



## Dental Management for Patient with Mondini Syndrome

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### ABSTRACT

Mondini syndrome is a rare congenital sensorineural deafness. Malformation of cochlea in inner ear is 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. We report 4 years old female child come to our dental clinic diagnosed with profound hearing loss bilateral with sever mondini deformity in the inner ear, status post brainstem implant done in Germany. Patient is 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 mild deformity in cochlea. Otoacoustic emissions and ABR consist of the diagnosis of severe to profound sensorineural hearing loss. Failed trial of cochlear implant, period of hearing aid trial failed to show any response. The communication with hearing impairment child has some difficulties and need some skills and awareness.

### INTRODUCTION

Mondini syndrome is a rare congenital sensorineural deafness. Malformation of cochlea in inner ear is unilateral or bilateral. May cause hear loss, recurrent meningitis and otorrhea. Mondini syndrome can be isolated or combined with other ear malformation or other syndromes. The condition occurs at seventh week of embryonic development due to arrested development of cochlea. Mondini dysplasia [MD] was first described in Latin by Carlo Mondini: Italian anatomist in 1791 after dissecting the inner ear of an 8 year old deaf boy. Mondini dysplasia is congenital malformation of cochlear with dilatation of the vestibule, aqueduct, and ampulla, and incomplete partition of the cochlea.

It can be unilateral or bilateral, profound sensorineural hearing loss or with residual hearing or normal hearing. Cerebrospinal fluid (CSF) leakage is relatively common, and CSF rhinorrhea or otorrhea may develop. Recurrent meningitis is easily complicated and leads to a diagnosis of Mondini dysplasia. As a health care provider there is difficulty to communicate with hear impairment patients due to little awareness with communication tools. The purpose of this case report to focus how to treat with Mondini syndrome and deaf patient in dental clinic [1-8].

### CASE REPORT

5 years old female child patient of 16 kg body weight come to the

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dental clinic complaining from pain in lower right second primary molar. First case in officer dental clinic diagnosed with profound hearing loss bilateral with sever Mondini deformity in the inner ear. Patient is 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 consist of 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 (Figure 1).



Figure 1: CT revealed deformity in cochlea.

## DISCUSSION

Our case was the first Mondini case in our dental officer clinic. The prevalence of sensorineural hearing loss (SNHL) is (1.5%) in all Saudi children who have been surveyed (1–4 in 1000). That means its high compared to the other developed countries. Although we have hearing and speech centers but still, we need more centers and more training, for training of personnel, parents' education and public awareness [9]. Sensorineural deafness constituted 16.2% of all hearing impairment cases in Saudi children in Riyadh [10]. "Cochlear malformations have been reported to occur in approximately 20% of children with congenital sensorineural hearing loss. Jensen studied 62 children enrolled in a school for the deaf with polysomnography and detected cochlear malformations in only 15%. In a recent series of 234 children who had SNHL of varying degrees of severity, Reilly found cochlear anomalies in only 4% of those evaluated by high resolution computed tomography (CT) scanning. In unilateral profound deafness, Ever berg detected only a 2% incidence of cochlear malformation among the 122 children studied. In analyzing each of these studies, children with radiographic signs of labyrinthine ossification were not included in the calculation of incidence of congenital malformation, as this finding is usually a marker for acquired deafness, typically due to meningitis.

Incomplete partition (mondini) malformation is the most common type of cochlear malformation, accounting for over 50% of all cochlear deformities. The others cochlear anomalies are common cavity, cochlear hypoplasia, cochlear aplasia, complete lab-

yrinthine aplasia. Removing masks while talking, reducing background noise and learning to use simple signs and pictures may improve communication with hearing impaired children. When communicating with a child, be sure to face the child, the right way up and ensure the light is not in the child's eyes or behind your head devise a deaf friendly call system such as the dentist going into the waiting room to take the child. Facial expressions and gestures may be very helpful, writing and lip reader [11-14]. 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 technique for dealing with such impairment like moving mask to allow for lip reading, writing or drawing what they want to say or using sign language.

## CONCLUSION

Dentists should learn simple technique for dealing with such impairment like moving mask to allow for lip reading, writing or drawing what they want to say or using sign language and more speech and hearing centers to provide care for those who have hear impairment.

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