla in LGS; the timing of regulatory submissions and meetings or other interactions with regulatory agencies related to Fintepla and MT1621; and Zogenix's expectations of the completion, timing and size of the proposed offering. These statements are based on Zogenix's current beliefs and expectations. The inclusion of forward-looking statements should not be regarded as a representation by Zogenix that any of its plans will be achieved. Actual results may differ from those set forth in this presentation due to the risks and uncertainties inherent in Zogenix’s business, including, without limitation: the top-line data Zogenix has reported is based on preliminary analysis of key efficacy and safety data, and such data may change following a more comprehensive review of the data related to the clinical trial and such top-line data may not accurately reflect the complete results of a clinical trial, and the U.S. Food and Drug Administration (FDA) may not agree with Zogenix’s interpretation of such results; later developments with the FDA that may be inconsistent with feedback received at prior meetings with the FDA; unexpected adverse side effects or inadequate therapeutic efficacy of Fintepla that could limit approval and/or commercialization, or that could result in recalls or product liability claims; and other risks described in Zogenix’s public periodic filings with the U.S. Securities and Exchange Commission. You are cautioned not to place undue reliance on these forward-looking statements, which speak only as of the date hereof, and Zogenix undertakes no obligation to revise or update this presentation to reflect events or circumstances after the date hereof. All forward-looking statements are qualified in their entirety by this cautionary statement. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.

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About Zogenix

A global pharmaceutical company focused on serious rare diseases

- **Fintepla® (ZX008)** lead product designed for the treatment of seizures associated with rare, catastrophic, difficult-to-treat epilepsies
- **MT1621** product candidate in late-stage development for TK2d, a rare, often fatal mitochondrial deficiency disease
- Headquarters in the San Francisco Bay Area
- Additional operations in Europe; partnership in Japan
- 180+ employees
- ~$390M cash and marketable securities on June 30, 2020; no debt on balance sheet
# Zogenix Product Portfolio

<table>
<thead>
<tr>
<th>Project &amp; Indication</th>
<th>Preclinical</th>
<th>Phase 1</th>
<th>Phase 2</th>
<th>Phase 3</th>
<th>Regulatory</th>
<th>Commercialization</th>
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<tbody>
<tr>
<td><strong>Rare, Treatment-Resistant Epilepsies</strong></td>
<td></td>
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<tr>
<td>Dravet syndrome – United States</td>
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<td>Approved on 06/25/20; Launched on 07/27/20</td>
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<tr>
<td>Dravet syndrome – Europe</td>
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<td>Under EMA review</td>
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<tr>
<td>Dravet syndrome – Japan</td>
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<td>Positive Phase 3 Trial</td>
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<td>Lennox-Gastaut syndrome</td>
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<td></td>
<td>Positive Phase 3 Trial</td>
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<tr>
<td>Other Rare Epilepsies</td>
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<td>P2 planned Q1 2021</td>
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<tr>
<td><strong>Thymidine Kinase 2 Deficiency (TK2d)</strong></td>
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<tr>
<td>MT1621 Substrate enhancement therapy</td>
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<td>TK2d Potential registration trial</td>
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<tr>
<td><strong>ZX008</strong> fenfluramine oral solution</td>
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Fintepla® | Dravet Syndrome
Lennox-Gastaut Syndrome
Additional Rare, Treatment-Resistant Epilepsies
Fintepla® (ZX008, fenfluramine oral solution)

- FDA approved on June 25th, 2020 for the treatment of seizures associated with Dravet syndrome in patients 2 year of age and older
- Three positive pivotal global Phase 3 trials completed
- Primary endpoint and all key secondary endpoints met with high statistical significance
- Ongoing OLE has shown robust efficacy results up to two years
  - MAA under active review, EU CHMP opinion anticipated in Q4 2020

Dravet syndrome

Lennox-Gastaut syndrome

Other Treatment-Resistant Epilepsies

- Positive pivotal global Phase 3 trial completed
- Top-line results announced Q1 2020
- Primary endpoint met with high statistical significance at 0.7 mg/kg dose; several key secondary endpoints met with statistical significance
  - sNDA submission planned in the U.S. for Q2 2021
  - Phase 2 “basket” efficacy study in multiple rare, difficult to treat epilepsies planned
  - Potential Indications include CDKL5 deficiency disorder, Doose syndrome, Dup15q syndrome, Tuberous Sclerosis Complex, mutations in PCDH19 gene, mutations in Na+ channel genes, and Dravet syndrome 1 to <2 years old
Dravet Syndrome

A rare and frequently catastrophic epilepsy

Affects 1/15,700 live births in the U.S. (about 200-250 infants/year)

Highly treatment resistant
  o 72% of patients take 3 or more antiepileptic drugs
  o >80% of patients are inadequately controlled on traditional antiepileptic treatment regimens

Significant morbidity
  o High seizure frequency; significant developmental and motor impairments

Increased risk of death
  o 6x-higher risk of death, most commonly due to SUDEP (sudden unexpected death in epilepsy) and Status Epilepticus
  o 46% of caregivers use pulse-ox at night due to fear of death from SUDEP

Urgent need to reduce or eliminate high seizure burden and associated comorbidities

Dravet Syndrome Phase 3 Trial Efficacy Results

Primary Endpoint Analysis of % Difference From Placebo in Reduction in Mean Monthly Convulsive Seizures

**Study 1**

<table>
<thead>
<tr>
<th>Dose</th>
<th>% Difference</th>
<th>p-value</th>
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</thead>
<tbody>
<tr>
<td>0.7 mg/kg/day</td>
<td>70.0%</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>0.2 mg/kg/day</td>
<td>31.7%</td>
<td>p=0.043</td>
</tr>
</tbody>
</table>

**Study 2**

<table>
<thead>
<tr>
<th>Dose</th>
<th>% Difference</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.4 mg/kg/day</td>
<td>59.5%</td>
<td>p&lt;0.001</td>
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</tbody>
</table>

p-value compared to placebo
Source: Fintepla prescribing information

1. All 0.4 mg/kg/day patients were also taking concomitant stiripentol, which increases the exposure of Fintepla.
Additional Study 1 and Study 2 Efficacy Results

**Study 1**

Proportion of Patients by Category of Seizure Response for FINTEPLA and Placebo in Patients with Dravet Syndrome (Study 1)

Cumulative 50% or >

0.7 mg/kg 70.0%  
0.2 mg/kg 34.2%  
Placebo 7.7%

**Study 2**

Proportion of Patients by Category of Seizure Response for FINTEPLA and Placebo in Patients with Dravet Syndrome (Study 2)

Cumulative 50% or >

0.4 mg/kg 53.3%  
Placebo 4.8%

Median Longest Interval Between Convulsive Seizures in Patients with Dravet Syndrome (Study 1)

- Placebo 7.8
- 0.2 mg/kg/day 12.9
- 0.7 mg/kg/day 20.5

p-value compared to placebo

Source: Fintepla prescribing information
All 0.4 mg/kg/day patients were also taking concomitant stiripentol, which increases the exposure of Fintepla.
Dravet Syndrome Study 1503 Open Label Extension Study

Robust seizure reduction sustained in OLE at up to two years with no evidence of waning therapeutic effect

Median Change in Monthly Convulsive Seizure Frequency
During Open Label Treatment over the first 24 months of the ongoing Open Label Extension (Study 1503)

- The decrease in patient number is primarily due to staggered entry into the study and not due to patient withdrawal.
- *p < 0.001 compared with no change (Wilcoxon signed rank test)
- Data disclosed as of April 19, 2019

Data originally presented at AAN 2020
Safety Highlights in Dravet Syndrome

Fintepla is only available through the Fintepla REMS Program

• Fintepla is available only through a restricted distribution program called the Fintepla REMS because of the risk of valvular heart disease and pulmonary arterial hypertension.

Boxed Warning: Valvular Heart Disease and Pulmonary Arterial Hypertension

• There is an association between serotonergic drugs with 5-HT2B receptor agonist activity, including fenfluramine (the active ingredient in Fintepla), and valvular heart disease and pulmonary arterial hypertension.

• Echocardiogram (ECHO) assessments are required before, during, and after treatment with Fintepla.

Contraindications

• Fintepla is contraindicated in patients with hypersensitivity to fenfluramine or any of the excipients in Fintepla and with concomitant use of, or with 14 days of, the administration of monoamine oxidase inhibitors because of an increased risk of serotonin syndrome.

No patient has developed valvular heart disease or pulmonary arterial hypertension in controlled or uncontrolled studies of up to 3 years in duration.

*Please see full Prescribing Information for additional Important Safety Information and a complete list of the adverse events reported to date
Fintepla Commercialization

HCP, caregiver awareness; integrated support from enrollment through continued use

**Clinical Education**
- Clinician and caregiver education about unmet medical needs and Fintepla® clinical data

**Zogenix Central Hub**
- Integrated support programs and specialty pharmacy distribution

**Long-Term Support**
- Dedicated case management to ensure proper monitoring and education

**U.S. & Europe:** Zogenix in-house commercial teams

**Japan:** Commercial partner Nippon Shinyaku Co. Ltd.
Fintepla U.S. Launch in Dravet Syndrome

HCP Segments

- Community HCPs: Treating ~10% Patients
- Secondary Epilepsy Centers: Treating ~10% Patients
- Top Epilepsy Centers: Treating ~80% Patients
- Clinical Trial and EAP Sites

Multi-Pronged Approach under COVID-19

- Reprint Carriers
- Live KAM details
- Websites
- Dosing & Titration Guidelines
- Journal Ads

Virtual and Digital Plan
- Virtual Call
- On Demand Resources
- Emails
- Speaker Program
- Social Media
- Digital Display
Zogenix Central

- HCP and Patient – Therapy Decision
- HCP and Patient Enrollment in REMS and Z Central
- Welcome Call – Case Manager
- Reimbursement Assistance
- Therapy Education by Case Manager & Pharmacist
- Specialty Pharmacy Ships Drug to Patient
- Z Central follow up with Patient for Refills
Study 3: Global Phase 3 Trial Including Japan

Primary Endpoint and Secondary Endpoints Met, Consistent with Prior Phase 3 Trials of ZX008 in Dravet Syndrome Patients

Fintepla was generally well-tolerated in this study, with adverse events consistent with those observed in Study 1 and Study 2 and with the known safety profile of fenfluramine.

Study 3 was a multi-national, randomized, double-blind, placebo-controlled, Phase 3 study enrolling 143 children and young adults with Dravet syndrome, whose seizures were not adequately controlled by existing anti-epileptic drugs planned to serve as pivotal study for a planned new drug application (J-NDA) in Japan. Top-line data announced September 10, 2020.
Lennox-Gastaut Syndrome

Rare, highly refractory, and notoriously difficult to treat

- Childhood onset, usually between 2-7 years of age
- Varied disease etiology
- Significant intellectual, behavioral, and motor disabilities
- Higher risk of status epilepticus and sudden death
- Most patients' seizures remain uncontrolled on current antiepileptic treatment regimens
- Accounts for ~1-4 % of all cases of childhood epilepsy¹
- 60,000-100,000 living with LGS in the U.S. and Europe combined²

Patients require constant care, creating high physical, emotional, and financial burden for caregivers and the entire family

¹ NORD website https://rarediseases.org/rare-diseases/lennox-gastaut-syndrome/ Accessed May 20, 2019
² Zogenix company estimates
LGS Phase 3 Study 1601 Primary Efficacy Endpoint

Primary endpoint met demonstrating Fintepla, at a dose of 0.7 mg/kg/day, was superior to placebo as adjunctive therapy in the treatment of LGS based on median percent change in monthly frequency of seizures that result in drops ($p=0.0012$).

Key secondary endpoint met: number of subjects with ≥50% reduction in drop seizures: Fintepla 0.7 mg/kg/day group compared to placebo.

Median Percent Reduction in Monthly Drop Seizures
(2 Week Titration + 12 Week Maintenance Period)

- **0.7 mg/kg/day**: 26.5% ($p=0.0012$)
- **0.2 mg/kg/day**: 13.2% ($p=0.0915$)
- **Placebo**: 7.8%

~3.4x greater reduction vs placebo

p-values are treatment compared with placebo.
Study 1601 Safety Results

- Generally well-tolerated with adverse events consistent with known safety profile of fenfluramine
- No cases of valvular heart disease or pulmonary arterial hypertension
- Six patients in the 0.7/mg/kg/d group had an AE that led to a discontinuation compared to four patients in the 0.2/mg/kg/d group and one patient in the placebo group
- One death during the trial caused by SUDEP in a patient in the 0.7/mg/kg/d/group assessed to be unrelated to study drug

<table>
<thead>
<tr>
<th></th>
<th>Placebo (n=87)</th>
<th>0.2 mg/kg/d (n=89)</th>
<th>0.7 mg/kg/d (n=87)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Subjects With at Least One Treatment Emergent Adverse Event (AE)</td>
<td>79.3%</td>
<td>76.4%</td>
<td>89.7%</td>
</tr>
<tr>
<td>Number of Subjects With at Least One Serious Treatment Emergent Adverse Event (SAE)</td>
<td>4.6%</td>
<td>4.5%</td>
<td>11.5%</td>
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<tr>
<td>Most Common Adverse Events (&gt;10%)</td>
<td>Placebo (n=87)</td>
<td>0.2 mg/kg/d (n=89)</td>
<td>0.7 mg/kg/d (n=87)</td>
</tr>
<tr>
<td>Decreased Appetite</td>
<td>13 (14.9%)</td>
<td>18 (20.2%)</td>
<td>32 (36.8%)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>4 (4.6%)</td>
<td>10 (11.2%)</td>
<td>11 (12.6%)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>5 (5.7%)</td>
<td>12 (13.5%)</td>
<td>9 (10.3%)</td>
</tr>
<tr>
<td>Fatigue</td>
<td>11 (12.6%)</td>
<td>8 (9.0%)</td>
<td>16 (18.4%)</td>
</tr>
<tr>
<td>Pyrexia</td>
<td>11 (12.6%)</td>
<td>11 (12.4%)</td>
<td>9 (10.3%)</td>
</tr>
<tr>
<td>Seizure</td>
<td>5 (5.7%)</td>
<td>11 (12.4%)</td>
<td>5 (5.7%)</td>
</tr>
<tr>
<td>Somnolence</td>
<td>10 (11.5%)</td>
<td>10 (11.2%)</td>
<td>15 (17.2%)</td>
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</table>
Fintepla Intellectual Property Position

Patents and regulatory exclusivity provides proprietary protection up to 2033 and beyond

Global Commercialization Rights
- Exclusive worldwide license
- Full data and patent rights in rare epileptic encephalopathies

Orphan Drug Exclusivity

<table>
<thead>
<tr>
<th>Region</th>
<th>Orphan Drug Exclusivity</th>
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<tbody>
<tr>
<td>U.S.</td>
<td>Dravet: 7.5-year Orphan &amp; Pediatric Drug exclusivity</td>
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<td>LGS: 7-year Orphan Drug exclusivity</td>
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<tr>
<td>Europe</td>
<td>Dravet: 12-year Orphan &amp; Pediatric Drug exclusivity</td>
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<td></td>
<td>LGS: 10-year Orphan Drug exclusivity</td>
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<tr>
<td>Japan</td>
<td>Dravet &amp; LGS: Seeking Orphan Drug exclusivity</td>
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</table>

Strong Patent Portfolio
- U.S. & International patents with expiries 2033-2039
- 8 Orange Book listed patents in the U.S.
- Coverage for Dravet syndrome and other rare epileptic encephalopathies
- Coverage for proprietary methods of manufacture (API), drug product formulation (pending), and Safety/REMs
MT-1621 | TK2 Deficiency
MT-1621: Thymidine kinase 2 (TK2) deficiency

Rare, genetic, progressive and often fatal mitochondrial DNA depletion syndrome

- Autosomal recessive mutations in TK2 gene
- First patient described in 2001
- Clinically characterized by severe muscle weakness, profoundly impairing movement, breathing, eating/nutrition and other normal functions
- Untreated patients show progressive decline which is often fatal. No spontaneous recovery has been reported
- Age of onset is prognostic for outcome; younger patients have high mortality
- Currently no approved therapies; treatment limited to supportive care

MT-1621: dC/dT Substrate Enhancement Therapy

An investigational deoxynucleoside combination therapy for TK2 deficiency

- TK2 phosphorylates deoxycytidine (dC) and deoxythymidine (dT), fundamental building blocks in production of mitochondrial DNA (mtDNA)
- MT-1621 uses alternative enzyme phosphorylation pathways and residual TK2 activity to potentially restore mitochondrial function
- Deoxynucleoside combination therapy improved nucleotide balance, increased mtDNA copy number, improved cell function, and prolonged life in preclinical models of TK2d
- Given orally, dissolved in solution
MT-1621: RETRO study preliminary data

Statistically significant survival in treated vs. untreated (natural history) groups

Survival Analysis Curves Modeled from Treated and Modified Untreated Groups

Survival Analysis comparing treated patients in RETRO study with untreated patients (TK2d Natural History data set) showed statistically significant effect of pyrimidine nucleosides on survival (p<0.004)

Functional Outcomes

- MT-1621 treated patients demonstrated improvements in functional domains (motor, respiratory, feeding)
- A number of MT-1621 treated patients re-acquired previously lost motor milestones (e.g., ambulation, discontinuing respiratory support)

Safety

- MT-1621 was generally well tolerated
- Most common treatment-related adverse event: mild to moderate diarrhea (63%)
- Four patients experienced Serious Adverse Events related to study drug
- Two patients discontinued due to increased liver enzymes
Key Financial Data

- $390M Cash and Marketable Securities as of June 30, 2020
- No debt as of June 30, 2020
- 1H2020 Operating Expenses of $120.7mm
  - R&D of $75.0mm\(^{(1)}\)
  - SG&A of $45.7mm
- 55.4mm basic shares outstanding as of June 30, 2020\(^{(2)}\)

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\(^{(1)}\) Includes acquired in-process R&D of $3mm and change in fair value of contingent consideration of $4.3mm.

\(^{(2)}\) Does not include 5.1mm options outstanding at a weighted average exercise price of $29.83 and 0.4mm restricted stock units as of June 30, 2020.
### Completed and Anticipated Future Milestones

<table>
<thead>
<tr>
<th></th>
<th>Dravet syndrome</th>
<th>Lennox-Gastaut syndrome</th>
<th>Rare, Treatment-Resistant Epilepsies</th>
<th>TK2 Deficiency</th>
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<tbody>
<tr>
<td><strong>Q1 2020</strong></td>
<td></td>
<td>Positive top-line results in Global Phase 3 trial ✓</td>
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<td><strong>Q2 2020</strong></td>
<td>FDA Approval, June 25, 2020 ✓</td>
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<td>FDA Type B and EMA meetings confirming paths for NDA and MAA submission ✓</td>
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<td><strong>Q3 2020</strong></td>
<td>Commercial launch in U.S. ✓</td>
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<td><strong>Q4 2020</strong></td>
<td>EMA CHMP Opinion</td>
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<tr>
<td><strong>Q1 2021</strong></td>
<td>Potential commercial launch in Germany</td>
<td>Ph2 Basket Study first patient screened</td>
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<tr>
<td><strong>Q2 2021</strong></td>
<td>sNDA submission in U.S.</td>
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<tr>
<td><strong>Q3 2021</strong></td>
<td>J-NDA submission in Japan</td>
<td>Study 1601 Japan top-line results</td>
<td>102 Study data compiled for NDA submission</td>
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</table>
Together we can bring hope and support to patients and families impacted by rare diseases.