

정상고막을 가진 선천성 이소골 기형

박기현 · 문성균 · 정연훈 · 최호석

Congenital Ossicular Anomaly with Intact Tympanic Membrane

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ABSTRACT

Background and Objectives : A nonprogressive and conductive hearing loss in the range of 40 to 60 dB with normal tympanic membrane which has no history of trauma or infection is highly suggestive of a congenital ossicular malformation. Stapes anomaly is the most common among such ossicular anomalies and it is clinically important because hearing loss can be corrected by appropriate procedures. In this study, we attempted to describe the patterns of ossicular anomalies encountered in patients who have a normal eardrum. We also analyzed the surgical result with special reference to the pattern of ossicular anomaly. **Materials and Method** : We experienced 28 ears (23 patients) of congenital ossicular anomalies which were confirmed through the exploratory tympanotomy at Ajou University Hospital between September 1994 and December 2001. The preoperative and postoperative audiologic findings, operative findings and CT findings were analyzed. **Result** : There were 7 patterns of ossicular anomalies among which stapes footplate fixation was the most common anomaly. We could get 64.3% of hearing improvement after ossiculoplasty with several kinds of prosthesis. **Conclusion** : There was a great diversity of patterns of ossicular anomalies without certain rule. Stapes anomaly was the most commonly detected. Stapes footplate fixation was the most common type which was usually bilateral. Anomalies associated with incus and malleus were usually unilateral. The surgical result of congenital ossicular anomaly was generally good. (Korean J Otolaryngol 2002;45:952-6)

KEY WORDS : Conductive hearing loss · Congenital anomaly · Ear ossicles.

60 dB

가

2)

1)

가

1994 9 2001 12 8

23

(28)

40

: 2002 1 8 / : 2002 8 7

: , 442 - 721 5

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24.8 , 가 13 , 가 10

As , 8 (29%) A , 7 (25%)
 Ad
 40 dB 2 , 41 50 dB 17 , 51 60 dB 8 , 61 dB
 1

(prosthesis)

Fisch type

(PORP)

(TORP)

2000, 3000 Hz

500, 1000,
 가 20 dB

(Fig. 1),

가 4 (Fig. 2),

15 가

가 3

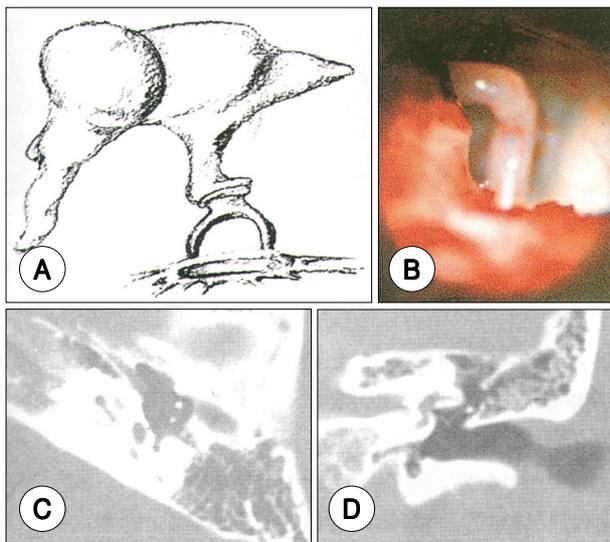


Fig. 1. Stapes footplate fixation A : Schematic view. B : Operative finding shows intact incudo-stapedial joint. C : Temporal bone CT shows normal ossicular chain on axial view. D : Temporal bone CT shows normal ossicular chain on coronal view.

(Fig. 3),

(Fig. 4),

가 1 (Fig. 6),

7가

가 2

가 2 (Fig. 5),

(obturator foramen)

가 1 (Fig. 7)

(Table 1).

16 (57.1%)

12 (42.9%)

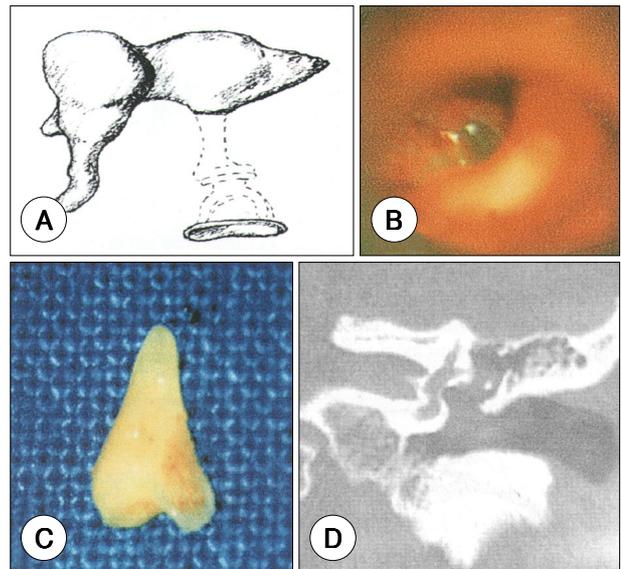


Fig. 2. No suprastructure of stapes and absent long process of incus. A : Schematic view. B : Operative finding shows no suprastructure of stapes. C : Removed incus shows missing long process. D : Temporal bone CT shows ossicular chain discontinuity on coronal view.

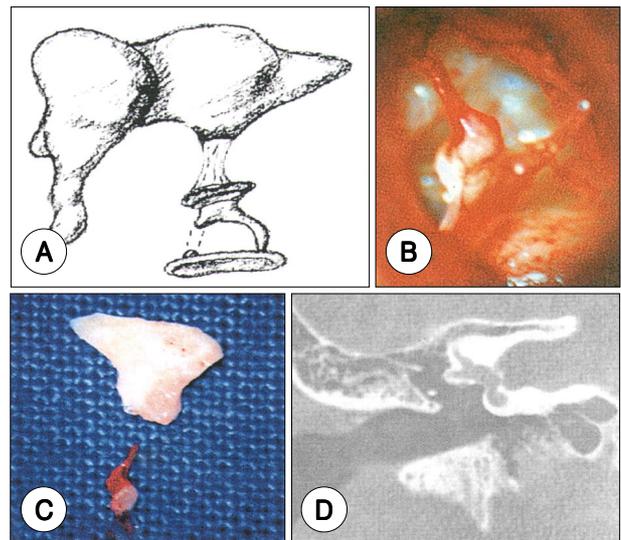


Fig. 3. No anterior crus and absent long process of incus. A : Schematic view. B : Operative finding shows no anterior crus and absent long process of incus. C : Removed incus shows no long process, and incidentally removed stapes shows no anterior crus. D : Temporal bone CT shows ossicular chain discontinuity on coronal view.

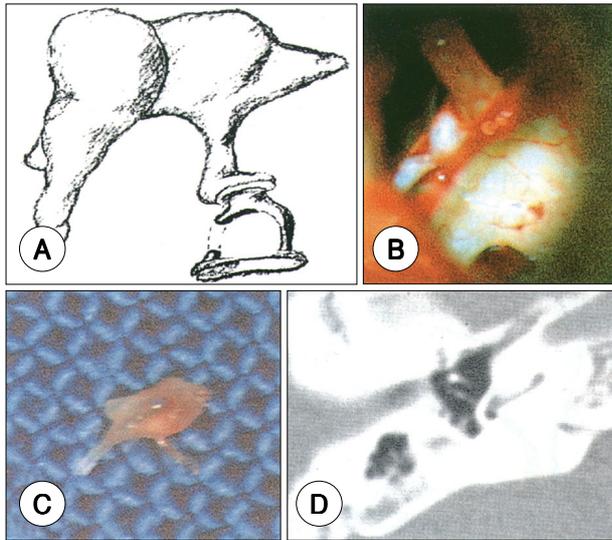


Fig. 4. No anterior crus of stapes. A : Schematic view. B : Operative finding shows no anterior crus of stapes. C : Incidentally removed stapes shows no anterior crus. D : Temporal bone CT shows only one crus of stapes on axial view.

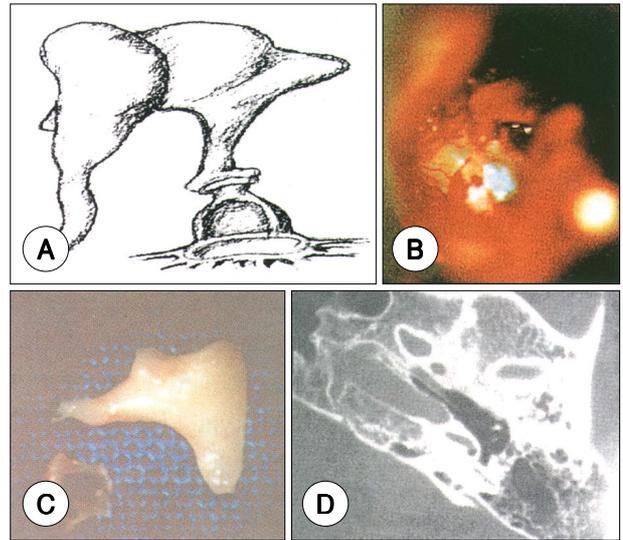


Fig. 6. Nonperforated obturator foramen with footplate fixation. A : Schematic view B : Operative finding shows an abnormal shape of the stapes. C : Removed ossicles show stapes with nonperforated obturator foramen and incidentally removed normal incus. D : Temporal bone CT shows intact ossicular chain on axial view.

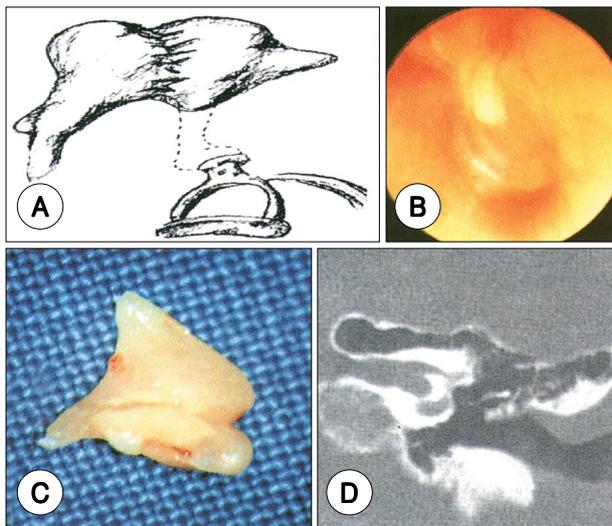


Fig. 5. Fused malleus and incus with absent long process of incus. A : Schematic view. B : Drum photograph shows intact tympanic membrane with abnormal malleus handle. C : Removed ossicle shows fused malleus and incus. D : Temporal bone CT shows lateral displacement of ossicular chain on coronal view.

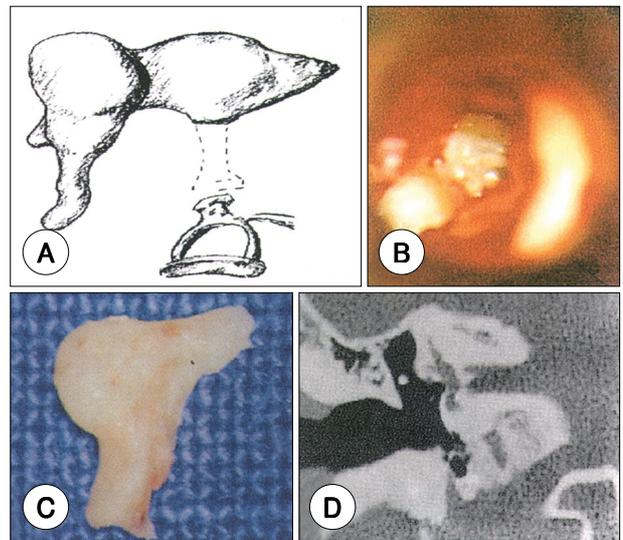


Fig. 7. Absent long process of incus. A : Schematic view. B : Operative finding shows absent long process of incus. C : Removed incus shows absent long process. D : Temporal bone CT shows ossicular chain discontinuity on coronal view.

			1	
		16	10 (63%)	As
			12	7
(58%)	Ad			
7가				
		10	8 가	

		가	28	
		64.3%	(Table 2),	
		가 91.7%		43.8%
		(Table 3).		
		5 dB	5 , 6	10 dB 1 , 11
				15 dB

Table 1. The patterns of ossicular anomalies (number of ears)

1. Stapedial footplate fixation (15)
2. No suprastructure of stapes and absent long process of incus (4)
3. No anterior crus and absent long process of incus (3)
4. No anterior crus of stapes (2)
5. Fused malleus and incus with absent long process of incus (2)
6. Nonperforated obturator foramen with footplate fixation (1)
7. Absent long process of incus (1)

Table 2. Hearing results according to patterns of anomalies (post-operative air-bone gap <20 dB)

Pattern	Number/Total (%)
1	7/15 (46.7)
2	4/ 4 (100)
3	3/ 3 (100)
4	1/ 2 (50.0)
5	2/ 2 (100)
6	0/ 1 (0)
7	1/ 1 (100)
Total	18/28 (64.3)

Table 3. Hearing results of according to status of footplate (post-operative air-bone gap <20 dB)

	Number/Total (%)
with stapes footplate fixation	7/16 (43.8)
without stapes footplate fixation	11/12 (91.7)
Total	18/28 (64.3)

Table 4. Hearing gain according to postoperative air-bone gap

	Number (%)
0 - 5 dB	5 (17.9)
6 - 10 dB	1 (3.5)
11 - 15 dB	3 (10.7)
16 - 20 dB	9 (32.1)
20 dB -	10 (35.7)
Total	28 (100)

3 , 16 20 dB 9 , 20 dB 10 (Table 4).

1 , 2 , 1 , 2 , 가

Jahrsdoerfer⁴⁾ (major type) (minor type) , Nomura⁵⁾ 52 가 가 29 (56%) Dasgupta Kacker⁶⁾ 11 , Koide⁷⁾ 6 가 가 1993 Teunissen Cremer⁸⁾ 144 4가 7가 Choi(1993)⁹⁾ 8 6가 , Park(1994)²⁾ 34 9가 18 가 Lee(2001)¹⁰⁾ 10 6가 4 가 가 Choi(1993)⁹⁾ 8 6가 , Park(1994)²⁾ 34 9가 18 가 Lee(2001)¹⁰⁾ 10 6가 4 가 가 laser drill 11)12) 가 11)13) 14)15) 가 14)16-18) 가 1)3) 20 dB Teunissen Cremer⁸⁾ 144 72.2% , Oh¹⁴⁾ 17 13 (77%) , Park¹³⁾ 19 15 (78.9%)

