

18. Waardenburg 증후군의 3예

연세의대

<지도: 김 기 령 교수>

김영명 · 조경열 · 이만용 · 박기현 · 김상기

Waardenburg 氏 증후군은 1951년에 Waardenburg 가 처음으로 기술한 이래 Partington(1959), Di George (1960)등에 의해 이 증후군에 대한 보고가 있었다.

선천성 난청의 1~7%를 차지하는 본 증후군은 선천성 난청 외에 內眼角의 측면전위, 백색증, 홍채이색증, 비근부의 편평 및 眉毛의 과도형성 등의 증상을 특징으로 한다.

본 증후군에서 청력장애는 출생후 서서히 나타날 수도 있고 청력장애의 정도 및 양상도 Fisch(1959)등의 보고에서 보듯이 다양하게 나타날 수 있다.

저자는 이 증후군의 3례를 체험하였기에 문헌적 고찰과 아울러 보고하는 바이다.

19. 우리나라 幼兒難聽의 原因에 관한 考察

서울醫大

盧寬澤 · 閔陽基 · 李熙培 · 高健成

言語는 聽覺을 통해서 발달하는 것으로서 難聽이 있어서 듣지 못하는 경우 그 사람의 言語發達은 停止되어 聾啞者가 되는 경우가 많다. 따라서 言語發達에 미치는 영향 때문에 幼兒難聽의 早期診斷 및 治療는 대단히 重要하다고 하겠다.

이에 著者들은 幼兒難聽의 原因을 調査함으로써 幼兒難聽의 豫防 및 治療에 도움을 주고자 1977年 1月부터 8月까지 8개월 동안에 本院 外來 難聽室로 來訪한 滿 5세 이하의 難聽兒 185명을 對象으로 하여 仔細한 病歷과 詳細한 臨床的 檢査 및 Impedance audiometer (Madsen ZO 70)에 의한 聽力檢査를 實施하고 그 原因에 대한 統計的 考察을 試圖하여 다음과 같은 結果를 얻었다.

1. 幼兒難聽 중 感覺神經性難聽의 가장 흔한 原因은 幼兒자신의 感染(123예 중, 44예, 35.8%)이었으며 妊娠中の 母體의 狀態(37예 30.1%), 分娩時 外傷 혹은 頭部外傷(18예, 14.7%)의 順이었다.

2. 幼兒難聽 중 傳音性難聽의 가장 흔한 原因은 急性 혹은 慢性中耳疾患이었으며(62예 중 32예, 51.6%), 다음으로서는 聾은 上氣道感染(19예, 30.6%), 아메노이드

增殖症(7예, 11.3%)의 順이었다.

3. 幼兒難聽은 外因性原因이 185예 중 179예(96.8%), 內因性原因이 6예(3.2%)로 대부분이 外因性 原因이었다.

20. Bing test 의 臨床聽覺學的 意義에 對한 考察

서울醫大

李熙培 · 車昌益 · 盧寬澤

傳音器管의 機能이 正常인 경우 1,000 Hz 以下の 低音域에서 骨導閾値는 外耳道를 閉塞하였을 때와 開放하였을 때 약 20 dB의 差異를 나타낸다. 그러나 外耳道를 閉塞하였을 때나 開放하였을 때 骨導閾値에 變化가 없으면 傳音器管에 病變이 있음을 意味한다.

1891년에 Bing이 처음으로 外耳道閉塞에 의한 骨導檢査를 發表한 以來 여러 學者들에 의하여 本檢査가 有用한 臨床聽力檢査의 한 方法으로서 認定되고 Bing test로 알려져 있다.

Sullivan은 1947년에 250 Hz, 500 Hz, 1,000 Hz에서 外耳道를 閉塞하였을 때와 開放하였을 때의 骨導閾値의 差異를 합한 것을 "Occlusion Index"라고 命名하였다.

이에 著者들은 Bing test의 臨床聽覺學的 意義를 檢討하고자 正常인 40耳, 慢性中耳炎患者 20耳, 感覺神經性難聽患者 14耳를 對象으로 純音聽力檢査, Bing test 및 impedance audiometry를 施行하고 occlusion index와의 相關關係를 檢討하여 다음과 같은 結果를 얻었다.

1. Occlusion index는 正常인 40耳에서는 33.10±1.68 dB, 慢性中耳炎患者 20耳에서는 3.10±0.69 dB, 感覺神經性難聽患者 14耳에서는 28.10±4.05 dB이었다.

2. 正常인 40耳의 static compliance는 범위가 0.22~0.75 cc, 平均値가 0.64 cc, S.D가 0.05이었다.

3. Static compliance가 작을수록 occlusion index는 컸고, static compliance가 클수록 occlusion index는 작아지는 影響을 나타내고 있음을 觀察하였다.

21. 최근 4년간에 시행한 중이수술 970예에 대한 임상적 고찰

연세의대

김기령 · 김영명 · 박인용 · 홍원표
정태인 · 조경열 · 이춘근

이미 100여년 전인 1873년에 Schwartze(1873)에 의

15. Microsurgery of the Laryngeal Lesions

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The microsurgery of the laryngeal lesions was introduced by Kleinsasser in 1965. This has been utilized for diagnosis, surgical management and education of the laryngeal lesions. The laryngomicroscopic technique appear to be useful in the evaluation of the lesions which may be precancerous, since minor degrees of epithelial thickening and alterations in the pattern of fine vessels beneath the epithelium may be observed with greater clarity.

With suspension laryngoscopy, the authors experienced 11 cases of the laryngeal lesions: polyp (4 cases), web formation between false cords and true cords (1), papilloma (4), laryngeal trauma (1), and squamous cell carcinoma (1).

16. Familial Hearing Loss

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There are over 60 types of hereditary hearing loss in man and most of these are distinguished from one another by outstanding associated defects caused the same gene producing hearing loss. However, there are at least 12 types of hereditary hearing loss with no known associated defects. The authors experienced two similar families with hereditary hearing loss.

The affected offsprings had inevitably an affected parent in type of transmissin (autosomal dominant) and the hearing loss developed in childhood and progressed. Audiologic findings showed bilateral symmetrical moderate to severe sensori-neural hearing loss with flattened configuration on the

audiogram. Vestibular function tests showed no abnormal findings and the associated anomalies could't be observed in history or clinical examinations.

17. A Case of Adenoid Cystic Carcinoma of the Middle Ear

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Adenoid cystic carcinoma that is found most frequently in the salivary gland is an extremely rare malignant tumor in middle ear and is a slow growing one with a characteristic tendency of recurrence after surgical removal. Recently we experienced a case of this rare adenoid cystic carcinoma arising from middle ear of 30 year old male patient. The patient complained of hearing impairment for two years duration and pulsating tinnitus for two weeks duration in the right ear. Under the local anesthesia, through exploratory tympanotomy, a grayish white mass measuring 1.5×1.5×1mm in the hypotympanic portion was removed out and a adenoid cystic carcinoma was verified histopathologically.

18. 3 Cases of Waardenburgs Syndrome

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Waardenburgs syndrome is estimated to account for 1 to 7% of all congenital deafness. The primary features of the syndrome include lateral displacement of the medial canthi and lacrimal punctae, a flat

nasal root, white forelock, unilateral or bilateral congenital deafness, some degree of heterochromia of the iris, and hyperplasia of the eyebrow.

This syndrome was described at first by Waardenburg in 1951, and since that time there have been reports of the same syndrome in both the English (Partington, 1953) and American (Di George) literature.

The authors have experienced 3 cases of Waardenburg's syndrome, and report these cases with literature review.

19. A Study on the Causes of Child Hearing Loss Under Age 5 in Korea

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This paper was designed to investigate the causes of child hearing loss which is essential to language development especially in young childhood.

Accurate history taking, precise otolaryngological examination and impedance audiometry were performed on 185 hard of hearing children under age 5 during last 8 months and investigated its causes.

The results were as follows:

1. Infections diseases were the most common causes of sensorineural hearing loss (44/123 cases, 30.1%), and mother's condition during pregnancy the second, and birth injury or head trauma the third.

2. Acute or chronic ear ear diseases were the most common causes of conductive hearing loss (32/62, 51.6%), frequent URI the second (19/62, 30.6%), and adenoid vegetation the third (7/62, 11.3%).

3. The majority of causes of child hearing loss under age 5 was exogenous (179/185 cases, 96.8%), and the minority was endogenous (6/185 cases, 3.2%).

20. Studies on Audiological Significance of the Bing Test

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In normal auditory systems, the difference between the bone conduction thresholds obtained with the test ear opened and occluded in low frequency signals below 1,000Hz is about 10~25 dB. But no difference in these thresholds suggests the presence of a lesion in the conductive mechanism.

Sullivan added the differences in these thresholds at 250Hz, 500Hz and 1,000Hz and called "Occlusion Index".

In order to study the audiological significance, we have measured the occlusion index in 40 ears of normal persons, 20 ears of chronic otitis media patients and 20 ears of sensori-neural hearing impairment patients. We also measured the static compliance in 40 normal ears and observed the correlation with the occlusion index.

The results are as follows:

1. Occlusion index was 33.10 ± 10.63 dB in normal group, 3.10 ± 3.03 dB in chronic otitis media group, 28.10 ± 15.17 dB in sensori-neural group.

2. Static compliance in normal group was 0.61 ± 0.31 cc (0.22~1.75cc)

3. Occlusion index showed inverse proportion to static compliance.

21. Clinical Analysis for 970 Cases of Middle Ear Surgery

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Since the first success of simple mastoidectomy,