Press Release

Sensorion Reports Data in the Audiogene Phase 1/2 Gene Therapy Clinical Trial

- **SENS-501 gene therapy product and surgical procedure are well tolerated by the first two treated patients and encouraging behavioural changes are observed in both toddlers**
- **Sensorion plans on hosting a KOL event in early 2025 to further comment and discuss the first cohort safety data, qualitative and quantitative efficacy measurements, and next steps for Audiogene Phase 1/2 clinical trial, including planned interactions with the U.S. Food and Drug Administration.**

Montpellier, December 18, 2024, 7.30 am CET – Sensorion (FR0012596468 – ALSEN) a pioneering clinical-stage biotechnology company specializing in the development of novel therapies to restore, treat and prevent hearing loss disorders, announced today latest data updates confirming the safety of SENS-501 in the two first patients injected in the Company's Audiogene study. This is a Phase 1/2 clinical trial evaluating SENS-501 in patients aged 6 to 31 months and naïve of cochlear implant, who are suffering from a specific form of congenital deafness linked to mutations in the OTOF (otoferlin) gene.

The Audiogene clinical trial assesses, as primary endpoint in the first part of the dose escalation study, the safety of an intra-cochlear injection of SENS-501 in infants and toddlers aged 6 to 31 months at the time of gene therapy treatment. Sensorion's objective in targeting the first years of life, when brain plasticity is optimal, is to maximize the chances of these young children with pre-linguistic hearing loss to acquire normal speech and language, thus, potentially transforming these toddlers' lives. For the first two toddlers treated in the first cohort, SENS-501 gene therapy product and surgical procedure were well tolerated: the intra-cochlear administration of the gene therapy product was uneventful, and no serious adverse events were reported. In addition, encouraging behavioural improvements were observed in both toddlers. Sensorion plans on hosting a KOL event in early 2025 to further comment and discuss the first cohort safety data, qualitative and quantitative efficacy measurements, and next steps for Audiogene Phase 1/2 clinical trial, including planned interactions with the U.S. Food and Drug Administration. Further details regarding the event shall be disclosed in due course.

The Company continues to anticipate the completion of the first cohort enrollment by the end of the year and the recruitment of the second cohort by the end of H1 2025.

Géraldine Honnet, M.D., Chief Medical Officer of Sensorion, said: "I'm very pleased with the progress made in the Audiogene Phase 1/2 clinical trial evaluating SENS-501, a potentially game changing hearing loss therapeutic being developed in collaboration with the Institut Pasteur. Today's data results confirm an excellent safety profile for the first toddlers treated with SENS-501 and I am happy to report early signs of encouraging behavioural changes in both patients. We look forward to treating the third patient of the first cohort imminently, thus achieving an important development milestone for Sensorion. We will host a KOL call in early 2025 and I am looking forward to presenting further safety and efficacy data for patients included in our first cohort.
I am confident that Sensorion's differentiated clinical approach will set new standards in the field of gene therapy for otoferlin deficiency as the study has indeed been designed to assess whether SENS-501 can demonstrate not only hearing restoration in a very homogeneous population of infants and toddlers but also enable language acquisition and development, and the resulting significant improvements in quality of life. I would like to reiterate my gratitude to the parents of the treated patients for their trust, and to the participating investigators for their ongoing commitment to the Audiogene study and conviction in the potential of SENS-501 to create a new treatment paradigm for this debilitating form of deafness."

Professor Natalie Loundon, M.D., Director of the Center for Research in Pediatric Audiology, Pediatric Otolaryngologist and Head and Neck Surgeon, Necker Enfants Malades, AP-HP, in Paris, France, Coordinating Investigator of the Audiogene clinical study, commented: "The

preliminary results provide satisfactory data on the safety of SENS-501 for patients. Gene therapy represents real hope for a therapeutic treatment and improvement hearing, speech acquisition and quality of life for children born deaf due to DFNB9 mutations. I look forward to the next data update on Audiogene's first cohort, where patients will have received the lowest dose investigated for the restoration of hearing in DFNB9 patients and I'm excited to continue our work on the Audiogene clinical study."

About the Audiogene Trial

Audiogene aims to evaluate the safety, tolerability and efficacy of intra-cochlear injection of SENS-501 for the treatment of OTOF gene-mediated hearing loss in infants and toddlers aged 6 to 31 months at the time of gene therapy treatment. By targeting the first years of life, when brain plasticity is optimal, the chances of these young children with pre-linguistic hearing loss acquiring normal speech and language are maximized. The study comprises two cohorts of two doses followed by an expansion cohort at the selected dose. While safety will be the primary endpoint of the first part of the dose escalation study, auditory brainstem response (ABR) will be the primary efficacy endpoint of the second part of the expansion. Audiogene will also evaluate the clinical safety, performance and ease-of-use of the delivery system developed by Sensorion.

About SENS-501

SENS-501 (OTOF-GT) is an innovative gene therapy program developed to treat a specific form of congenital deafness linked to mutations in the OTOF (otoferlin) gene. This gene plays a key role in the transmission of auditory signals between the hair cells of the inner ear and the auditory nerve. When this gene is defective, affected individuals are born with severe to profound hearing loss.

The aim of SENS-501 (OTOF-GT) is to restore hearing by introducing a functional copy of the OTOF gene directly into hair cells via viral vector technology (AAV). By replacing the defective gene, this therapy aims to restore the normal process of converting sound into electrical signals, enabling patients to regain their hearing ability. Currently in clinical research phase, this gene therapy program represents significant hope for families affected by this rare form of genetic deafness. SENS-501 (OTOF-GT) embodies a commitment to scientific innovation in the field of hearing, with the potential to dramatically improve the quality of life of patients suffering from genetic deafness.

This gene therapy for patients suffering from otoferlin deficiency has been developed in the framework of RHU AUDINNOVE, a consortium composed of Sensorion with the Necker Enfants Malades Hospital, the Institut Pasteur, and the Fondation pour l'Audition. The project is partially financed by the French National Research Agency, through the "investing for the future" program (ref: ANR-18-RHUS-0007).

About Sensorion

Sensorion is a pioneering clinical-stage biotech company, which specializes in the development of novel therapies to restore, treat, and prevent hearing loss disorders, a significant global unmet medical need. 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 mechanisms of action for drug candidates.

It has two gene therapy programs aimed at correcting hereditary monogenic forms of deafness, developed in the framework of its broad strategic collaboration focused on the genetics of hearing with the Institut Pasteur. SENS-501 (OTOF-GT) currently being developed in a Phase 1/2 clinical trial, targets deafness caused by mutations of the gene encoding for otoferlin and GJB2-GT targets hearing loss related to mutations in GJB2 gene to potentially address important hearing loss segments in adults and children. The Company is also working on the identification of biomarkers to improve diagnosis of these underserved illnesses.

Sensorion's portfolio also comprises clinical-stage small molecule programs for the treatment and prevention of hearing loss disorders. Sensorion's clinical-stage portfolio includes one Phase 2 product: SENS-401 (Arazasetron) progressing in a planned Phase 2 proof of concept clinical study of SENS-401 in Cisplatin-Induced Ototoxicity (CIO) and, with partner Cochlear Limited, has completed in a study of SENS-401 in patients scheduled for cochlear implantation. A Phase 2 study of SENS-401 was also completed in Sudden Sensorineural Hearing Loss (SSNHL) in January 2022.

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