Bilateral cochlear implantation in a patient with osteopathia striata Voorhoeve and cranial sclerosis

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Introduction
Osteopathia striata Voorhoeve with cranial sclerosis is a rare X-linked dominant inherited bone dysplasia, characterized by longitudinal striations of long bones and cranial sclerosis. Patients can be asymptomatic or present with typical facial dysmorphism, sensory defects, internal organs anomalies, growth and mental retardation, depending on the severity of the disease. The WTX gene has been recently identified as the disease causing gene.

Case Report
We present here the case of a 13 year old girl with osteopathia striata exhibiting bilateral sensorineural hearing loss, microcephaly with mental retardation, hyperopia, astigmatisms and strabismus. The mother also suffers from osteopathia striata and sensorineural hearing loss however not as severe. Due to her mental retardation the girl's vocabulary was limited. However, over the last month her pronunciation grew sloppy and her vocabulary seemed even more restricted. Cochlea implantation was performed on the left ear first and showed good results with hearing gain between 20 and 30 dB HL. With progressive hearing loss despite hearing aid on the right side cochlear implantation was performed about eight months after the left side. Clinical evaluation showed functional deafness on the left ear and severe sensorineural hearing loss on the right ear.

Radiology
CT and MRI diagnostic showed sclerosis of the temporal and petrous bone on both sides with narrowing of the inner ear canal. Despite this the labyrinth showed normal configuration with normal fluid signal on both sides. The auditory nerve was intact and showed no sign of degeneration.

Results
Postoperative CT scan shows both electrodes in the intended intracochlear position on both sides.

Discussion
This case is to our knowledge, the first describing bilateral cochlear implantation in a case of Osteopathia striata Voorhoeve with cranial sclerosis. The hearing loss could be attributed to the massive sclerosis of the cochlear capsule or due to compression of the blood supplying artery in the narrow inner auditory canal. Despite mental retardation the patients benefit was immense. However we are aware that progressive sclerosis can lead to further narrowing of the internal canal or even complete closure thereby damaging the auditory nerve irreversibly.

Literature
1Osteopathia striata-cranial sclerosis: otorhinolaryngologic clinical presentation and radiologic findings. Maglulo G et al. Am J Otolaryngol. 2007 Jan-Feb;28(1):59-63.
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