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Japan takes step forward towards treating hereditary deafness

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Researchers at Juntendo University in Japan report in the journal *Human Molecular Genetics* that a stem-cell technique may be used to develop a treatment of a common mutation-related type of deafness.

The method involves reproducing cells occurring in the human inner ear where the researchers emphasize that 'the pathological condition could be reproduced with iPS derived from a typical patient.'

Deafness is the most common sensory impairment in newborn children — about 1 child in 1000 is born with hearing loss, or develops it in early childhood. Half of these cases have a genetic cause; very often, this type of deafness is related to a mutation of a gene called 'gap junction beta 2 (GJB2)', which encodes a protein called 'connexin 26 (CX26)'. This protein occurs in cells in the cochlea, the part of the inner ear enabling hearing.

One way of treating GJB2-related deafness would be to reproduce properly functioning cochlear cells and introduce them into the inner ear. Now, Kazusaku Kamiya from Juntendo University and colleagues have made an important step forward toward making this treatment possible: using a stem-cell technique, they succeeded in reproducing cochlear supporting cells, a type of cell known to contain CX26 proteins.

The researchers started from human induced pluripotent stem cells (iPSCs) — artificially generated cells that can differentiate into any other cell type of the human body.

According to the scientists: "Such iPSC-derived cells should be particularly useful for drug screening and inner-ear cell therapies with genome editing targeting GJB2-related hearing loss and the pathological condition could be reproduced with iPS derived from a typical patient".