Osteopetrosis is a rare and heterogeneous group of hereditary skeletal dysplasia syndromes. Manifestations of this condition are common in the temporal bone, particularly with regards to hearing loss. We present the case of a 41 year old male with a well-documented history of osteopetrosis and longstanding, profound, bilateral sensorineural hearing loss who underwent cochlear implantation.

**Abstract**

**Background**

Osteopetrosis is a heterogeneous family of rare genetic disorders characterized by increased bone density due to defective osteoclast function. Sclerotic bone formation in the temporal bone can lead to manifestations of hearing loss in multiple ways. For example, compression of the cochlear blood supply or compression of the cochlea within the narrowed IAC, can lead to a sensorineural loss, while fixation of the ossicular chain and obliteration of the round and oval windows can lead to a conductive loss.

**Background**

The patient was counseled extensively on the possible risks and outcomes of cochlear implantation, including lack of improvement in speech perception post-operatively as well as risks associated with obliteration of the external auditory canal. A transcanal approach to the cochlea with a blind sac of the external auditory canal was performed. Intraoperatively, the entire medial wall of the mesotympanum was found to be ossified and the round window was not visualized. A cochleostomy was performed in the basilar turn and a Cochlear Corporation Contour Advance electrode was fully inserted. Intra-cochlear placement was confirmed by an intra-operative X-ray and intraoperative neural response telemetry detected an auditory response to electrical stimulation. Post-operative high resolution CT scan confirmed correct placement and verified full insertion of the patient’s electrode (Figures 3 and 4). The patient tolerated the procedure well with postoperative complications. He demonstrated significant improvement in his access to speech sounds with an average aided threshold of 20 dB in his implanted ear at his two month follow-up visit, with

1. Pre-operative axial CT scan showing diffuse bony sclerosis and thickening of the temporal bone around the otherwise normal appearing bilateral cochlea and labyrinth
2. Pre-operative T2 MRI demonstrating an intact but poorly defined and notably small cochleovestibular nerve (white arrow)
3. Post-operative axial CT scan showing CI electrode positioned within the right cochlea (black arrow)
4. Post-operative coronal CT scan, again showing CI electrode within the right cochlea (black arrow), as well as its connecting wire traveling through the blind sac external ear canal (red arrow)

**References**