

## CONGENITAL ABSENCE OF OVAL WINDOW

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Oval window agenesis with obvious craniofacial ear deformity is a rarely diagnosed condition. This finding is noticeable nowadays as a result of the development of microsurgery of the ear.<sup>1</sup>

The vestibule is the central chamber of the bony labyrinth of the inner ear. It has a lateral opening at the medial wall of the middle ear, which is called the oval window (fenestra ovale), and lies above and slightly behind the promontory, and below the horizontal portion of the facial nerve. It is closed in life by the footplate of the stapes and its annular ligament. It has an area of approximately  $3 \times 1.4 \text{ mm}^2$  and a depth of 2.5-3 mm.<sup>2</sup>

The stapes has a double origin. The main structures derive from the second branchial arch, but the inner (vestibular) aspect of the footplate differentiates from the otic capsule itself. This area is termed the "lamina stapediale." As development proceeds, the cells immediately adjacent to this lamina normally form the annular ligament, and the lamina becomes incorporated into the footplate of the stapes. Teratogenic agents might affect any stage of the process, producing anomalies including congenital absence of the oval window and others.<sup>3</sup> The oval window is indispensable for the transmission of sound waves into the inner ear (cochlea). In the case of oval window occlusion or absence, the stapes is not normally developed and may be rudimentary, hanging above an undifferentiated oval window niche, causing conductive loss of hearing. The adjacent lateral wall of the bony canal of the facial nerve is also developed from the second branchial arch.<sup>4,5</sup>

The facial nerve may be exposed or displaced downwards. Congenital facial nerve dehiscence of the bony facial canal in the oval window region, a very common finding at tympanotomy, was present at 31% of normal temporal bones examined by Leonard and Alexander in 1968.<sup>6</sup>

Differentiation of the otocyst and otic capsule to inner ear is developed independently of the middle ear, which explains the presence of agenesis of footplate as a separate

anomaly, and the presence of normal inner ear with deformity of the ossicular chain.<sup>5</sup> If the stapes is totally absent, there may be no depression or other indication of the site of the vestibular window, and the facial nerve may be displaced downwards.<sup>7</sup>

### Case Report

A 24-year-old Jordanian female presented to the Otorhinolaryngology Department at King Hussein Medical Centre, Amman, Jordan, in 1995, with a history of bilateral loss of hearing since childhood. There was no positive family history of hearing loss or history of trauma. She was a good lip reader. ENT examination revealed normal tympanic membranes with outstanding small auricles (microtia). Pure tone audiogram showed bilateral, almost symmetrical conductive loss of hearing of 45 dB (Figure 1). Cochlear function and caloric test were normal. Tympanogram showed normal middle ear pressure, and compliance was within the wide normal variation range.

After the condition was explained to the patient, she was asked to decide between having a hearing aid fitted or treatment by surgical intervention via exploration of the middle ear. The patient preferred the latter, accepting any likely risk or complication of this type of surgery as explained to her.

Exploration of the right ear was performed in April 1995, following full radiological investigations, including CT scan of the temporal bones, which were reported to be normal (Figure 2). Operative findings showed rudimentary, almost absent stapes hanging freely above a shallow



FIGURE 1. Preoperative conductive loss of hearing (45 dB).

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FIGURE 2. CT scan of the temporal bones showed them to be normal.



FIGURE 3. Postoperative restoration of hearing giving 40 dB.

depression area. Above and slightly behind the promontory, this shallow area was supposed to be the oval window. The bony canal of the facial nerve was found to be dehiscant, and slightly displaced downwards, but still above the shallow depression. The round window and other two ossicles were found to be normal.

A small hole was created using a bur (0.7 mm) over the area which was supposed to be the oval window, and a teflon piston (0.6 mm x 4.5 mm) was inserted in the hole and hung on the long process of the incus, followed by the application of periosteal graft around the piston at the oval window area.

The patient's hearing improved clinically two weeks following surgery, and audiotically six weeks later, with a gain of 40 dB (Figure 3). She still has the same level of improved hearing, after three years of follow-up.

The surgical treatment of choice should be to create a vestibular fenestra, accompanied with periosteal graft and teflon piston placement. This reconstructive surgery seems to give a good possibility of restoration of hearing. In such cases, the surgeon should be aware of the possibility of facial nerve abnormality (dehiscence of the bony canal, or displacement of its course) during surgery. No complication has yet been reported in this case.

### Discussion

Absence of the oval window as well as stapes is a very rare condition. A review of the literature showed only 57 cases, the first of which was reported in 1958 by Hough.<sup>2</sup> Our report is an addition to the current total.

Jahrodeorfer<sup>8</sup> has suggested that the oval window absence is related to the abnormal development of the facial nerve, while Lambert<sup>9</sup> stated that it is related to the anterior displacement of the facial nerve between stapes and otic capsule, preventing the free development of the oval window and lamina stapedialis. Sterkers<sup>10</sup> has also suggested the morphodysplasia of autosomal recessive transmission, which occurs during the development of the second branchial arch and ossification of the otic capsule. However, a congenital deformity should be suspected in patients with an intact, normal-appearing tympanic membrane which is accompanied by a history of conductive hearing loss. Once surgery is decided upon, then vestibulotomy fascial graft and piston prosthesis should be the choice.

In this case, we preferred to use periosteal homograft because it is available from the same surgical field (endaural) and easy to apply, resulting in good sealing with little risk of fistula formation later, preventing sensorineural hearing loss. Hough<sup>2</sup> used perichondrium, Scheer<sup>11</sup> and Kamal and Palkar<sup>1</sup> used original vein graft, and McGee used fat to seal the oval window.<sup>12</sup>

Hough has suggested loss of hearing since birth greater than the usual as indication that the middle ear malformation is present. If other anomalies in the branchial arches are present, a unilateral conductive hearing loss is more likely to be of congenital origin of the middle ear. Unilateral or bilateral cases of congenital oval window absence are not exactly clear in all reported cases.

Only 39 out of the 57 reported cases in the literature were operated upon, and almost all had good postoperative hearing restoration. Some authors decide not to perform cochleostomy either because of the young age of most of the patients, or the fact that some patients prefer hearing aids to surgery. Advanced ear microsurgery can play a major role in correcting such conditions.

Every case should be evaluated and discussed with the patient and his/her relatives, who should be given the choice of whether or not to proceed with treatment with the use of hearing aids or by undergoing surgery.<sup>13</sup>

In children, surgery should be limited to cases with bilateral deformation. In adults, surgery may be performed when the patient accepts surgery after being informed of all possible risks, such as fistula formation with vertigo, and sensorineural hearing loss. Other possible risks include facial nerve affection, surgical trauma to the inner ear, infection of the middle or inner ear, postoperative granuloma (early complication) and others. Some of the complications which were mentioned in the reported operated cases in the literature were temporary facial paralysis, and over time, loss of much of the initial hearing gain.

CT scan can be of value in the evaluation of the oval window absence, but exploration of the ear is the basis of the diagnosis and treatment, using placement of periosteal graft and teflon piston, which is a simple and safe procedure providing valuable hearing restoration.

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