

한국 Turner 증후군 환자에서의 이과적 발현

김 상 렬

Otologic Manifestations in Turner Syndrome in Korea

Sang Ryul Kim, MD

*Department of Otorhinolaryngology-Head & Neck Surgery, College of Medicine, Inje University,
Pusan Paik Hospital, Busan, Korea*

-ABSTRACT-

Background and Objectives : Patients with Turner's syndrome have been frequently suspected to have associated otologic disorders and hearing loss. The purpose of this study was to elucidate the otologic problems in Turner's Syndrome Patients, such as incidence, degree, and types of hearing loss. **Patients and Methods** : The 12 subjects aged 5 and 30 years who were previous diagnosed with Turner's syndrome were investigated for the otologic manifestations. All the patients were evaluated audiologically. **Results** : Clinical examination revealed the histories of otitis media in 3 subjects (25%). A audiological examination were performed for all subjects and revealed normal hearing in 8 subjects (62%), unilateral hearing loss in 2 subjects (17%), bilateral hearing loss in 2 subjects (17%). Type of the hearing loss was conductive type in 2 ears (8%), mixed type in 2 ears (8%), sensorineural type in 2 ears (8%). Degree of hearing loss was mild in 5 ears, moderate in 1 ear. **Conclusion** : Our results demonstrated that the otitis media and hearing loss were common for Turner's syndrome patients. The findings emphasize the importance of regular otological examinations and audiological evaluation of all girls with Turner's syndrome early in life. (J Clinical Otolaryngol 2005;16:41-44)

KEY WORDS : Turner syndrome · Hearing loss · Otitis media.

서 론

Turner

ear)

(rotated

.1) Turner

가

.2-5)

가

X

.1)

: 2005 3 25

: 2005 4 21

: , 614 - 735

633 - 165

.6)

Turner

12

: (051) 890 - 6379 · : (051) 892 - 3831

E - mail : iment1@yahoo.co.kr

Turner

대상 및 방법

1998 2 2004 3
Turner
12 3
5 30 13.9
Impedance Audiometer AT235h(Interacoustics, Denmark)
AUDIO screener(GSI, USA)
Biologic Navigator(Bio - logic Systems Corp., USA)
Clinical Audiometer AC40(Interacoustics, Denmark)

가 3
1 가
가 1 가
가
2
, 1
1
(Table 1).

청력 검사

4
1 B type
A type
1 (fail)
(pass)
가 8 , 2 , 2
가 2 , 가 2 ,
2 8000 Hz

결 과

병력 및 진찰 소견

가 1

Table 1. Clinical characteristics of Turner's syndrome

	Age	MEE Hx	Otoscopic findings	ABR	Karyotype
1	5	-	Normal	20/20	45, X (20)
2	8	+	Lt. Retraction	10/20	45, X (12)/47, XXX (16)
3	8	+	Both Retraction, Myringosclerosis	30/40	45, X (20)
4	11	-	Both Retraction	30/25	45, X/46, XX/47, XXX
5	12	-	Normal	25/10	45, X (20)
6	13	-	Normal	15/20	45, X (25)/46, XY (6)
7	13	+	Both Retraction, Myringosclerosis	40/40	45, X/46, Xi (Xq)
8	15	-	Normal	15/20	45, X (20)
9	15	-	Normal	10/25	45, X/46, XY
10	18	-	Normal	35/25	45, X/46, Xi (Xq)
11	19	-	Normal	25/15	45, X/46, Xr (X)
12	30	-	Both CholeOM	45/35	45, X

MEE : middle ear effusion

: Turner

고 찰

Turner 1,500~2,500 1 Anderson¹³⁾ 79 70% 64% Hulterantz⁷⁾ 50~90%

가 1 45,X가 , 45,X/46,XX, 45X/46,Xi(Xq), 45,X/46,X,r(X), 45,X/46,XY¹⁾ Watkin³⁾ Turner 4.5 7 Hulterantz⁷⁾ Turner 30~50%

Turner

가⁸⁾ , 2 , 2⁷⁾¹⁴⁾¹⁵⁾

⁹⁾ Schulerati¹⁰⁾ 가 1 5 Turner 가

Turner 53~90% 가

가 1¹¹⁾¹²⁾ Turner 가

Turner 가 2¹⁾

⁹⁾ Hulterantz⁹⁾ 50~80% 3 , 1

1 가 **결 론** Turner 12

가 2

4 , 1
 6 , 2 가 , 2 가
 , 2 가 , 2 가
 4 2
 Turner
 가 가
 중심 단어 : Turner
 2004

REFERENCES

- 1) Rapaport R. *Disease of the Gonads*. In: Behrman RE, Kliegman RM, Jenson BB. *NELSON'S TEXTBOOK of PEDIATRICS*. 17rd ed. Philadelphia: W.B. Saunders;2003. p.1921-46.
- 2) Turner HH. *Asyndrome of infantilism, congenital webbed neck and cubitus valgus*. *Endocrinology* 1938;23:566-74.
- 3) Watkin PM. *Otological disease. in Turner's syndrome*. *J Laryngol Otol* 1989;103:731-8.
- 4) Lippe BM. *Turner syndrome*. In: Sperling MA, ed. *pedi-*

- atric Endocrinology*. 1st ed. Philadelphia: W.B. Saunders; 1996. p.387-421.
- 5) Saenger P. *Turner's syndrome*. *N Engl J Med* 1996;335:1749-54.
- 6) Roush J, Davenport ML, Carlson-Smith C. *Early-onset sensorineural hearing loss in a child with Turner syndrome*. *J Am Acad Audiol* 2000;11:446-53.
- 7) Hulterantz M, Sylven L, Borg E. *Ear and hearing problems in 44 middle-aged women with Turner's syndrome*. *Hear Res* 1994;76:127-32.
- 8) Perrin P, Leheup B, Perrin C, Pierson M. *Turner's syndrome and ear*. *Rev Laryngol Otol Rhinol* 1988;109:55-9.
- 9) Stenberg AE, Nysten O, Windh M, Hulterantz M. *Otological problems in children with Turner's syndrome*. *Hear Res* 1998;124:85-90.
- 10) Schulerati N, Ledesma-Medina J, Finegold N, Stool S. *Otitis media and hearing loss in Turner's syndrome*. *Arch Otolaryngol Head Neck Surg* 1990;116:704-7.
- 11) Szpunar J, Rybak M. *Middle ear disease in Turner's syndrome*. *Arch Otolaryngol* 1968;87:34-40.
- 12) Leheup BP, Perrin P, Perrin C, Pierson M. *Otologic signs and early diagnosis of Turner's syndrome. Reevaluation of 30 cases*. *J Genet Hum* 1988;36:315-21.
- 13) Anderson H, Filipsson R, Fluor E, Koch B, Lindsten J, Wedenberg E. *Hearing impairment in Turner's syndrome*. *Acta Otolaryngol Suppl* 1969;247:1-26.
- 14) Leon Y, Vazquez E, Sanz C, Vega JA, Mato JM, Giraldez F, et al. *Insulin-like growth factor-1 regulates cell proliferation in the developing inner ear, activating glycosyl-phosphatidylinositol hydrolysis and Fos expression*. *Endocrinology* 1995;136:3494-503.
- 15) Bony CA. *Transient IGF-I gene expression during the maturation of functionally related central projection neurons*. *J Neurosci* 1991;11:3442-55.