

이구개지단 증후군에서 나타난 선천성 등골 고정증

박기현 · 이동훈 · 김성균

Congenital Stapedial Fixation in Oto-Palato-Digital Syndrome ; A Case Report

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ABSTRACT

The Oto-palato-digital syndrome is a generalized skeletal dysplasia resulting in short stature, characteristic facies, and unusual deformities of the hands and feet. In addition, cleft palate and other congenital malformations are present. Some patients have conductive deafness due to dysplastic auditory ossicle which is sometimes stapes fixation. We experienced a case of Oto-palato-digital syndrome which showed bilateral stapes fixation, high-arched palate and syndactyly of toes. Congenital stapes fixation was treated through partial stapedectomy with good hearing gain. (**Korean J Otolaryngol 1998;41(3):405-408**)

KEY WORDS : Oto-palato-digital syndrome · Congenital stapes fixation · Syndactyly.

1962 Taybi¹⁾가
 50 dB(Fig. 3) 45 dB(Fig. 4)
 , tympanogram type
 As (Fig. 5).
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 1995 7 27
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 suprastructure drill
 Cause loop piston(Xomed)
 (stapes prosthesis)(Fig. 7)
 Fisch stainless steel piston(Xomed)
 (Fig. 8)
 가 25 dB,
 15 dB
 (Figs. 3 and 4).

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Hanson²⁾ (1962)

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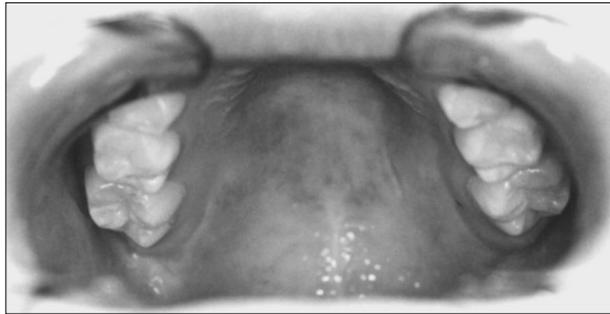


Fig. 1. High arched palate.



Fig. 2. Syndactylism of both second and third toes.

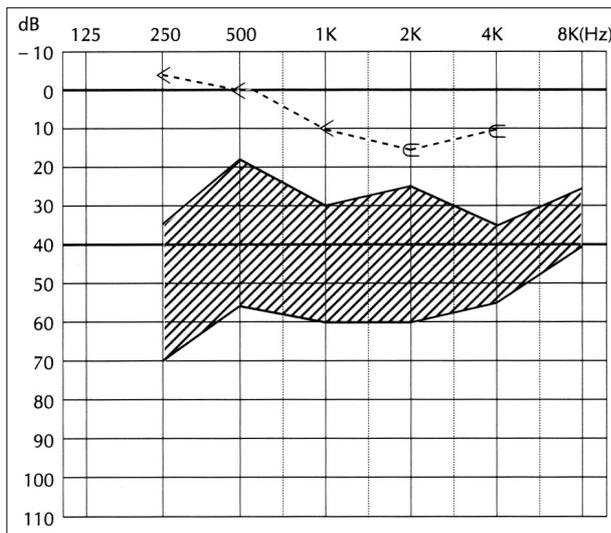


Fig. 3. Preoperative and postoperative pure tone audiogram, right dash lines represent hearing improvement after surgery.

Bast³⁾ (1949) Anson⁴⁾ (1959)
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Schlosser⁵⁾ (1964)

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House⁶⁾(1958)

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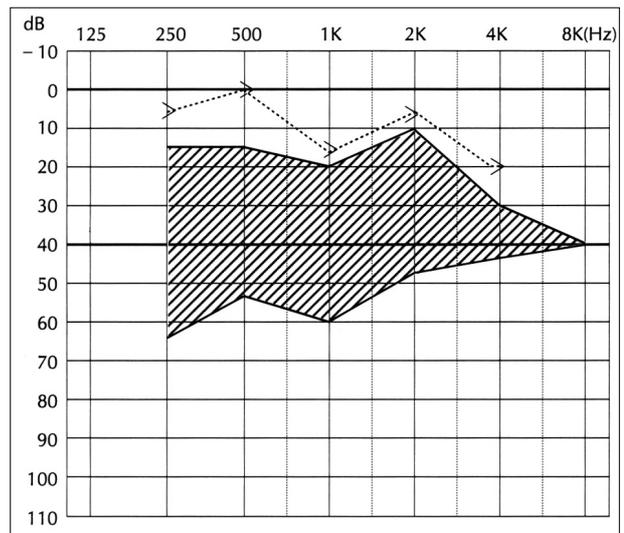


Fig. 4. Preoperative and postoperative pure tone audiogram, left dash lines represent hearing improvement after surgery.

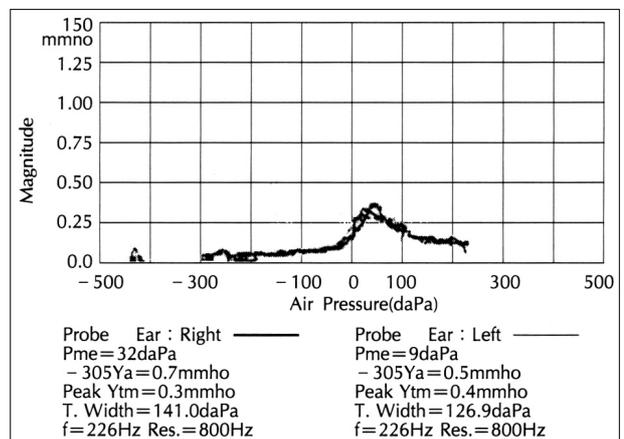


Fig. 5. Tympanogram : both type as.

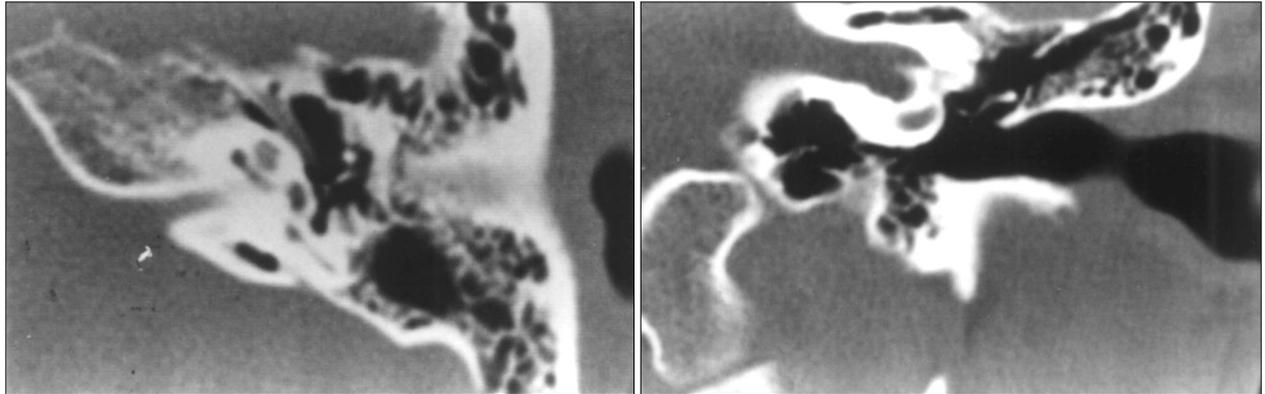


Fig. 6. Axial and coronal temporal bone CT show normal ossicular finding.



Fig. 7. Causse loop piston (Xomed).

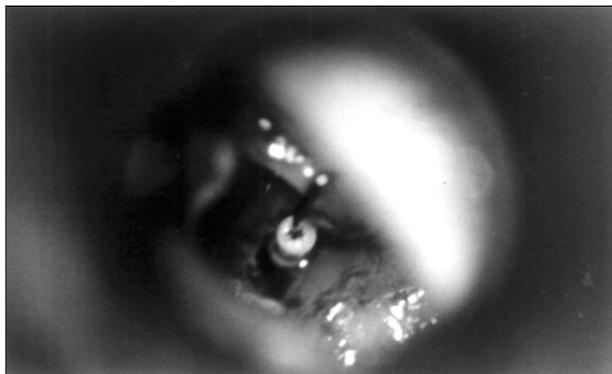


Fig. 8. Fisch stainless steel piston (Xomed).

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 Kim¹¹⁾ (1981)
 , Lee¹²⁾ (1990) 1

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1970 Gorlin⁷⁾

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