

"CONGENITAL MALFORMATIONS OF THE OSSICULAR CHAIN: A REVIEW OF 62 CASES"

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Congenital conductive hearing loss in most cases is due to anomalies of the external and middle ear that usually occur together to a variable extent because of their closely related embryologic development; less frequently, congenital conductive hearing loss is caused by an isolated malformation of the ossicular chain. Congenital ossicular anomalies include abnormalities of either one isolated ossicle or a combination of more than one. Among the different classifications proposed in the literature, the most useful is that proposed by Teunissen and Cremers. These authors distinguish 4 classes of ossicular chain malformations: Class 1: congenital isolated stapes ankylosis; Class 2: congenital stapes ankylosis in combination with an associated anomaly of the incus or malleus. Class 3: include ears with mobile stapes footplate. Anomalies comprising deformed malleus, incus or stapes suprastructure which causes discontinuity of the ossicular chain are classified as class 3A, while anomalies involving fixation of the incus or malleus but a mobile stapes are classified as class 3B. Class 4: aplasia or severe dysplasia of the oval window or round window.

In this study we have retrospectively evaluated 62 cases of isolated ossicular malformations (52 patients) surgically treated in our Department between 1990 and 2005 in order to report on clinical and surgical findings and hearing results. There were 28 males and 24 females; patients ages ranged from 9 to 43 years and the mean age was 23 years.

According to Teunissen and Cremers classification system, 14 cases (11 patients) were classified as class 1, suffering from congenital stapes ankylosis; in all these cases a small fenestra stapedotomy was performed. Fifteen cases (12 patients) were classified as class 2, being patients with congenitally ankylosed footplate in association with malformation of incus and stapedia suprastructure (6 cases), malleus fixation (5 cases), incus fixation (4 cases). Twenty-five cases presented a mobile footplate with an associated malformation of the ossicles and thus were classified as class 3; among these cases, there were 13 cases with an ossicular chain discontinuity and 12 cases with malleus or incus fixation. Finally, in 8 cases a complete or partial absence of the oval window with a malposition of the facial nerve was present. Clinical presentation, role of high resolution computed tomography in diagnosis, intraoperative findings and surgical strategies as well as hearing results will be discussed in detail.