As in all forms of MPS, reduction in hearing in patients with MPS IVA can be attributed to multiple causes. Firstly, conductive hearing loss can be present and is most likely secondary to recurrent upper respiratory tract infections and frequent serous otitis media. Conductive loss can also be caused by deformity of the ossicles. Secondly, sensorineural loss may occur as a result of GAG accumulation. Abnormal auditory brainstem response (ABR) results have been described and are thought to be a combination of middle ear, cochlear, eighth nerve, and lower brainstem pathology. Reduced GALNS activity results in impaired catabolism of two glycosaminoglycans (GAGs), chondroitin-6-sulfate (C6S) and keratan sulfate (KS). Clinical presentations of MPS IVA reflect a spectrum of progression from a severe “classical” phenotype to a mild “attenuated” phenotype. 180 mutations have been identified in the GALNS gene, which likely explains the phenotypic heterogeneity of the disorder. **MATERIALS AND METHODS:** Fourteen patients were evaluated. All the patients underwent a tonal audiometry to determine their auditory thresholds, both aerial and osseous, likewise they evaluated low, medium and acute frequencies, thus being able to identify the type and degree of hearing loss. **RESULTS:** The following alterations were identified: mixed hearing loss 50% (7/14), sensorineural hearing loss 29% (4/14) and 21% (3/14) normal audiometry. All patients had the 901G> T (GLY301CY) mutation (11 in homozygotes and 3 in heterozygosis). 100% of patients in heterozygosis did not have hearing impairment. **CONCLUSIONS:** The hearing losses generate in the patients difficulties in their school development and in the acquisition of language. It is important to perform hearing evaluations at an early age to obtain a diagnosis and timely treatment. Prophylactic antibiotics may be useful to manage recurrent infections due to abnormalities in the ears, nose, and throat. As the disease progresses, patients may experience mixed hearing loss and may require tympanostomy tube insertion and/or hearing aids. It is important to know the genotype to identify the hearing prognosis of patients.