Press release

Sensorion Announces Candidate Selection for GJB2 Gene Therapy Program

- **Hosts R&D Day showcasing gene therapy for inner ear diseases April 6, 2023**

Montpellier, April 6, 2023 – Sensorion (FR0012596468 – ALSEN) a pioneering clinical-stage biotechnology company which specializes in gene therapies in the inner ear, will present new data supporting GJB2-GT program candidate selection to address hearing loss related to mutations in the *GJB2* gene during the Company's R&D Day taking place today, Thursday, April 6, 2023 (event details below).

The candidate designed with a specific adeno-associated virus (AAV) capsid targets key cells in the ear that normally express GJB2 and avoids ototoxicity.

Sensorion will advance the candidate into IND-enabling activities to enable clinical development in three pathologies related to *GJB2* mutations: early onset of age-related hearing loss in adults, progressive forms of hearing loss in children, and pediatric congenital deafness.

Géraldine Honnet, M.D., Chief Medical Officer of Sensorion, said: "We are very proud to have selected a candidate for our GJB2-GT program. This program is the second in an exciting portfolio of gene therapy candidates, developed in collaboration with the Institut Pasteur, and it has the potential to provide long-term therapeutic solutions to hearing loss for patients with *GJB2* mutations.

"Sensorion believes gene therapy has the potential to transform lives and we are delighted to be moving this promising candidate forward towards clinical development. We believe this is the optimal candidate and is well supported by our internal CMC capabilities, allowing streamlined tech transfer to CDMOs."

Sensorion, in collaboration with the Institut Pasteur, has generated positive Proof of Concept (POC) data in Non-Human Primates and mouse models recapitulating both human congenital and progressive conditions, confirming the potential of the candidate to treat GJB2-related hearing loss.

Professor Christine Petit, Professor at the Institut Pasteur and Professor at the Collège de France, said: "Genetic forms of hearing loss remain incurable. The knowledge of the genetic architecture of human deafness has paved the way for the development of treatments, based on gene therapy in particular. The design, optimization and accurate definition of the range of application of gene therapy agents are critically dependent on relevant models for the human deafness forms caused by diverse pathogenic variants of deafness genes.

"In our collaboration with Sensorion on gene therapy for the various forms of deafness due to defects of the GJB2 gene encoding connexin 26, the development of the various GJB2 deficits-related mouse models recapitulating clinical indications have allowed us to advance the preclinical development of a promising gene therapy product targeting cochlear cells that critically express connexin 26."

GJB2 mutations are the most prevalent form of congenital deafness and are thought to affect around 300,000 paediatric and adult patients in the US, Europe and Japan. Severity varies from mild to profound but is typically severe to profound.

Sensorion also plans to file a Clinical Trial Application for its OTOF-GT program, a dual-vector AAV gene therapy program for the treatment of children born with hearing loss caused by otoferlin deficiency this quarter. Sensorion has been granted Orphan Drug Designate status for OTOF-GT by the US Food and Drug Administration and by the European Medicines Agency.

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R&D Day showcasing gene therapy for inner ear diseases

Sensorion will host an R&D Day today, 6 April 2023, at the Hearing Institute (Institut de l'Audition), an Institut Pasteur center, located at 63, rue de Charenton, 75012, Paris, France. The event will take place at 8am EDT / 2pm CEST, a live webcast and a replay of the presentation will also be accessible. To register for the webcast, please sign up here.

Sensorion's management team, including CEO Nawal Ouzren, Géraldine Honnet, M.D., Chief Medical Officer, Laurent Désiré, Ph.D., Head of Preclinical Development, and Christine Le Bec, Ph.D., Head of CMC Gene Therapy, will provide an update on the Company's development programs and expected milestones in 2023.

Key Opinion Leaders (KOLs) from the field of hearing therapeutics will also give presentations related to patient needs, therapeutics and market landscape in key areas including hereditary monogenic forms of deafness.

External speakers participating in this event include:

- Professor Christine Petit, M.D, Ph.D., Professor at College de France, Professor "Classe exceptionnelle" at Institut Pasteur, Director of the Laboratory for innovation in hearing therapies at the Hearing Institute, Paris, France
- Barbara Kelley, Executive Director of the Hearing Loss Association of America (HLAA), the nation's leading organization representing patients with hearing loss, Washington D.C., USA
- 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, Paris, France

Agenda Overview:

- **Welcome and Introduction**, *Nawal Ouzren*
- **Incorporating the Patient Voice for Tomorrow's Care**, *Barbara Kelley*
- **Deafness: From Genetic Architecture to Gene Therapy**, *Pr. Christine Petit*
- **GJB2-GT Program: Data to Drive Next Steps***, Dr. Laurent Désiré*
- **GJB2-GT Program: Natural History Studies to Prepare Execution of Clinical Trials**, *Dr. Géraldine Honnet*
- **Otoferlin Deficiency: Approaches towards Hearing Restoration**, *Pr. Natalie Loundon*
- **OTOF-GT Program: SENS-501, Sensorion's Lead Gene Therapy Program**, *Dr. Laurent Désiré*
- **Enabling Reliable Gene Therapy Manufacturing and Analytical Control**, *Dr. Christine Le Bec*
- **Q&A Session and Closure**

About Sensorion

Sensorion is a pioneering clinical-stage biotech company, which specializes in the development of gene therapies to restore 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. OTOF-GT 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, 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.
www.sensorion.com

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