Press release

Sensorion reports 2021 first half results

- ***SENS-401 trial protocol for SSNHL successfully amended, clinical trial application filing in CIO in adults confirmed for H2 2021***
- ***Plans to file Clinical Trial Application (CTA) in H1 2023 for OTOF-GT***
- ***Launch of a third GT program targeting GJB2-GT for adult and pediatric segments***
- ***Cash position of c.€55m at June 30, 2021 provides unchanged runway until the end of H2 2022***

Montpellier, September 27, 2020 – Sensorion (FR0012596468 – ALSEN), a pioneering clinical-stage and gene therapy biotech company specializing in the development of novel therapies to restore, treat, and prevent within the field of hearing disorders, today announces its interim annual results as of June 30, 2021 alongside its outlook for the remainder of 2021.

"Many elements of Sensorion's strategic planning came together in a successful first half of 2021. Our otoprotectant small molecule SENS-401 continued to make clinical and regulatory progress. Regulators accepted our proposal to reduce the sample size of our study in Sudden Sensorineural Hearing Loss; we will open a new combined study on SENS-401 in cisplatin-induced ototoxicity in H2 2021; and our partnership with Cochlear Limited will take SENS-401 into the clinic in combination with cochlear implantation. Sensorion's gene therapy programs, made progress too. Recently we received feedback from regulatory advisors of the EMA that was aligned with the assumptions of our otoferlin gene therapy development program. At the start of 2021, we had announced our third and potentially largest gene therapy collaboration with our long-term partners at the Institut Pasteur, on GJB2. This program creates significant opportunities in both adult and pediatric hearing loss. In addition, part of the deal we announced with Sonova will involve genotyping to survey gene variants associated with progressive hearing loss in adults, including GJB2. This might represent a first step in bringing together routine diagnosis of hearing loss with advanced treatments such as gene therapy" said **Nawal Ouzren, CEO of Sensorion.**

Key developments in H1 2021: science and operational

Gene Therapy programs

- **Third collaboration with Institut Pasteur: GJB2 gene therapy targeting adult and pediatric segments**

On February 15, 2021, Sensorion announced its largest gene therapy opportunity to date, a collaboration with Institut Pasteur, targeting the GJB2 gene in pediatric and adult deafness. Research by the Institut Pasteur demonstrated anomalies in GJB2 are both the most common cause of congenital deafness as well as a wide contributor in adult cases of severe age-related hearing loss. Although the types of GJB2 mutation in children and adults may differ, gene therapy could potentially provide solutions in both spheres.

Sensorion's GJB2 gene therapy programs have the potential to address three pathologies related to GJB2 mutations: age-related hearing loss in adults, progressive forms of hearing loss in children, and pediatric congenital deafness. Initially, the focus will be on the first two populations and in particular the

adult population with GJB2-associated hearing loss where the unmet medical need is well-defined, becoming the first company to address this important and potentially large market segment. Sensorion will later target the pediatric need related to GJB2.

- **OTOF-GT gene therapy program: clear advice from the European Medicines Agency to progress the program**

Sensorion sought scientific advice from EMA on the preclinical and clinical development plans for OTOF-GT, the Company's dual vector AAV gene therapy program for the treatment of children born with hearing loss caused by otoferlin deficiency.

In Q3 2021, Sensorion received clear feedback from the EMA's advisors on the overall development plan for OTOF-GT, which was consistent with the Company's preclinical and clinical development plans. The agency's advisors also welcomed the ongoing Natural History Study, Audioferline (NCT04202185), a component of the AUDINNOVE project coordinated by researchers at Hôpital Necker-Enfants malades (Necker Hospital) in partnership with Sensorion. Sensorion's aim is to document the natural course of disease progression in otoferlin deficiency patients, define clinically meaningful endpoints suitable for market approval, and identify the patient populations that would benefit the most from Sensorion's OTOF-GT treatment.

The Natural History Study will enable Sensorion to select the most relevant and clinically meaningful endpoints and clinical trial design as OTOF-GT progresses into the clinic.

Sensorion expects to file a Clinical Trial Application (CTA) for its OTOF-GT program in H1 2023.

- **Usher-T1-GT project**

We have completed the preclinical proof-of-concept study for our gene therapy approach in Usher's syndrome (USHER-GT). The study was designed to test whether a gene therapy approach would be effective in older mice, thereby opening the possibility of extended treatment window for clinical studies.

The results of the completed study require further analysis with some indications of hearing restoration in the older animals and complete restoration of the vestibular function. The company will continue to review the results before providing a more detailed update on the implications of the study in the coming months.

Drug candidate SENS-401

Our clinical program with otoprotective small molecule SENS-401 continued to progress in 2021 in different indications: Sudden Sensorineural Hearing Loss (SSNHL), Cisplatin-Induced Ototoxicity (CIO) and cochlear implantation.

- **SENS-401 in SSNHL: AUDIBLE-S Phase 2 protocol amendment approved**

In March 2021, Sensorion announced plans to approach regulators to reduce the study sample size following a review of the trial design and statistical analysis plan for the AUDIBLE-S study in patients with Sudden Sensorineural Hearing Loss (SSNHL).

In September 2021, Sensorion announced that these amendments to AUDIBLE-S were approved by 9 out of 10 regulatory authorities of all participating countries, the last one still being under evaluation. The recruitment target had been significantly reduced without compromising the quality or potential outcome

of the trial. The study has now a recruitment target of 111 patients and has enrolled 112 patients. Since not all of the patients have yet completed their 28-day end of treatment visit, Sensorion will continue the recruitment until the end of October 2021. Top line data continues to be expected around year end (see post-closing events).

- **SENS-401 in CIO: clinical trial application filing confirmed in H2 2021**

Following SENS-401's demonstrated ability to significantly reduce hearing loss in a rat model of cisplatin-induced ototoxicity (CIO, Petremann et al., 2017), Sensorion will initiate a Phase 2 clinical study of SENS-401 in adults with CIO in H2 2021, as previously announced.

The trial will have three arms, including a control arm without SENS-401. The natural history study (NHS) of CIO the company had planned to begin at the end of H1 2021 is now included as the control arm. Sensorion's modified strategy condenses the clinical timelines of the CIO program, provides a more robust and relevant control arm for the study and makes SENS-401 available sooner to patients being treated with cisplatin therapies.

- **SENS-401 for hearing preservation in combination with cochlear implantation: to begin first clinical trial with Cochlear Limited**

On January 19, 2021, Sensorion released positive preclinical data demonstrating that the combination of its SENS-401 molecule and a cochlear implantation helped reduce loss of residual hearing at a frequency located beyond the electrode array. Preservation of 'natural' hearing is particularly important in speech recognition.

Following this initial success, Sensorion and Cochlear announced the initiation of a Phase 1 clinical trial of SENS-401 (Arazasetron) in patients scheduled for cochlear implantation on September 8, 2021. The study will be sponsored by Sensorion and the proposed design is expected to be submitted to regulatory authorities during H2 2021 (see post-closing events).

Pipeline summary

Treatment	Mechanism of Action	Indication	Status
GJB2-GT	Gene therapy	Progressive pediatric hearing loss	Candidate selection
GJB2-GT	Gene therapy	Pediatric congenital deafness	Candidate selection
GJB2-GT	Gene therapy	Adult age-related hearing loss	Candidate selection
OTOF-GT	Gene therapy	Otoferlin deficiency	CTA in H1 2023
USHER-GT	Gene therapy	Usher's syndrome Type 1	Preclinical PoC
SENS-401	Small molecule	Sudden Sensorineural Hearing Loss (SSNHL) - Otorestoration	Top line data release around end of 2021
SENS-401	Small molecule	CIO Cisplatin Induced Ototoxicity - Otoprotection Otorestoration	Clinical trial application in H2 21
SENS-401	Small molecule	Combined with cochlear implantation - Otoprotection	Clinical trial application in H2 21

Multi-year strategic collaboration with Sonova

On September 15, 2021, Sensorion announced the signing of an important multi-year collaboration with Sonova, a leading international player in the hearing solutions market. The collaboration aims to create new diagnostic and therapeutic solutions for hearing loss and it brings to fruition the exclusive negotiations commitment made between the two companies in December 2020, when Sonova acquired a 3.7% stake in Sensorion.

Part of the deal is a jointly-funded study of natural history in age-related progressive hearing loss (presbycusis) in adults. It will involve the collection of disease information and samples via selected Sonova Audiological Care stores. This will be followed by genotyping to better understand the prevalence of gene variants thought to be associated with progressive hearing loss in adults, including GJB2. The collaboration could lead to the introduction of genetic analysis to the routine diagnosis of progressive hearing loss in adults and subsequently open the way for improved care through a combination of advanced therapeutic interventions and traditional hearing solutions including hearing aids. Sonova and Sensorion will jointly fund the study with €7.0 million, split 70/30 between the two companies (see below post-closing events).

Strengthened scientific and medical leadership

As Sensorion expands and builds its product development capabilities, it has expanded its senior management team with the creation of two new senior executive positions.

In June, gene therapy and rare disease expert, Dr Nora Yang, was named as the company's new Chief Scientific Officer based in the USA. Dr Yang leads the preclinical organization and spearheads the development of collaborations between fundamental and clinical research, including working with Sensorion's partner, Institut Pasteur.

In July, Dr Otmane Boussif was appointed to the newly created position of Chief Technology Officer. Dr Boussif, formerly headed Gene Therapy CMC at Novartis, and oversees now all of Sensorion's technical operations and CMC functions.

Both positions will be critical in taking Sensorion's pipeline forward and securing regulatory approvals.

Scientific communications

Sensorion presented at various scientific congresses and hosted one Key Opinion Leader (KOL) event in H1 2021 including:

- Christine Le Bec, Head of CMC Gene Therapy, spoke in the "*Gene Therapy Analytics and Manufacturing*" session at the 4th Annual Bioprocessing Summit Europe, in March 2021. Dr Le Bec's talk on "*Quality Control for a Dual AAV Vector*" focused on product characterization and quality control aspects of Sensorion's gene therapy program in Otoferlin deficiency, where the size of the Otoferlin gene requires the use of a dual AAV vector system.

- Sensorion hosted a Key Opinion Leader (KOL) Webinar with Dr. Thomas Lenarz on the GJB2 Gene Related Hearing Loss on May 10, 2021. Dr Lenarz discussed the clinical aspects, current treatment landscape and unmet medical need in treating patients with pediatric onset of GJB2–related hearing loss as well as the role of the GJB2 gene.

- Christine le Bec co-chaired a scientific session on the *"Development of AAV Capsid Variants*" at the American Society of Gene & Cell Therapy (ASGCT) Annual meeting, that took place virtually on 11-14 May 2021.
  In addition, Sensorion's Research Team presented an overview of the company's Gene Therapy platform focused on hearing disorders at the ASGCT Annual Meeting, held in May 2021.

Others

During the first half of 2021, Sensorion Inc subsidiary was created in the United States.

H2 2021 announcements

Since the end of June 2021, the key business updates are as follows:

- The company incorporated a wholly owned subsidiary in Australia, Sensorion Australia, in July 2021.

- On September 8, 2021, Sensorion and Cochlear reported the initiation of the first clinical trial of SENS-401 for hearing preservation in combination with cochlear implantation. The study aims to measure the perilymph PK of SENS-401 and to explore the therapeutic effect of SENS-401 on early hearing outcomes upon cochlear implantation. The trial will be fully funded under the existing agreement between Cochlear and Sensorion. The study will be sponsored by Sensorion and the proposed design is expected to be submitted to the regulatory authorities in H2 2021.

- On September 15, 2021, Sensorion and hearing care leader Sonova signed a strategic research collaboration which covers a jointly funded Natural History Study in age-related hearing loss (presbycusis). It is intended that large-scale genetic screening and clinical follow-up will be performed in collaboration with Sonova Audiological Care stores. The key element of the collaboration between Sensorion and Sonova is a natural history study including genotyping thousands of patients suffering from early onset of severe presbycusis with the aim of confirming approximately a hundred patients for specific variants of gene mutations.

- On September 24, 2021, Sensorion announced that 9 out of 10 regulatory authorities of all participating countries have approved Sensorion's amendment to the Phase 2 AUDIBLE-S study protocol with SENS-401 in Sudden Sensorineural Hearing Loss (SSNHL). Sensorion had submitted to the regulatory authorities an amendment to the statistical analysis plan for the AUDIBLE-S study that significantly reduced the recruitment target to 111 patients without compromising the quality or potential outcome of the trial. The study has now enrolled 112 patients. Since not all patients have yet completed their 28-day end of treatment visit, Sensorion will continue the recruitment until the end of October 2021. Top line data continue to be expected around year end.

H2 2021 strategy and prospects

As of June 30, 2021, the Company had c. €55 million in cash. Sensorion intends to use the new funds to develop its current gene therapy programs, support its pharmacology and clinical studies of SENS-401 and for general corporate purposes.

Expected future milestones and estimated timelines:

- H2 2021 – Submission of the clinical trial application for the SENS-401 CIO study in adults
- H2 2021 – Submission of the clinical trial application for the pilot study SENS-401 with cochlear implant
- End of 2021 – Top line data readout for the SENS-401 Phase 2 clinical study in SSNHL
- H1 2022 – Start of the pilot study SENS-401 in combination with cochlear implants
- H1 2022 – Sensorion and Institut Pasteur disclosing next steps for the USHER-GT program
- H1 2022 – GJB2-GT Candidate selection
- H1 2023 - Submission of the clinical trial application for the OTOF-GT program (CTA/IND)

First-half 2021 financial results

The half-year accounts as of June 30, 2021, drawn up according to IFRS standards and approved by the Board of Directors on September 24, 2021, have been duly reviewed by statutory auditors.

The simplified income statement as of June 30, 2021 is as follows:

In Euros – IFRS Standards	**30.06.2021**	**30.06.2020**
Operating income	**1,603,749**	**902,203**
Research and Development expenses	6,003,596	3,661,766
General and Administrative expenses	1,748,922	1,915,400
Total operating expenses	**7,752,518**	**5,577,166**
Operating profit/loss	**-6,148,769**	**-4,674,963**
Financial profit/loss	-94,152	-44,031
Net profit/loss	**-6,242,921**	**-4,718,994**

On June 30, 2021, Sensorion's **operating income** amounts to €1.6m including the research tax credit for €1.0m, and grants related to Audinnove (RHU) and Patriot (PSPC) programs for €0.6m. The € 0.7m increase in operating income is mainly due to grants for €0.5m, tax credit for €0.1m and refundable advance for €0.1m

Operating expenses increased by 39%, from €5.6m on June 30, 2020 to €7.8m on June 30, 2021.

The research and development expenses increased by 64%, from €3.7m on June 30, 2020 to €6.0m on June 30, 2021. The increase is mainly due to the development of preclinical and clinical studies and the increase of R&D headcounts.

G&A expenses are down 9%; from €1.9m on June 30, 2020 to €1.7m on June 30, 2021, mainly due to the decrease in consulting fees and travel costs.

Operating losses for the period ending June 30, 2021 thus amounted to €6.1m, compared with a loss of €4.7m in the prior year.

Net loss amounted to -€6.2m as of June 30, 2021, compared with -€4.7m for the period ended June 30, 2020.

As of June 30, 2021, the company employed 33 people.

Financial structure

The simplified balance sheet at June 30, 2021 is as follows:

In Euros – IFRS standards	**30.06.2021**	**31.12.2020**
Non-current Assets	**1,658,090**	**1,474,119**
Other Current Assets	5,282,431	4,254,909
Cash & cash equivalent	54,998,592	62,174,948
Total Assets	**61,939,113**	**67,903,976**
Equity	**52,261,833**	**58,379,653**
Non-current Liabilities	5,202,019	5,246,408
Current Liabilities	4,475,260	4,277,915
Total Liabilities	**61,939,113**	**67,903,976**

Total **Equity** amounted to €52.3m on June 30, 2021, compared with €58.4m on June 30, 2020, the decrease corresponds mainly to the loss of the period for €-6.2m.

As of June 30, 2021, cash and cash equivalent amounted to €54.9m, compared to €62.2m at December 31, 2020.

Based on its forecasted expenses, the cash position of €54.9m in June 30, 2021, the Company believes it will be able to fund its operations until the end of the second half of 2022.

First-Half 2021 certified accounts

The first-half 2021 detailed accounts can be found on Sensorion's website (https://www.sensorion.com/en/home/) in the investor section under financial information. The first-half year accounts as of June 30, 2021 have been duly reviewed by statutory auditors and the certification report has been issued.

Capital Breakdown

The company's capital breakdown as of June 30, 2021 is described in the table below:

	Non-diluted basis		**Fully diluted basis** [2]	
	Number of shares	**Equity holding**	**Number of shares**	**Equity holding**
Inserm Transfert Initiative	982,911	1.23%	982,911	1.19%
Innobio	3,499,874	4.39%	3,499,874	4.23%
Management, employees, directors [1]	160,000	0.20%	2,140,041	2.59%
Cochlear	533,755	0.67%	533,755	0.65%
Invus Public Equities LP	26,490,415	33.22%	26,490,415	32.02%
Sofinnova Partners	15,469,458	19.40%	15,469,458	18.70%
WuXi AppTec	5,249,608	6.58%	5,249,608	6.34%
SONOVA AG	2,941,176	3.69%	2,941,176	3.55%
3SBio	4,055,150	5.09%	4,055,150	4.90%
Floating (including former officers and directors)	20,357,881	25.53%	21,376,612	25.84%
Total	**79,740,228**	**100.00%**	**82,739,000**	**100.00%**

(1) Including 160,000 free shares allocated on May 29, 2018

(2) Including securities giving access to the capital and stock-options described below

Press release

About Sensorion

Sensorion is a pioneering clinical-stage biotech company, which specializes in the development of novel therapies to treat, prevent and restore within the field of hearing disorders. Its clinical-stage portfolio includes one Phase 2 product: SENS-401 (Arazasetron) for Sudden Sensorineural Hearing Loss (SSNHL). Sensorion has built a unique R&D technology platform to expand its understanding of the pathophysiology and etiology of inner ear related diseases enabling it to select the best targets and modalities for drug candidates. The Company is also working on the identification of biomarkers to improve diagnosis of these underserved illnesses. Sensorion has launched three gene therapy programs, currently at preclinical stage, aimed at correcting hereditary monogenic forms of deafness including deafness caused by a mutation of the gene encoding for Otoferlin, hearing loss related to gene target GJB2 as well as Usher Syndrome Type 1 to potentially address important hearing loss segments in adults and children. The Company is potentially uniquely placed, through its platforms and pipeline of potential therapeutics, to make a lasting positive impact on hundreds of thousands of people with inner ear related disorders, a significant global unmet medical need.

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