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### Leader in Solving Unmet Medical Needs in the Area of Inner Ear Disorders

Sensorion is leading the way in the field of hearing loss with the most diversified pipeline to **Restore**, **Treat** and **Prevent**.

<table>
<thead>
<tr>
<th>Unique R&amp;D platform</th>
<th>Strong management and governance</th>
<th>Large market with different unmet medical needs</th>
<th>Disciplined financial processes</th>
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<tbody>
<tr>
<td><strong>Skills and know-how within Inner Ear Diseases:</strong></td>
<td><strong>Experienced management team with biotech and pharma industry backgrounds</strong></td>
<td><strong>Global untapped market evaluated at $10bn+</strong></td>
<td><strong>Selective capital allocation between programs</strong></td>
</tr>
<tr>
<td>• Pathophysiology</td>
<td>• Solid R&amp;D team with experience in inner ear and CNS</td>
<td>• 48m US patients suffering from hearing loss</td>
<td>• €9.7m non dilutive financing awarded in 2019 to finance the OTOF gene therapy program up to first patient</td>
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<tr>
<td>• Etiology</td>
<td>• Strong industry advisors such as John Furey – ex Spark Tx COO</td>
<td>• Hearing Care market CAGR estimated at c.4-5%</td>
<td>• €5.6m non dilutive financing (PSPC) awarded in Dec 2019 to support SENS-401 Phase 2 in SSNHL with involvement of the French Army</td>
</tr>
<tr>
<td>• Biomarkers</td>
<td>• Biotech specialists investors such as Invus and Sofinnova</td>
<td></td>
<td>• Cash runway until mid-Q1 2021*</td>
</tr>
</tbody>
</table>

*to be extended based on Dec 2019 PSPC award – update to be provided based on final contractualization with Bpifrance (French public investment bank)

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The Inner Ear is One of the Most Delicate Organ in the Human Body

KEY FACTS

- Inner ear disorders affect around 360m patients worldwide
- Every human is born with a definite number of sensory hair cells
  - 3,500 Inner Hair Cells
  - 15,000 Outer Hair Cells
- Hair cells do not regenerate

Source: OMS report 2015
Addressing Major Causes of Hearing Loss through our Restore, Treat and Prevent strategy

The most frequent causes of hearing loss

**TREAT**
Through our SENS-401 Program in Phase II on SSNHL

- Exposure to loud noise
- Natural ageing
- Heredity
- Head injury
- Ototoxic medications
- Illness

**PREVENT**
Through our SENS-401 programs targeting Cisplatin and Aminoglycoside induced ototoxicity

**RESTORE**
Through our gene therapy programs in collaboration with Institut Pasteur

Source: Mayo Clinic
### Building an Attractive Pipeline in the Hearing Therapeutics Space

<table>
<thead>
<tr>
<th>PRODUCT</th>
<th>IN-VITRO POC</th>
<th>IN-VIVO POC</th>
<th>PRECLINICAL</th>
<th>PHASE 1</th>
<th>PHASE 2 CLINICAL POC</th>
<th>PHASE 3</th>
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<tr>
<td><strong>TREAT</strong></td>
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<td>SENS - 401</td>
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<td>Sudden Sensorineural Hearing Loss</td>
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<td><strong>PREVENT</strong></td>
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<td>SENS - 401</td>
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<td>Cisplatin Induced Ototoxicity</td>
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<td>Hearing preservation after cochlear implantation</td>
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<td>SENS - 401</td>
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<td>Aminoglycoside Induced Ototox.</td>
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<tr>
<td><strong>RESTORE</strong></td>
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<td>USHER - GT</td>
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<td>Usher Syndrome</td>
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<td>OTOF - GT</td>
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<td>Otoferlin deficiency</td>
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</tbody>
</table>
**SENS-401 Mechanism of Action Creates the Opportunity to Target Multiple Indications with One Compound**

**Disrupted Ca\(^{2+}\) Homeostasis**
- Excitotoxicity
- Neuro Inflammation

**Calcineurin Activation**
- NFAT translocation: oxidative stress
- Actin (cytoskeleton) depolymerization
- BAD translocation/mPTP opening/cytochrome C + AIF release/Caspase 9/3 activation

**Structural degeneration, swelling and synaptic uncoupling & Apoptosis (cell death)**

- 5HT3R antagonist

**INSULT**

**SENS-401**
Partnering with Top Experts of the Inner Ear Space

**Institut Pasteur**
- Institut Pasteur is an internationally renowned center for biomedical research
- Partnership framework agreement with Sensorion on Gene Therapy programs targeting hearing loss
- First two programs are on Otoferlin & Usher Type 1
- Rights of first negotiation for the research pipeline
- World-class scientific team led by Pr. Christine Petit, who received the Kavli prize in 2018

**Cochlear**
- Cochlear is the global leader in implantable hearing solutions
- In exchange of equity investment Cochlear received a right of first negotiation for a global license to use SENS-401 in patients with certain implantable devices
- Ongoing Preclinical study on SENS-401 in combination with cochlear implants

**Necker**
- Necker Hospital has one of the biggest pediatric ENT unit in Europe
- Together with Necker Hospital, Institut Pasteur and Fondation pour l’Audition, we were awarded a RHU prize (Hospital research university) with a €9.7m grant to develop a gene therapy program addressing Otoferlin deficiency

**French Armed Forces Biomedical Research Institute (IRBA)**
- Conducts research for prevention and care improvement in military personnel
- Consortium including IRBA and Sensorion was awarded a €10.8m non dilutive financing. Sensorion will receive €56m to further develop SENS-401 in SSNHL
- Volunteer military personnel exposed to impulse noise will be recruited in the ongoing SENS-401 Phase 2 study
Sudden Sensorineural Hearing Loss is an Otological Emergency

**What is SSNHL?**

The sudden onset of a significant hearing loss due to dysfunction of the cells of the cochlea and central auditory structures

Hearing loss develops over less than 72 hrs, hearing sensitivity is reduced by at least 1,000 fold in the affected ear(s)

**Incidence**

Between 27 to 35 per 100,000 people (218,000 patients in 2017 in G7 countries)\(^1\). >70% cases are idiopathic, known causes include noise/head trauma, ischemia and infection

\(^1\) Company estimates based on publicly available data (in the US, Japan, Germany, France, the UK, Italy and Spain)

**Unmet medical needs**

More than 50% suffer from permanent, disabling hearing loss, mostly those with initial severe to profound hearing loss

Complications significantly impact patients’ quality of life due to:
- Difficulties in communicating, social isolation, cognitive decline
- Accompanying tinnitus
SENS-401 Developed to Treat Sudden Sensorineural Hearing Loss

First-in-class treatment

- First-in-class oral 5HT3 receptor antagonist & calcineurin inhibitor
- The Mode of Action is well-defined and understood
- SENS-401 acts through reduction of cochlear cell death and neurodegeneration

SENS-401 market exclusivity

- Strong IP
  - 2 patent families filed
- Orphan Drug Designation from EMA
- Pediatric Investigation Plan approved in EU

SENS-401 demonstrated safety and PK in phase 1

- 36 healthy volunteers enrolled in a double-blind, randomized, multiple ascending dose design (7 days)
- No serious or significant adverse events reported, safety profile comparable to placebo
- Pharmacokinetics match effective systemic exposures in preclinical model

Phase 2 trial started in 2019

- Opening of centers is ongoing
- Primary endpoint: audiometry with a 10db improvement over the 3 most commonly affected frequencies vs baseline.
- Results aimed for Q2 2020
Daily Administration of SENS-401 Reduces Auditory Deficit

A daily oral administration of SENS-401 reduces auditory deficit, improves recovery and reduces hair cell loss

**MODEL**
- Randomized treatment post-noise induced trauma (2h exposure at 120 dB) in rats receiving either twice daily placebo or SENS-401 PO for 28 days

**BENEFIT**
- Regulatory threshold for efficacy (>10 dB improvement)
- Significant effects with treatment initiation delay up to 96 hrs

**Histology of hair cell layers**
- Significant hair cell loss
- Limited hair cell loss

**Cochleograms**
- Placebo
- SENS-401

**ABR threshold shift from baseline to day 32/33 after acoustic trauma**

**ABR threshold recovery from 24h to day 32/33 after acoustic trauma**

Petremann et al. 2018
401-SSNHL Phase 2: a Multicentered, Randomized, Double-blind vs Placebo Study

**Screening**
- 50 Clinical sites
  - Global

**Primary Endpoint**
- 1 Primary Endpoint
  - Audiometry

**10 dB Improvement**
- 3 most frequencies
  - Vs baseline 260 patients

**Planning**
- Q1 2019
  - Center openings
- Q2 2020
  - Final results
- Q4 2019
  - Interim. safety readout

**Dose 1**: 43.5mg BID
**Dose 2**: 29mg BID
**Placebo**

**Randomization**

**Visit 1**: Screening
**Visit 2**: Inclusion
**Visit 3**: Visit
**Visit 4**: Visit
**Visit 5**: End of Treatment
**Visit 6**: End of Study
Cisplatin Induced Ototoxicity (CIO) Leads to Permanent Hearing Loss

What is CIO?

Cisplatin administration for chemotherapeutic treatment of cancer damages the inner-ear and leads to hearing loss, tinnitus and dizziness.

Risks factors for CIO include young age as well as individual and cumulative cisplatin doses during chemotherapy.

Incidence

Between 350 to 450 per 100,000 people (~500,000 patients in 2017 in G7 countries)\(^1\)

Unmet medical needs

CIO leads to permanent inner ear problems in 50-60% of cases. These complications significantly impact patients’ quality of life due to:

- Hearing loss, tinnitus and dizziness impacting daily life activities
- Problems in language acquisition and learning for pediatric patients
- Difficulties in communicating, social isolation, cognitive decline

Potential treatments must not interfere with cisplatin efficacy.

\(^1\) Company estimates based on publicly available data (in the US, Japan, Germany, France, the UK, Italy and Spain)
**SENS-401 Developed to Prevent Cisplatin Induced Ototoxicity**

**First-in-class treatment**
- First-in-class oral 5HT3 receptor antagonist & calcineurin inhibitor
- The Mode of Action is well-defined and understood
- SENS-401 acts through reduction of cochlear cell death and neurodegeneration

**SENS-401 demonstrated safety and PK in phase 1**
- 36 healthy volunteers enrolled in a double-blind, randomized, multiple ascending dose design (7 days)
- No serious or significant adverse events reported, safety profile comparable to placebo
- Pharmacokinetics match effective systemic exposures in preclinical model

**SENS-401 market exclusivity**
- Strong IP
  - 2 patent families filed
- Orphan Drug Designation for pediatric patients from US FDA
- Pediatric Investigation Plan approved in EU

**Phase 2 ready in 2020**
- Trial to be conducted in the US and Europe in pediatric population
- Assessing the opportunity to lead a clinical trial in the adults population
SENS-401 Significantly Reduces Cisplatin-Induced Hearing Loss and Outer Hair Cell Death in Preclinical Models

Treatment Protocol
Placebo and SENS-401 at 6.6 mg/kg, 13.2 mg/kg or placebo once daily before and for 13 consecutive days after cisplatin infusion

Significant improvement versus placebo
• 23-29 dB, up to 65% reduction with 6.6 mg/kg
• 22-29 dB, up to 73% reduction with 13.2 mg/kg

Significant enhancement of OHC survival 22-264% for both doses

Petrenom et al. 2017
**SENS-401 Investigated to Prevent Cell Death Post Cochlear Implants Procedure**

**Healthy ageing**

Growing understanding of the link between healthy hearing and healthy ageing

- Cognitive decline
- Depression
- Falls
- Isolation
- Ability to work
- Loss of independence

**Cochlear collaboration**

- Preclinical studies designed to preserve hearing before and after cochlear implants surgery
- Several animal models were developed

**Next steps**

- Cochlear has a right of first negotiation to license SENS-401 to be used in combination with their implants
- Interim cat data results are expected in H1 2020
- Full data package in H2 2020

Source: COCHLEAR market day presentation
Sensorion Leading the Way with New Gene Therapy Options to Restore Auditory Functions

2 programs initiated with Pasteur within our Restore segment

**Usher Syndrome Type 1 (USH1)**
Patients with USH1 are born with severe to profound congenital bilateral sensorineural hearing loss and congenital vestibular dysfunction

Retinitis pigmentosa develops during childhood

**Otoferlin Deficiency (OTOF)**
Patients with mutations in OTOF suffer from profound sensorineural prelingual non-syndromic hearing loss

Sources: Morton 2006, Koshlaret al., 2007; Shearer et al. 2017

We believe developing gene therapy for those diseases has a life-changing potential for patients

Right of first refusal on all GT programs from Institut Pasteur

- World-class scientific minds having significantly contributed to gene and protein discoveries in the hearing field
- Team managed by Pr Petit who will lead the future French Hearing Institute
- Vectors bank
Usher Syndrome Type 1: Most Severe Expression of the Disease

**Usher type 1 – a rare genetic disorder**
- **Biological description**
  - Mutation on USH1C, MYO7A, CDH23, PCDH15, USH1G, CIB2 gene, autosomal recessive
  - Inner hair cell lacking USH1G/Sans protein cannot develop, maintain and have a functional hair bundle
- **We are addressing the USH1G mutation**

**Gap in care and burden of care**
- **Delayed diagnosis** – not suspected at first sight
- **No standard of care**, cochlear implant in less than 10% of the patients

**Affected population**
- **Average Prevalence of USH1 1:10 000**
- Represent 4-6% of people with hereditary deafness
- **EU = 25 700 (1/29 000) and US = 15 000 (1/23 000)**

---

Sources: Papsin et al. 2007; Sahin et al. 2017

Sources: Morton 2007; Shearer et al. 2017; Scheffield et al. 2018
Institut Pasteur Restored Hearing & Vestibular Functions in Mouse Model of Usher Type 1G

Cochlear stereocilia

Normal mice

Usgh1-/mice

Treated Usgh1-/mice

Source: Emptoz et al. 2017

Vestibular stereocilia

Histological improvements were electrophysiologically validated

Restoration of stereocilia physiology using AAV8-SANS allows an increase in electric function

Source: Emptoz et al. 2017

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Otoferlin Deficiency Responsible for up to 8% of All Prelingual Non-Syndromic Deafness

**Otoferlin deficiency - a rare genetic disorder**
- **Biological description**
  - Mutation of OTOF gene (locus DFNB9) with autosomal recessive transmission
  - Synaptopathy affecting ribbon on inner hair cells. Cochlear nerve is not affected

**Unmet medical need and burden of care**
- **Delayed diagnosis** – not suspected at first sight
- **No standard of care**, cochlear implant in less than 10% of the patients

**Affected population**
- **Estimated prevalence in 0-4 year population suffering from non-syndromic hereditary hearing loss**: 4-8%
- 15,000 – 20,000 new cases of non-syndromic hereditary hearing loss per year (US + EU)

Sources: Shearer et al. 2017; Scheffield et al. 2018

Sources: Papsin et al. 2007; Cosetti et al. 2010; Sahin et al. 2017

Sources: Choi et al; 2009; Iwasa et al. 2013; Shearer et al., 2015
Local Gene Therapy Durably Restores Cochlear Receptors Function in a Mouse Model of Otoferlin Deficiency

Cochlear stereocilia in wild type, Otof -/- and Otof -/- injected with AAV2-OTOF vector

Expression of Otof protein in cochlear receptors

Normal mice

Otof-/mice

Treated Otof -/-mice

Restoration of auditory function in Otof -/- using Dual AAV2-OTOF

Akil et al. 2019
Sensorion is Well-Positioned with its **Restore, Treat, Prevent** franchise

---

**Strong foundations...**

**R&D platform**
- Small molecules & Gene therapy
- Preclinical & clinical capabilities
- Biomarkers identification

**Ecosystem**
- Key players from the field (Pasteur, Cochlear, Necker, IRBA)

**Finance**
- Supportive experienced biotech investors

---

**... to lead the field**

**Treat**
- SENS-401 Phase 2 trial in SSNHL

**Prevent**
- SENS-401 Phase 2 ready in CIo
- SENS-401 investigated in combination with cochlear implant

**Restore**
- Usher Type I gene therapy
- Otoferlin deficiency
Financial profile and Shareholder Overview

Cash position
- At June 30th, the cash position was €22.3m
- In September 2019, completion of a capital raise of €18.1m in equity

Key renowned investors joined Sensorion in 2019
- Invus and Sofinnova Partners
- Wuxi AppTec and 3SBio

Non dilutive funding
- €9.7m grant awarded in June 2019 to finance the development of the OTOF gene therapy program up to first patient in (“AUDINNOVE” consortium)
- €5.6m non dilutive financing (PSPC) awarded in Dec 2019 to support SENS-401 Phase 2 in SSNHL with involvement of the French Army

Shareholder base before convertible bonds conversion
Total shares October 17th, 2019 : 32,247,263
## 2020 Key Goals

<table>
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<tr>
<th>Goals</th>
<th>Expected timing</th>
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<td>Intermediate preclinical data read out with Cochlear</td>
<td>H1 2020</td>
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<tr>
<td>Preclinical milestones of the two gene therapy programs</td>
<td>H1 2020</td>
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<tr>
<td>SENS-401 SSNHL phase 2 data readout</td>
<td>H1 2020</td>
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</table>
Thank you
Shareholder base post-conversion

Fully diluted, based on an OC conversion price of €1.3662

- Invus Public Equities: 26%
- Sofinnova Partners: 15%
- WuXi AppTec: 8%
- 3SBio: 8%
- BPIFrance Investissements (Innobio): 7%
- Novalis: 3%
- ITI: 2%
- Cochlear: 1%
- Free Float: 30%