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Case report of dental management for a child with Mondini syndrome

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Mondini syndrome is a rare congenital sensorineural deafness. Malformation of cochlea in inner ear unilateral or bilateral may cause hear loss, recurrent meningitis and otorrhea. Mondini syndrome can be isolated or combined with other ear malformation or other syndrome. The condition occurs at seventh week of embryonic development. It is a case report of 5 years old female child patient came to the dental clinic complaining from pain in lower right second molar. First case in officer clinic diagnosed with profound hearing loss bilateral with sever Mondini deformity in the inner ear. Patients were not responding to any sounds. There is no history of any other congenital problems. There is positive history of consanguinity and there is no family history of congenital hearing loss. No history of trauma or meningitis, no significant perinatal history. The patient looks well, no dysmorphic features, normal external auditory canal and bilateral dull tympanic membrane. Examination was otherwise normal. CT revealed deformity in cochlea. Otoacoustic emissions and ABR was used for the diagnosis of severe to profound sensorineural hearing loss. Failed trial of cochlear implant, period of hearing aid trial failed to show any response status post brainstem implant done in Germany. Communication with hearing impaired patient has some difficulties and needs a good level of skills and awareness. In this case, it was the father helped with sign language and communicating with the child patient easier. Dentists should learn simple techniques for dealing with such impairment like removing mask to allow for lip reading, writing or drawing what they want to say or using sign language.

Biography

Badria Al Matrafi has completed her BDS in 1991 in King Saud University in Riyadh KSA and AGD certificate in 2000 from University of South California, USA. She is a Consultant Restorative Dentistry in Prince Sultan Military Medical City in Riyadh. She is the Director of dental clinic and has a year of teaching and clinical supervision experience.

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