

전음성 난청이 동반된 하악안면이골증 1례

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A Case of Conductive Hearing Difficulty on Treacher Collins Syndrome

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—ABSTRACT—

Treacher-Collins Syndrome, also called mandibulofacial dysostosis, affects the shape of the head and face. Small and slanting lower jaw, down slanting eyes, notched lower eyelids, lack or absence of the lower eyerash, hypoplasia or absence of the zygomatic processes and flattening of the frontonasal angle are characteristics of Treacher-Collins Syndrome. Most children with Treacher-Collins have normal development and intelligence. They have 40% hearing loss in each ear due to abnormalities of the outer and middle ear, which conduct sound to the nerve endings. This paper reviewed a case of bilateral conductive hearing loss on Treacher-Collins Syndrome. (J Clinical Otolaryngol 2003;14:302-305)

KEY WORDS : Mandibulofacial dysostosis · Conductive · Hearing loss.

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1900 Treacher Collins

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Treacher - Collins

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Syndrome Franceschetti - Klein

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증례

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(Treacher Collins)

70 dB,

68%, 66%

(Fig. 1).

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46XY

(Fig. 2).

As , C

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50 dB

(Fig. 3). 가

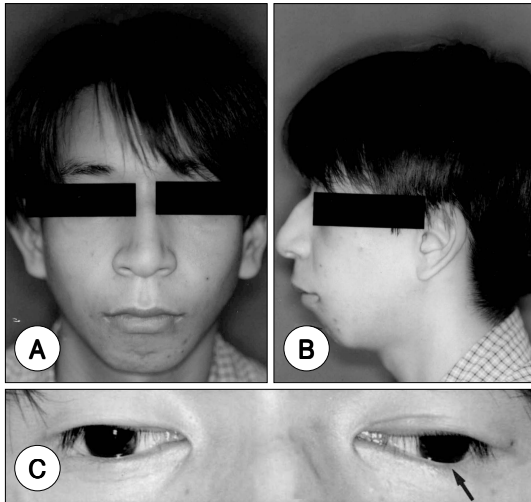


Fig. 1. A : 20-years old man who have malformed cheek bones. B : Receding lower jaw (arrow) C : Drooping eyelid, absent of lower eyelashes.

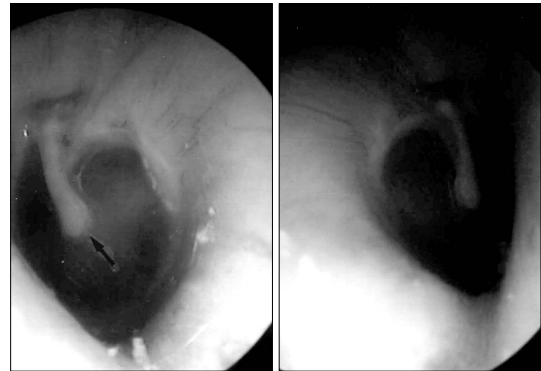


Fig. 2. Otoscopic finding. Dark gray colored tympanic membrane, loss of cone of light, deformed flat appearance of manubrium (arrow) and external auditory canal narrowing are noted.

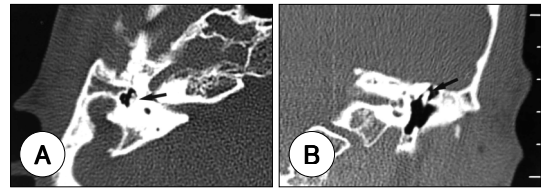


Fig. 4. A : Axial CT image shows metallic screw (arrow) on right zygomaticomaxillary junction area. B : Axial CT image shows also metallic screw (arrow) on left zygomaticomaxillary junction area, defect of posterior portion of zygomatic arch and hypoplastic both temporal bone.

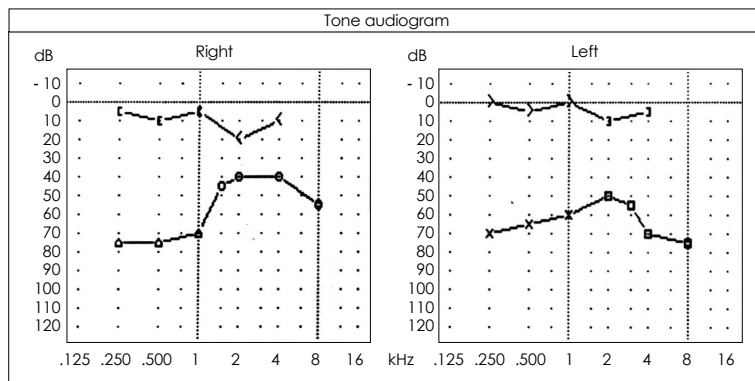


Fig. 3. Pure tone audiogram shows severe conductive hearing loss pattern.

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