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Vestibulotomy With Ossiculoplasty Versus Round Window Vibroplasty Procedure in Children With Oval Window Aplasia

*Liliana Colletti, *Marco Mandalà, †Giacomo Colletti, and *Vittorio Colletti

**ENT Department, University of Verona, Verona; and †Department of Maxillofacial Surgery, University of Milan, San Paolo Hospital, Milan, Italy*

Objective: To review the surgical procedures and outcomes in children with bilateral oval window aplasia (OWA).
Study Design: Retrospective cohort review.
Setting: Tertiary referral center.
Patients: Children suffering from OWA between 1990 and 2010.
Intervention: Vestibulotomy with ossiculoplasty (V-OPL) or round window vibroplasty (RWV).
Main Outcome Measures: Findings at radiology and surgery, preoperative and postoperative bone conduction (BC), air conduction (AC), and RWV-air conduction (RWV-AC) thresholds and speech discrimination scores (SDSs).
Results: Among 23 children, 11 underwent V-OPL and 8 RWV. Four children in the V-OPL group had aborted surgery and were excluded from the study. In all the remaining 19 children, the 6-month follow-up time showed postoperative AC and SDS values significantly better than the preoperative thresholds in both groups. At the 36-month long-term follow-up, AC and SDS were stable in the RWV group but showed a significant worsening in the V-OPL children compared with the 6-month follow-up results. Preoperative versus postoperative BC values showed a significant difference between the 2 groups at 36 months; 5 of the V-OPL group underwent revision following the same surgical principles, which did not result in improved outcome.
Conclusion: In children with OWA, V-OPL provides modest long-term results and carries higher risks of BC degradation compared to RWV. Both procedures are technically challenging but considering the respective hearing results and morbidity of primary and revision surgery, we have abandoned the V-OPL procedure in favor of RWV. In infants and children younger than 5 years with OWA previously not considered candidates for hearing restoration, we consider RWV as the first-choice surgery. It has shown to provide significantly better hearing outcomes than traditional atresia surgery with minimal complication rate.
Key Words: Aural atresia—Fenestration—Ossiculoplasty—Round window vibroplasty—Severe oval window malformation.

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Congenital absence of the oval window (OW) is an uncommon embryologic defect (1). Oval window aplasia (OWA) involves failure of the otic capsule bone to open into the vestibule either because the footplate fails to develop or because the developing footplate does not fuse with the primitive vestibule (2). Typically, there is no footplate or annular ligament, and the OW area is characterized by a complete osseous obliteration, either as a thick bony plate or as a concentric osseous narrowing (1). Deriving from the first branchial arch structures, in the setting of an aural atresia, the lateral ossicular chain may be normal, abnormal as the single stapes crus, dysmorphic or fused malleus and incus, or absent. The associated hearing loss may be conductive or mixed and varies from mild to severe.

With otoscopy, OWA is often not readily apparent and most children present after failing either a newborn or school hearing screening. From a surgical perspective, children with minor malformations of the middle ear (Jardorfer scale >7) (3) are considered good candidates for ossiculoplasty (OPL) surgery to rehabilitate hearing. Conversely, malformations of the oval and round window, deriving from underdevelopment of the second branchial arch, often include facial nerve abnormalities and are generally considered poor candidates for surgery. Attempts at surgical repair have met with mixed and conflicting outcomes in the very small series described in the literature (4–6).

Address correspondence and reprint requests to Vittorio Colletti, M.D., Piazzale L. A. Scuro, 10, 37134 Verona, Italy; E-mail: vittoriocolletti@yahoo.com

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Recently, the use of active middle ear implants (AMEIs) has been expanded from sensorineural to conductive and mixed hearing losses (7) with the aim of offering a better hearing outcome to patients with severe ossicular chain defects than that obtained with OPL. Furthermore, AMEIs provide better cosmesis, easier maintenance, and a lower risk of local inflammation than provided by a bone-anchored hearing aid (BAHA) in these patients (8).

Since the new approaches of coupling the actuator to the round window membrane or to any residual middle ear ossicle, also called vibroplasty, have been clinically proven in adult patients with severe mixed hearing loss (9), the use of AMEIs (10) was expanded to infants and children with congenital aural atresia (CAA).

Currently, no data on the application of AMEIs in children with OWA have been presented in the literature, and the purpose of this study was to compare the feasibility, outcome, and complications of round window vibroplasty (RWV) to vestibulotomy with ossiculoplasty (V-OPL) to assess which technique offers the best option for long-term restoration of hearing.

MATERIALS AND METHODS

A retrospective review of the radiologic evaluation, surgical management, and audiometric outcomes of children with severe OWA who underwent surgery by the senior author (V. Colletti) at the University of Verona and elsewhere for the last 20 years identified 23 children. From 1990 to 2004, 15 children underwent V-OPL (Group A), and from 2005 to 2010, 8 children underwent RWV (Group B). Surgery was aborted in 4 children in Group A, and these children were fitted with conventional hearing aids and excluded from analysis of the hearing results. During the same period, 2 other OWA subjects were fitted with conventional hearing aids because their parents refused the surgical option. These 6 subjects and most of the children presented in this series had a previous unsatisfactory experience with hearing aids. No children in the present series opted for BAHA as a result of parental refusal.

The Verona University Ethics Board approved the study, and all patients gave their informed consent.

Preoperative and Postoperative Measurements

The following preoperative and postoperative measurements at 0.5, 1, 2, and 4 kHz were collected at 6 and 36 months of follow-up in 15 children at our institution and in 6 children from outside our institution: 1) bone conduction (BC) and air conduction (AC) thresholds; 2) air-bone gap (ABG) as the difference between preoperative AC and BC thresholds and between postoperative AC and aided AC and BC thresholds; 3) percentage of speech discrimination scores (SDSs) obtained with bisyllabic words correctly repeated at 65 dB hearing level (HL) in the Italian language available in the 15 children followed at our institution; and 4) postoperative complications and floating mass transducer and V-OPL displacement or extrusion rate of the prosthesis.

Surgery and Devices

V-OPL Procedure

A transcanal approach was used for the V-OPL procedure in this cohort of children. After elevating the tympanomeatal flap, the middle ear was entered and the ossicular chain was identified and evaluated for anatomy and mobility. The relationship between the facial nerve and the ossicular chain/stapes remnant was clearly assessed.

Fenestration into the vestibule was created using a low-speed drill starting with a 3-mm diamond burr in the area corresponding to the normal stapes position in a slight depression, just behind the Jacobson nerve and inferior to the tympanic facial nerve. Drilling of the otic capsule bone continued to the endosteum of the vestibule using a 2-mm and then a 0.7-mm burr. The opening of the fenestration into the vestibule was closed with a vein or fascia graft to avoid contaminating the membranous labyrinth with blood. The distance from the neck of the malleus or incus to the location of the vestibulotomy was measured, and the prostheses were modified to the appropriate length. The fenestrated window measured approximately 0.7 to 0.9 mm to accommodate either a personally constructed malleus to footplate prosthesis (11) to be connected to the neck of the malleus (6 children; Fig. 1) or a classic wire-piston-Teflon stapes prosthesis for the long process of the incus, although dysmorphic and without the lenticular process (5 children). The prosthesis was carefully placed over the vein or temporalis fascia down into the new OW fenestra, and the drum was returned to its normal position (round window reflex verified). The graft was large enough to completely cover the fenestration and prevent its migration into the vestibule.

RWV Procedure

For RWV, the Vibrant SoundBridge (VSB) (manufactured by MED-EL Hearing Technology, Innsbruck, Austria) was used. Device characteristics, surgical principles, and device activation have been described previously (9,10,12,13) (Fig. 2).

Statistical comparisons between preoperative and postoperative outcome measurements at 6- and 36-month follow-up were conducted by paired Student's *t* test, Wilcoxon-Mann-Whitney test, or Fisher's exact test as appropriate (significance: $p < 0.05$).

RESULTS

The mean ± standard deviation age of the whole population was 7.4 ± 4.7 years, with a male/female ratio of 1.3. Demographic and clinical data for the 2 subgroups in the study population are shown in Table 1.

For the 15 V-OPL children followed up at our institution, computed tomographic scans were available for review for only 7 children operated on during the period of 1990 to 2004; for the remaining 8 children followed up at our institution or elsewhere, we used only the radiologic reports. Computed tomographic scans and radiologic reports were available for all RWV subjects.

The scans showed that the OW was absent in all ears (all surgically confirmed) with concomitant ossicular anomalies and an aberrant course of the facial nerve. The main anatomic abnormalities observed at surgery are included in Table 2. Surprisingly, the round window membrane was present in all patients.

Ossicular and facial abnormalities are included in Table 2. A correlation was demonstrated between computed tomographic reports and surgical findings. This was despite the fact that, in 2 children in Group A and 1 in Group B, the radiologic reports were misleading, overestimating the angle of the second genu of the facial nerve

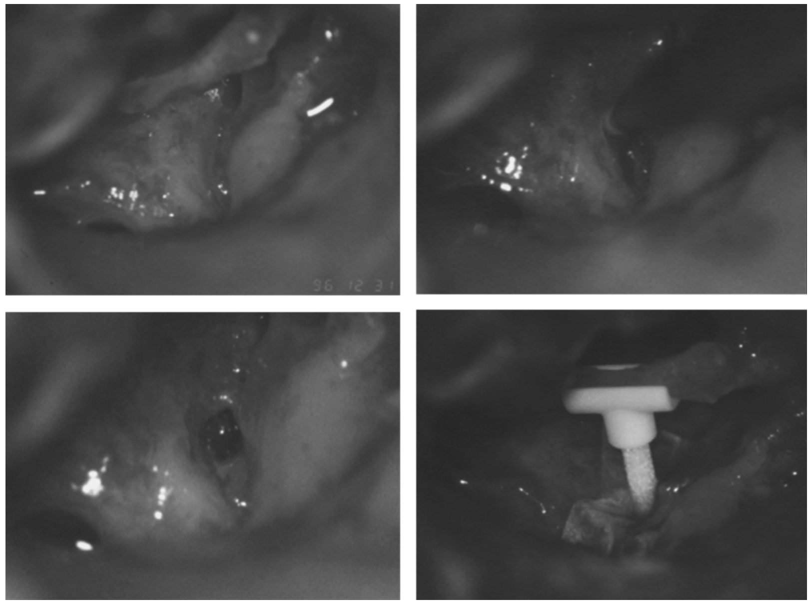

FIG. 1. Surgical view of a vestibulotomy with ossiculoplasty procedure.

and not accurately describing its aberrant course in the mastoid portion.

In the 4 patients in Group A, where surgery was aborted, the radiologic reports were misleading underestimating the dimensions of the stapedial artery overlying the OW in 1 child and poorly describing the entity of the dehiscent facial nerve covering the OW area in 3 children.

The radiologic reports did not concur with the surgical findings in 30% (7/23) of patients.

Audiologic Outcomes

The means and standard deviations of the outcomes of all of the test procedures for both groups preoperatively and at 6- and 36-month follow-up times are reported in Table 3. In both groups (A and B), there was a statistically

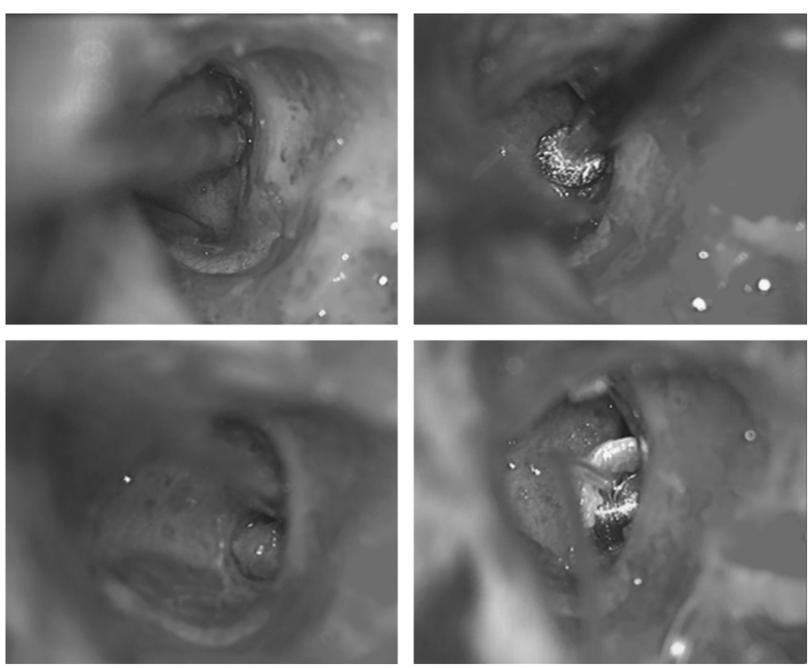

FIG. 2. Surgical view of a round window vibroplasty procedure.

TABLE 1. *Demographics of the patient population with severe OWA who were submitted to surgery*

	V-OPL group (15 patients)	RWV group (8 patients)	*p*
Age, mean ± SD, yr	7.9 ± 4.3	6.8 ± 5.5	>0.05
Male:female	9:6	4:4	>0.05
Bilateral:unilateral	9:6	5:3	>0.05
Aborted surgery	4	—	
Revision surgery	5	—	
Syndromic patients	2 Goldenhar 1 Hemifacial microsomia	1 Goldenhar 1 Hemifacial microsomia	

OWA indicates oval window aplasia; RWV, round window vibroplasty; SD, standard deviation; V-OPL, vestibulotomy with ossiculoplasty.

significant improvement in hearing when comparing the preoperative average AC and SDS to the 6-month follow-up ($p = 0.0234$ and $p < 0.0001$, respectively).

A statistically significant deterioration in average AC and SDS was observed in the V-OPL group when comparing the 36-month follow-up to the 6-month follow-up ($p = 0.0104$).

No statistically significant differences were identified when comparing the postoperative AC thresholds in the RWV patients at 6- and 36-month follow-ups ($p > 0.05$).

There was no statistically significant difference in BC among the average preoperative and 6- and 36-month postoperative follow-up values in the RWV group alone ($p > 0.05$). The V-OPL group showed a significant deterioration of BC from the preoperative threshold to the last follow-up ($p = 0.0025$).

The mean ABG worsened significantly in the V-OPL subjects between the 6- and 36-month follow-ups from 13.9 ± 13.1 to 19.2 ± 19.1 dB HL ($p = 0.0315$). No significant difference was observed for RWV subjects at the 2 follow-up times ($p > 0.05$).

Postoperative AC thresholds at 0.5, 1, 2, and 4 kHz evaluated at the 6- and 36-month follow-ups for both groups (V-OPL and RWV) are shown in Figure 3. The final mean magnitude of hearing improvement from preoperative AC threshold to the 36-month follow-up was 1.9 ± 27 and 38.9 ± 13.8 dB HL in the V-OPL and RWV groups ($p < 0.0001$), respectively, with only 4 subjects in the V-OPL group compared to all the subjects in the RWV group having at least a 10-dB HL improvement in AC hearing threshold from preoperatively to the last follow-up.

TABLE 2. *Main anatomic abnormalities observed at surgery*

Findings	V-OPL group (15 patients)	RWV group (8 patients)
Malleus fused to incus	3	3
Malleus fused to the anterior or medial attic wall	4	3
Incus with distorted and with a short long process	7	4
Incus without lenticular process	4	3
No stapes superstructure, no footplate, no annular ligaments, and OW obliterated by a thick bony plate	13	8
OW overlain by artery	1	1
VII cranial nerve dehiscent and overlying the oval window	3	2
VII cranial nerve inferiorly and anteriorly displaced limiting the view of the round window	1	1

OW indicates oval window; OWA, oval window aplasia; RWV, round window vibroplasty; V-OPL, vestibulotomy with ossiculoplasty.

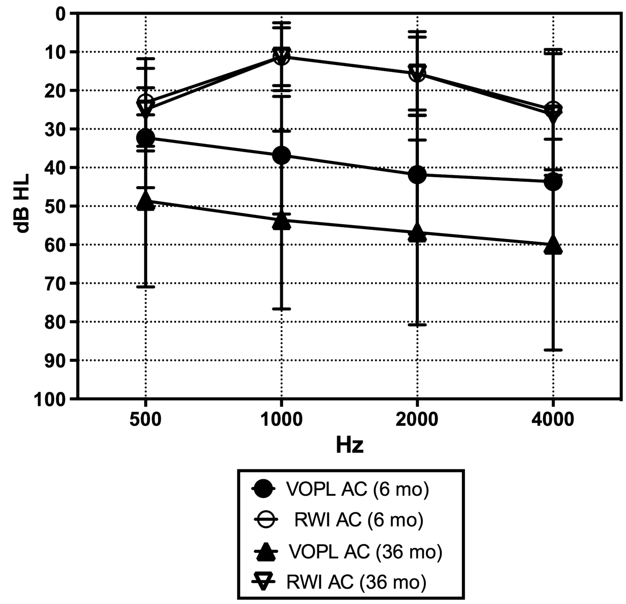

FIG. 3. Air conduction thresholds at 0.5, 1, 2, and 4 kHz evaluated at 6- and 36-month follow-ups for vestibulotomy with ossiculoplasty and round window vibroplasty subjects.

Statistically significant differences were observed when comparing the SDS outcomes between the 2 groups with the RWV subjects consistently reaching values above 80% of speech intelligibility ($p < 0.01$).

Revision Surgery

Reasons for Revision Surgery

The decision for a revision procedure was taken at least 36 months after surgery on the basis of the outcome of the BC, AC, and SDS tests that showed a significant deterioration of hearing compared with the preoperative and short-term values. Analysis of the revision cases for V-OPL performed using the same procedure showed a further poor outcome in all patients, with no significant ($p > 0.05$) gain in hearing thresholds after revision. After 2005, all of the revisions were performed with RWV. Patients who had revision surgery with RWV were excluded from this study. A detailed description of the outcomes of all patients who received a revision procedure will be presented in a subsequent article.

Reasons for Aborted Surgery

One patient had an OW area overlain by a large stapedial artery (Fig. 4) and 3 patients had a dehiscent facial

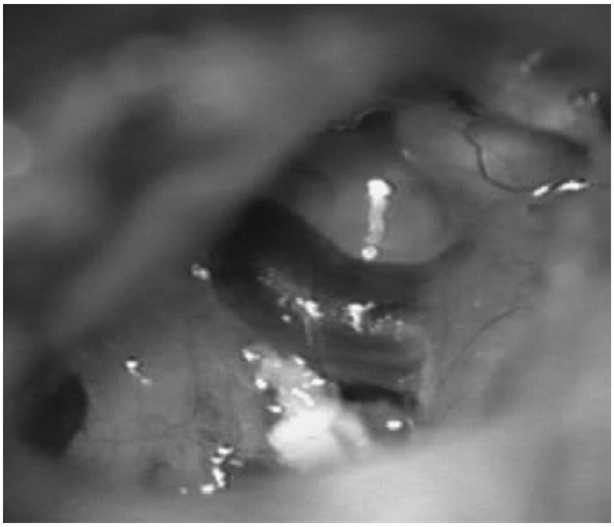

FIG. 4. Oval window area overlain by a large stapedial artery (aborted vestibulotomy with ossiculoplasty).

nerve overlying the OW area. The experience acquired so far in this special population suggests that surgery might have been avoided if the preoperative radiologic analysis could have focused specifically on the anatomic location of the facial nerve and on the presence of any vascular structure overlying the OW area.

Complications

Three minor (1 facial paresis lasting 6 months, 1 prosthesis extrusion, and 1 tympanic membrane perforation) and 1 major complication (sudden deafness after an acute otitis media) were all observed in Group A.

DISCUSSION

In recent years, depending on the anatomic complexity encountered in OWA, different modalities of V-OPL have been proposed for hearing restoration. Briefly, after a vestibulotomy in the round window area, a classic OPL was performed with a prosthesis connecting either the residual incus, malleus, or chorda tympani (incudostapedotomy, malleostapedotomy, or chordostapedotomy) to the vestibule (6). When an inferiorly displaced facial nerve was obscuring the OW area, a fenestration of the horizontal semicircular canal or again a vestibulotomy was performed either above or below the displaced facial nerve (14).

For many patients in these series, the long-term hearing results were mixed, with very few patients obtaining an ABG closure to within 20 dB and most to within 30 dB or more. Furthermore, the good initial hearing results were followed by a long-term deterioration (1,3–6), suggesting that the results from V-OPLs did not justify the risks involved for the minimal gains in hearing. In addition, since revision surgery was rarely successful, most of these patients were then recommended conventional hearing amplification or osseointegrated auditory implants (e.g., BAHA, Cochlear Corp., Englewood, CO, USA) for an improvement in their hearing.

Recently, it has been reported that unilateral CAA and hearing loss can lead to speech development and learning difficulties and can be detrimental to the academic success of children (15,16).

At the time of the present surgical experience on children with OWA, alternative options for these subjects were limited to conventional hearing aids, BAHA Softband, and traditional BAHA in children older than 5 years. However, in this context, our personal experience indicated that a large proportion of these children were unable to tolerate the BAHA Softband and were unwilling to be fitted with BAHAs despite very good outcomes reported in CAA (17,18). Significant difficulties in acceptance by children and parents were attributed to their limited output level and bandwidth, their poor cosmetic appeal, the need for patient feedback for optimal fitting, and the impossibility of treating the sensorineural component of hearing loss that can be present in these subjects (19–21). Furthermore, children younger than 5 years were not recommended to be implanted with BAHA because of problems with the thickness of the temporal bone and osseointegration (17). To the best of our knowledge, no report of children affected by OWA fitted with BAHA has been described in the literature. At the same time, conventional hearing aids were not providing significant benefit in these patients owing to the large ABG.

For all these reasons, we therefore looked for a surgical procedure that could provide good long-term hearing results in OWA patients and retrospectively compared the outcome from 2 groups of children undergoing either V-OPLor RWV procedures.

The RWV group showed significant improvements in AC hearing tests compared with the V-OPL group, both at short- and long-term analyses. Furthermore, in the V-OPL group, the initial improvement in hearing at the 6-month follow-up was short-lived, and the deterioration of all hearing gains was highly statistically significant in the long-term analysis. Conversely, the RWV group maintained a substantially similar level of improvement over time (Table 3 and Fig. 3).

The major limitations of the present study are the low number of subjects enrolled and the great variability of procedures adopted and outcomes in the V-OPL group. Both of these limitations could not be overcome since a great variability in the results of traditional functional surgery for OWA and aural atresia is an intrinsic factor in the literature and this report presents the largest number of subjects with OWA in the literature.

The different concepts and approaches that characterize the 2 procedures shed light as to why hearing outcomes with V-OPL are inferior to RWV and why the good short-term outcomes of V-OPL significantly deteriorate over time. The outer and middle ear functions are bypassed with RWV and mechanical energy is delivered directly to the inner ear by the actuator coupled to the round window membrane, making it possible to compensate simultaneously for the conductive and sensorineural

TABLE 3. *Outcomes of primary surgery: means and standard deviations of the hearing thresholds observed for all subjects before surgery and at the 6- and 36-month follow-ups*

	BC		AC		ABG		SDS	
	V-OPL	RWV	V-OPL	RWV	V-OPL	RWV	V-OPL	RWV
Before surgery	13.9 ± 6.1	14.6 ± 6	56.7 ± 8.4	58.3 ± 6.5	42.8 ± 8.8	43.8 ± 9.7	14.5 ± 11.3	12.5 ± 10.4
p	>0.5		>0.5		>0.5		>0.5	
6-mo follow-up	24.6 ± 18.6	16.4 ± 6.4	38.6 ± 17.4	18.8 ± 9.9	13.9 ± 13.1	2.4 ± 10.8	45.4 ± 29.8	83.8 ± 10.6
p	>0.05		0.0103		0.0289		0.003	
36-mo follow-up	35.6 ± 16.1	17.6 ± 4.6	54.8 ± 23.9	19.5 ± 10.7	19.2 ± 19.1	1.9 ± 9.5	29.1 ± 25.9	87.5 ± 10.3
p	0.0043		0.0012		0.0315		<0.0001	

ABG indicates air-bone gap between BC and AC thresholds (dB HL); AC, air conduction threshold (dB HL); BC, bone conduction threshold (dB HL); RWV, round window vibroplasty; SDS, percentage of bisyllabic words at 65 dB HL in the Italian language; V-OPL, vestibulotomy with ossiculoplasty.

component, as the comparison in the amount of ABG gains between the 2 procedures shows. In addition, with the obliteration of the middle ear space with soft tissue, the risk of extrusion or dislodgement of the floating mass transducer is reduced as demonstrated by the consistent long-term results with the RWV approach.

Conversely, to be successful, V-OPL requires a precise and stable coupling of all the interconnected components of the chain of transmission: from the tympanic membrane to the malleus or incus and to the prosthetic device and finally to the inner ear via the vestibulotomy (22,23). Failure to achieve good coupling even in 1 single step puts the success of the whole procedure at risk.

Probably the most significant biologic ''stumbling block'' in OWA is the new bone growth observed in the drilled area of the vestibulotomy in patients undergoing revision surgery. This bone growth is very similar to that observed in the bony ear canals of children with CAA submitted to atresioplasty surgery and is similarly responsible for significant progressive hearing loss in terms of increased ABG up to 50 to 60 dB HL from the 6-month audiogram.

A review of the intraoperative picture at revision surgery showed that, in the drilled ''OW'' area, new bone had regenerated under the prosthesis, progressively restricting the vestibulotomy and pushing the prosthetic device upward. This resulted in a disconnection of the prosthetic device from the long process of the incus or from the neck malleus. This implies that the otic capsule may regenerate new bone matrix either under the mechanical stress of the drilling or from the dust produced by the drilling. If these assumptions are correct, it might be wise to limit the V-OPL approach to elderly children because the rate of bone growth seems to parallel the body's overall rapid bone growth in children.

These findings at revision surgery did not indicate a favorable long-term outcome for V-OPL and motivated a change in our surgical strategy. We bypassed the middle ear and used 1 single physiological point of entry to the inner ear represented by the round window membrane. The outcomes of the present study seem to corroborate this new approach.

Patients with OWA present unique and challenging anatomic features so, before undergoing surgery, differences in the possible surgical options for hearing rehabilitation in children with OWA must be discussed and elucidated with patients and family. For V-OPL, the family must be made aware that:

1. given the difficult surgical anatomy, and in light of the finding of misleading radiologic reports in approximately 30% of our patients, aborting the case rather than risking sensorineural hearing loss or facial nerve injury is prudent (4,24–26);
2. the improved hearing may significantly deteriorate over time; and
3. if initial surgery does not improve hearing to serviceable levels, revision surgery is not recommended because it is rarely successful in restoring hearing.

Similarly, when discussing RWV, the informed consent should indicate that:

1. the procedure is new and the experience limited to very few centers, but
2. so far, the improvement in hearing has been shown to be stable over a long period (7 yr),
3. device failure is very rare (27), and
4. revision surgery for hearing deterioration is an exception but mostly successful with RWV.

In conclusion, in children with OWA, V-OPL provides modest results and carries higher risks of BC degradation compared to RWV. Both procedures are technically challenging, but considering the respective hearing results and the morbidity of primary and revision surgery, we have abandoned the V-OPL approach, and for the last 10 years, our preferred method of hearing reconstruction for infants and children younger than 5 years with severe aural atresia and OWA has been RWV, and this study validates this practice. In children older than 5 years, today we indicate the new nonpercutaneous implantable bone-anchored hearing devices as good alternatives. The new bone conducting hearing devices only need 1 operation, have a very low risk for complication, and a hearing gain similar to other bone conducting hearing aids.

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