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(71) Applicant: **RESCUE HEARING INC.** [US/US]; 2148
Northwest 29th Avenue, Gainesville, FL 32605 (US).

(72) Inventors: **STAECKER, Hinrich**; 2209 West 125th
Street, Leawood, KS 66209 (US). **AYALA, Caesar,
James**; 2148 NW 29th Avenue, Gainesville, FL 32605
(US).

(74) Agent: **PATEL, Rikesh P.**; Wiggin and Dana LLP, One
Century Tower, 265 Church Street, New Haven, CT 06510
(US).

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(54) Title: GENE THERAPY SYSTEMS AND RELATED METHODS FOR TREATMENT OF HEARING LOSS

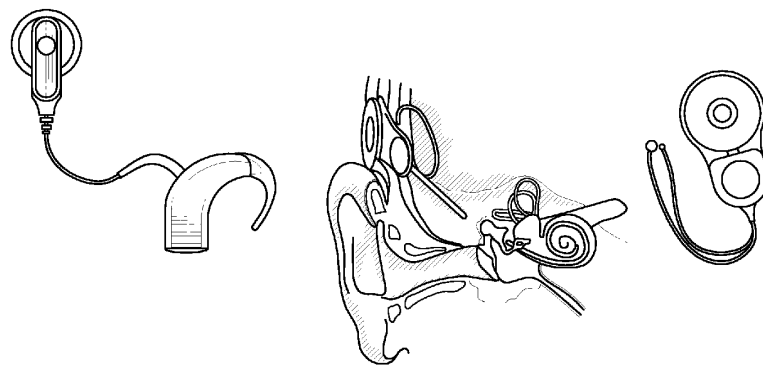


Figure 6

(57) Abstract: The present disclosure describes gene therapy systems, and related methods, useful for treating and/or preventing deafness caused by genetic mutation of the *TMPRSS3* gene or the *LOXHD1* gene. The compositions and methods disclosed herein use adeno-associated viral (AAV) vector gene delivery of *TRMPSS3* or *LOXHD1* into the inner ear to restore activity of the *TMPRSS3* gene or the *LOXHD1* gene, respectively, promote hair cell survival and restore hearing in patients suffering from hearing loss. As disclosed herein, the systems and methods may utilize a combination of gene therapy (e.g., molecular therapeutics) for hearing loss caused by a genetic mutation together with implantation of a cochlear implant.

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