Management of Otodysplasia

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Epidemiology and etiology of otodysplasia

Otodysplasia can be defined as any congenital malformation affecting the ear. Otodysplasias are relatively rare, with an incidence of approximately 1/10.000 live newborns and a peak incidence in some specific areas, such as Scandinavia (2.35/10.000) and California (2/10.000)[1,2], and specific ethnic groups, such as the Asian (2/10.000) and the Hispanic one (3.25/10.000)[1]. Unilateral presentation is more common than bilateral, with a 6:1 ratio. In unilateral cases, the right side is more frequently affected (57-67% of cases), especially when the ear malformation is isolated.

Otodysplasias are associated with major comorbidity in 30-40% of cases, the most commonly reported diseases being congenital heart defects and cleft lip/palate (30%), followed by eye malformation (anophthalmia and microphthalmia in 14% of cases), liver and kidney disease (11%) and holoprosencephaly (7%)

The typical association with an abnormal development of branchial-derived organs, such as the mandibular, zygomatic and maxillary bone (10%), can be found in specific genetic syndromes/sequences, the most frequent of which are:

- *Pierre-Robin sequence (1/8500 births)[3]:* micrognathia, glossoptosis and upper airway obstruction.
- *Treacher-Collins syndrome* (1/50.000 births)[4]: also known as mandibulo-facial dysostosis, it is an autosomal dominant disorder caused by mutations of the TCOF-1 gene; its main features include bilateral microtia and aural atresia, zygomatic bone and mandibular hypoplasia, lower eyelid coloboma.
- *Brachio-oto-renal (BOR) syndrome:* autosomal dominant disease caused by a mutation of the EYA1 and SIX1 genes, characterized by variable degree otodysplasia, ranging from I to III degree microtia with aural atresia, and by kidney malformation[5].
- *Hemi-facial microsomia,* also known as Goldenhar syndrome or I-II branchial arch syndrome, consists of facial asymmetry due to hypoplasia of facial bones on one side, unilateral microtia and aural atresia, plus eye, spine and heart malformations[6,7].
- *CHARGE syndrome (0.1–1.2/10,000 live newborns)*: caused by a mutation of the CHD7 gene in 75% of patients, its leading features include eye coloboma, heart malformation, choanal atresia, growth/development retardation, genital hypoplasia, hearing loss. It can be associated with both sensorineural and conductive hearing loss[8].

- *Velocardiofacial syndrome:* caused by 22q11.2 microdeletion, its main traits are cranio-facial malformations (90%), soft palate insufficiency (90%), hypoparathyroidism and hypocalcemia (65%), psychiatric disorders (60%), mental retardation (35%). Hearing loss is present in 40-50% of cases and is mostly of a conductive type, being caused by chronic otitis media, and in rare cases of a sensorineural type[9].
- *Nager syndrome*: mandibulo-facial dysostosis associated with defective development of the radial bone, lower eyelid coloboma, maxillary and mandibular hypoplasia and cleft palate of variable severity[10].

Other than genetic defects, causes of otodysplasia also include pre-natal factors such as trauma (fetal microhemorrhages), placental dysfunction, hypoxia, infectious agents (rubella) and toxic drugs (thalidomide, vitamin A, etc.).

Classification

Abnormalities of development of the external and middle ear present with such variability that it is difficult to provide a simple and exhaustive classification. Over decades, several classifications have been suggested[11-12], as illustrated by **Table 1**. Oldest classifications were based exclusively on the malformation of the pinna, thus distinguishing three grades of microtia:

Table 1. Classifications of otodysplasias

	Marx, 1926	Altmann, 1955	Meurman 1957	Lapchenko, 1967	Gill, 1969	Kruchinsky, 1959	Ombredanne, 1971	Nager, 1973	Colman 1976	Marquet, 1978	Pulec, 1978	ECEAI 2009
Auricle	■		■		■	■	■	■	■			■
External auditory canal		■	■		■		■	■	■	■	■	■
Middle ear		■		■	■		■	■	■	■	■	■
Inner ear								■	■			■

- *I degree microtia:* all structures of the normal auricle are present, with modest deformity such as protruding auricle, macrotia, cryptotia, cleft ear.
- *II degree microtia:* dysmorphic aspect is more evident, because some anatomical parts such as helix and lobule are absent.
- *III degree microtia:* the auricle is represented only by a remnant; this type of microtia can be further divided into “concha” type and “lobule” type.

Other classifications take into account middle ear morphology, whereas other ones include coexisting inner ear malformations.

In the Audiology and Otology Unit of "Bambino Gesù" Pediatric Hospital in Rome, Italy, an original classification, is currently used with the aim to be simple and at the same time functional to surgical treatment; this has been published by Pasquale Marsella[13] and divides otodysplasias in two categories:

Minor otodysplasia, characterized by a *patent external ear canal (including cases in which a tight stenosis may mimics an atresia)* generally includes:

- ✓ 1st or 2nd degree microtia;
- ✓ variable degree of stenosis to apparent atresia of the external auditory canal;
- ✓ malformed or disjointed ossicular chain, with related conductive hearing loss;
- ✓ tympanic cavity and mastoid volume are within normal limits, facial nerve is normal.

Furthermore, based on the severity of malformation of the above mentioned anatomical landmarks, minor otodysplasia can be further classified into **I** and **II** type (**Figure 1**), useful as far as prognosis and functional results after surgery are concerned.

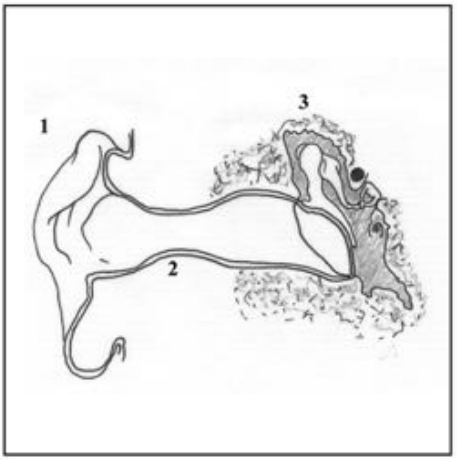


Figure 1. (left) Minor otodysplasia type I: 1st degree microtia, (1), mild stenosis of the external auditory canal (2), slightly malformed ossicular chain (3).

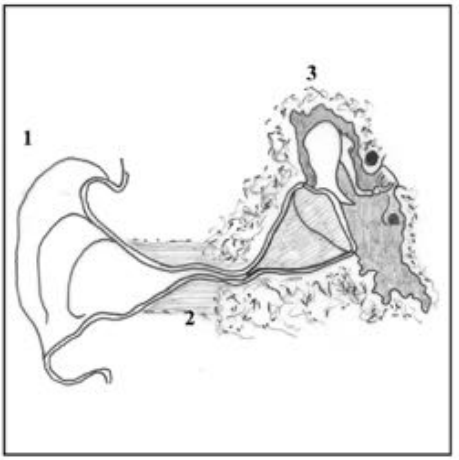


Figure 1. (right) Minor otodysplasia type II: 2nd degree microtia (1); severe stenosis of the external auditory canal (2) dysmorphic ossicular chain (3).

Major otodisplasia or aural atresia, characterized by an *atretic external ear*, presents:

- ✓ 2nd or 3rd degree microtia;
- ✓ fibrous or bony atresia of the external auditory canal;
- ✓ severe malformation of the ossicular chain;
- ✓ abnormal conformation of the tympanic cavity and the mastoid, aberrant facial nerve.

The degree of malformation enables distinction between two types of major otodysplasia (**Figure 2**). In the author's opinion, such classification is important since surgical correction of major otodysplasia type II has a high rate of unsuccessful outcome and complications, that recommend against it.

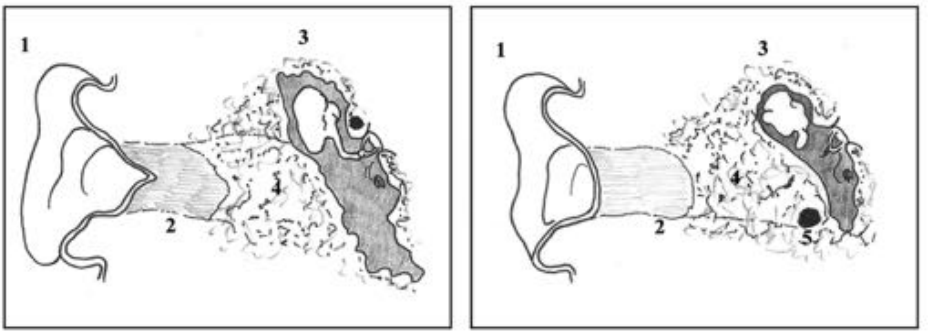


Figure 2. Major otodysplasia type I (left) and II (right). Left: Major otodysplasia type I: 2nd-3rd degree microtia (1); bony aural atresia (2, 4); malformed ossicular chain (3), well-shaped tympanic cavity and normal route of VII nerve (5). Right: Major otodysplasia type II: 3rd degree microtia (1); very thick bony layer causing external auditory canal atresia (2, 4); small tympanic cavity with grossly malformed ossicular chain elements, often fused in one block (3); aberrant VII nerve, sometimes found in the atretic plaque (5).

Embryogenesis and important anatomical landmarks

In order to thoroughly understand the challenges presented by functional surgery of otodysplasia, it is important to review some important anatomical features deriving from the peculiar embryogenetic defects of malformed ears. Such tips allow the clinician to assess the malformation in its complexity and to give a correct indication to surgery, thereby minimizing the risk of intra- and postoperative complications and of a poor functional outcome.

Auricle

External and middle ear have a different embryogenesis form the inner ear; therefore, auricle malformations are seldom associated to inner ear dysplasias. Consequently, in a newborn with a malformed auricle one can expect a conductive hearing loss, whereas a sensorineural hearing loss is seldom detected. In fact, the degree of microtia is in itself a reliable predictor of the severity of the middle ear malformation[11]. Under this respect, micro-otoscopy is the main exam. Sometimes ossicular chain malformation can be found in the presence of a normal auricle, which can be explained by the earlier development of the latter during organogenesis: this condition is generally referred to as *minimal otodysplasia,* its features including incudo-malleolar fusion, incudo-stapedial disjunction, stapes superstructure malformation, stapes agenesis, together with a moderate-to-severe degree conductive hearing loss showing a normal pinna.

External auditory canal

In case of aural atresia, the bony layer of atresia can have variable shape and thickness, depending on the stage of organogenesis at which the involution of the "meatal plug" (i.e. the embryonic tissue obliterating the primitive external auditory canal) stopped: the earlier the interruption of the process, the thicker the bony layer of atresia. In cases of very late interruption, the atresia plaque is merely fibrous tissue.

Mandible

A deficient development of the tympanic bone, originating from the II branchial arch, causes a backward position of the mandible condyle, which is thus in the way of the surgical approach to the tympanic cavity. In these cases, access to the middle ear will necessarily be as near as possible to the temporo-mandibular joint and the canaloplasy more posterior than usual.

Mastoid and facial nerv

The stage of mastoid development influences the VII cranial nerve route. A poor vertical growth of the mastoid bone and its styloid process shortens the mastoid tract of the facial nerve, which becomes "horizontal" and emerges from the mastoid more superiorly than normal. In turn, this causes the facial nerve to run through the tympanic cavity rather than surround it as usual. As a consequence of this, intraoperatively the otologist will find the third tract of the facial nerve covering the round and sometimes even the oval window (**Figure 3**), or bifurcating in a way that one branch will sneak in between the stapes crura.

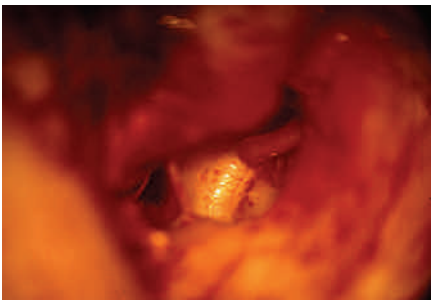


Figure 3. Aberrant facial nerve and agenesia of staper superstructure.

Ossicular chain

The compound embryogenesis of the ossicular chain accounts for the number of possible malformations that may affect it. Deformity of the malleus head and incus body are often associated, owing to their common origin from the I branchial arch, whereas defects of the long arm of the incus are often associated with abnormal stapes superstructure, both elements stemming from the II branchial arch. Instead, despite the frequency of impaired oval window motility, malformations of this element are rare due to its independent embryogenesis.

Management protocol

Considering the common association between otodysplasia and other organ disease, a multidisciplinary approach is always mandatory. Therefore, a newborn with otodysplasia should receive a definite diagnostic protocol aiming to detect comorbidities and at the same time to clarify the priorities of treatment and rehabilitation.

ENT examination and an assessment of hearing are among the most important points: in most cases, the impossibility to perform tympanometry and to obtain otoacoustic emissions makes it necessary to proceed with air- and bone-conduction auditory brainstem responses test, which allow the clinician to evaluate the indication for a hearing aid.

Besides being a very important tool for an appropriate hearing and speech rehabilitation of these subjects, hearing aids can also be a precious support to diagnosis, enabling a better definition of the audiogram in those frequencies that are not reliably investigated by the auditory evoked potentials. Finally, a speech assessment is helpful to investigate the language learning progresses and any eventual cognitive delay.

Importantly, CT scan imaging of the ear finds no indication in the first years of life of these patients, in that it adds no useful information for treatment and/or rehabilitation. The rare conditions in which imaging is mandatory include suspicion of cholesteatoma or neoplasms. In all other cases the authors recommend that the CT scan should be postponed until a surgical plan is defined, such as Baha surgery or functional reconstruction.

Defining the appropriate indication to surgical correction of otodysplasia and its timing may be a challenging task. To date, the elements predicting success and candidacy criteria are not well defined. The only absolute indication to surgery (i.e. a condition in which the operation cannot be postponed) is represented by cholesteatomatous or otherwise complicated otomastoiditis. In all other conditions the opportunity of a functional surgery must be carefully weighed against other simpler functional solutions such as implantable bone-conduction hearing aids.

In fact, several factors are known to carry a relevant prognostic significance:

- **Severity of otodysplasia**: generally, the milder the dysplasia, the higher the likelihood of a good functional outcome. Authors unanimously agree that *minor otodysplasia* type I and type II represents an indication to surgical correction; instead, there is no agreement concerning surgical treatment of *major otodysplasia*. In the latter, Jahrsdoerfer's[14] and Siegert's[15] grading systems, illustrated in **Table 2**, are still considered as a reference, because they bear prognostic significance: a Jahrsdoerfer score > 7 or a Siegert score > 15 relate to a good probability of success. Importantly, even the highest resolution CT scan may fail to correctly score ear malformations. In fact, it allows to detect gross abnormalities of the middle ear, but often misses on minimal defects such as ossicle dysmorphies, ossicular chain discontinuity or oval/round window abnormalities which can be identified only intraoperatively. Under this respect, the above mentioned classification of otodysplasia, proposed by Marsella[13] and used in Bambino Gesù Pediatric Hospital, allows to distinguish between cases in which a surgical correction is feasible (minor otodysplasias and major otodysplasias type I), and those in which it would most probably be unsuccessful.
- **Age**: independently of the degree of dysplasia, a surgical indication must be considered starting from 6-8 years, the same as in all other cases where functional otosurgery is an option. Subjects presenting with a moderate/severe conductive hearing loss may benefit from hearing aids before they become old enough to be considered suitable for a surgical treatment.
- **Bilateral vs unilateral otodysplasia:** in the literature, there is a general agreement that bilateral conductive hearing loss constitutes an indication to functional surgery as long as patient's age and anatomical conditions are favorable. On the other hand, unilateral otodysplasia is seldom an indication.

➢ **Auditory threshold:** it is a widely shared opinion that the finding of a sensorineural component with bone conduction threshold (PTA 0.5-4 kHz) > 30 dB hearing loss (HL) represents a relative contraindication to surgery because of the decreased cochlear reserve.

Table 2. Grading systems by Jahrsdoerfer and Siegert

JAHRSDOERFER		**SIEGERT**		
Malformation	**Score**	**Anatomical site**	**CT scan**	**Score**
Stapes present	2	EAC	Atretic-stenotic-normal	0-2
Patento oval window	1	Mastoid pneumatization	None-good	0-2
Tympanic cavity volume	1	Tympanic cavity volume	Small-big	0-2
VII C.N. route	1		Small-big	0-1
Incudo-malleolar complex	1	Tympanic cavity pneumatizazion	Modest-big	0-2
Mastoid pneumatization	1	VII C.N.	Aberrant-normal	0-4
Incudo-stapedial joint	1	Sigmoid sinus	Aberrant-normal	0-4
Round window	1	Malleus + incus	Absent-Dysmorphic-Normal	0-2
External ear	1	Stapes	Absent-Dysmorphic-Normal	0-2
Total	**10**	Oval window	Ossified-normal	0-4
		Round window	Ossified-normal	0-4
		Total		**0-28**

Surgery

In attempting surgical reconstruction in otodysplasia, the surgeon must have a perfect knowledge of the anatomical variants described in the literature and take care that an accurate pre-operative clinical and radiological assessment is carried out.

As far as ***minor otodysplasia*** is concerned, surgical techniques hardly differ from those employed in ossiculoplasties and in canaloplasties. Often, the tympanic membrane is of normal dimensions and structure; a tympano-meatal flap will permit tympanic cavity exploration and ossicular replacement surgery. The rate of success is comparable to the one reported in the treatment of chronic otitis.

Canaloplasty can be sometimes challenging in the cases that present a very tight stenosis, as compared with the treatment of obstructive canal esostosis; the main post-operative risk is restenosis, therefore we suggest a particularly long packing and silastic sheathing of the ear canal, together with frequent local medications under microscope control.

On the contrary, while treating ***major otodysplasia***, the surgeon may face several challenges, including the absence of an external auditory canal, a malformed ossicular chain with welding of malleus and incus, absence of common surgical landmarks such as tensor tympani tendon, pyrmidalis process, oval window; furthermore, the facial nerve is often aberrant, sometimes appears bifurcated and running outside the Fallopian canal.

Two surgical approaches are described to gain access to the middle ear:

1. Posterior transmastoid approach: it is the old one, which consists of drilling a wide mastoidectomy that allows to identify the sino-dural angle first, and then to proceed along the tegmina into the superior portion of the tympanic cavity. The advantages of this technique include an easier identification of anatomic landmarks and a lower risk of VII cranial nerve injury. Drawbacks are poor re-epithelization of the large surgical cavity and mastoid cavity infection, which may complicate the post-operative course; moreover, functional ossiculoplasty is unlikely to offer a significant gain, as in all canal wall down tympanoplasties.

2. Anterior transcanal approach (**Figure 4**): it is the currently used one, and consists of direct opening of the tympanic cavity after drilling of a new external auditory canal, with no drilling of mastoid cells. Once the small triangular area between the temporo-mandibular joint and the zygomatic root has been exposed, the tympanic tegmen is used as the superior landmark and the mandibular fossa as the antero-inferior one. Then, drilling of the atresia layer toward a medial direction allows to reach the tympanic cavity at an epitympanic level. The main advantage of the technique is the possibility to reconstruct a new external auditory canal and tympanic cavity as close as possible to a normal morphology and function.

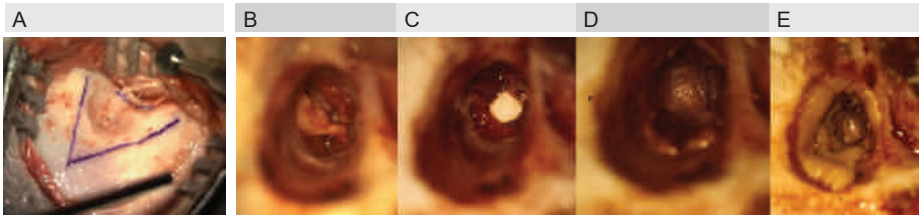


Figure 4. Intraoperative photographs showing some important surgical steps of the "anterior approach" atresiaplasty. **a)** landmarks for anterior approach canaloplasty; **b)** after drilling out the atresia plate the ossicular chain is exposed; **c)** replacement of the malformed not functioning ossicular chain with a TORP; **d)** miringoplasty; **e)** sheathing the ear canal with Tiersch flap.

Independent of the technique, some details deserve particular attention. Special care is recommended while drilling the most medial portion of the bony layer of atresia, in order to avoid trauma to the ossicular chain. To the purpose, circular dissectors placed medially to the atresia plaques can be useful to protect the ossicular chain, especially during drilling of the neo-anulus. This step is important to improve tympanic flap adherence and to prevent "blunting".

Once the middle ear has been exposed, it is very important to identify the facial nerve medially to the incudo-malleolar block, and successively the oval and round window. The latter is sometimes very difficult to expose, hiding behind the third tract of the facial nerve.

While attempting functional reconstruction the surgeon should, if possible, try to preserve and use the existing elements. Thus, even if malformed, the ossicles can be used if they are functionally valid, serving for sound transmission in contact with the neotympanic membrane.

In case the incudo-malleolar block is fixated, it will be removed and stapes

motility will be tested. In such situations a TORP or a PORP, or autologous incus remodeling, will serve the purpose. Footplate fixation won't always be effectively treated by means of a platinectomy, owing to difficulties finding a stable support for the prosthesis.

In order to create a stable tympanic membrane with minimal risk of lateralization, it is recommendable to drill the bony annulus medially to the ossicular chain. The tympanic flap can be then laid on the neo-canal walls, so as to cover the mastoid cells that have accidentally been exposed by drilling. The canal must be lined with Thiersch free skin flaps, harvested from hair-free body areas such as forearm and hip.

Meatoplasty is yet another crucial step. Connecting it with the neo-canal skin is of paramount importance, so that post-operative stenosis from scars is prevented.

For this specific purpose some authors suggested to suture the free skin flap to the margins of the external meatus, although this is a difficult task. Other authors[16] suggested that a skin incision should be performed at the meatus level, thus creating a posteriorly-based skin triangle that is sutured to the posterior bony margin of the neo-canal by means of 5-0 nylon stitches. Such a precaution is supposed to reduce the risk of re-stenosis. In our experience the best solution is to avoid difficult attempts of suturing and to use a sigarette shaped silatic sheathing of the ear canal that will guide riepithelization towards the meatoplasty.

Complications

In approaching functional surgery of otodysplasia it is important to inform the child's parents in detail about intra- and post-operative complications, although their incidence is different according to case series in the literature.

- *VII nerve injury*: although rare, it is the most dreaded surgery-related risk; in case of otodysplasia (both major and minor), it is higher than in anatomically normal ears owing to aberrant route, unexpected bifurcations, etc.
- *Neotympanic membrane lateralization:* this is the most frequently encountered complication, being found in 30% of cases[16,] and appears to be related to each individual patient's scar formation. When it occurs, it compromises the functional gain.
- *Post-operative stenosis of the new auditory canal:* reported in 15-31% cases[17], on one side it depends on the circular scar, and on the other side on the absence of the physiological self-cleaning properties of the skin lining a normal external auditory canal. The ongoing stacking of epithelial debris causes recurrent inflammatory processes, that constitute a continuous stimulus to scar formation: thus, granulation tissue tends to grow, subsequently replaced by retracting fibrosis. In the light of this, periodic and meticulous cleansing is strongly recommended, together with instillation of oily droplets that facilitate debris removal.
- *Purulent otorrhea:* unfortunately, this complication is not uncommon in operated patients, although its incidence is much lower with the direct transcanal approach. In the past, the posterior transmastoid approach caused this complication quite often, owing to a too large surgical cavity.

➢ *Sensorineural hearing loss:* this is a rare complication that can be effectively prevented by avoiding drilling on the ossicular chain and working directly on the footplate, and abstaining from platinotomy when there is no adequate support for the prosthesis.

In the literature, about 30 to 40% of cases are reported as needing revision surgery because of post-operative complications.

Functional results

Functional results present a high variability across case series, possibly due to the heterogeneity of described cohorts [17-19], to differences in surgical techniques and to a lack of commonly accepted outcome measures. **Table 3** describes the results reported in the literature and the experience of ENT Department from "Bambino Gesù" Pediatric Hospital.

Table 3. Published case series of functional otologic surgery of major and minor otodysplasias.

AUTHOR	PATIENTS/ OPERATED EARS	PERIOD	UNI/ BILATERAL	R/L	M/F	MINOR/ MAJOR
Pinelli '80	57 / 81	20 (1960-1980)	58%-42%	80%-20%	64%/36%	30%-70%
Manach '87	109 / 125	7 (1979-1986)	76%-24%	67%-33%	55%-45%	0%-100%
Schuknecht '89	55 / 69	24 (1962-1986)	75%-25%	51%-49%	55%-45%	26%-73%
Crabtree '93	29 / 39	12 (1978-1990)	32%-68%	100%-0%	52%-48%	23%-77%
Chang '94	21 / 25	5 (1987-1992)	57%-43%	58%-42%	85%-15%	20%-80%
De la Cruz '95	70 / 92	9 (1986-1995)	63%-37%	77%-23%	63%-37%	0%-100%
De la Cruz, '03	116/116	17 (1985-2002)	65%-35%	59%/41%	60%/40%	
Div.ORL - OPBG	75 / 90	30 (1981-2010)	52%-48%	55%-45%	57%-43%	59%/41%

Protocol for the management of otodysplasia

In the management of otodysplasia, it is necessary to integrate solutions offered by **modern otomicrosurgery, semi-implantable hearing aids** and **auricle reconstruction**. For this reason, the definition of a protocol is important for an appropriate indication and timing of the numerous treatment options, and in order to obtain an early hearing and language rehabilitation. In particular, cosmetic and functional surgery must be well planned, so as to avoid interference between the otologist and the plastic surgeon.

As a general rule, the functional rehabilitation of the newborn with bilateral otodysplasia has to be considered an absolute priority, with bone conduction hearing aids fitted from the first months of life, replacing them with implantable

devices as soon as possible. Functional otosurgery is not indicated before age 10, that is, before plastic surgery can be undertaken. For this reason, application of external bone conduction hearing aids as Softbands must be prescribed as early as the first months of life. As soon as the patient's skull bone is thick enough (in general, starting from 3-4 years of age), a bone conduction implant can be used to optimize hearing rehabilitation. Modern solutions include the classic percutaneous Baha and the latest generation transcutaneous devices such as Bonebridge™ (MED-EL), Sophono™ (Sophono Inc.) and Baha Attract™ (Cochlear).

Once hearing is optimized, patients will be able to choose whether to keep the bone conduction implant or to attempt functional reconstruction through surgery, which is recommended on the less malformed side. Some patients will receive significant benefit from both solutions, for example a Baha implant on one side and functional reconstruction of the contralateral ear.

Another general rule, consists of planning the first step of auricle reconstruction before functional micro-otosurgery, which may irreversibly alter the surgical field and thus make plastic surgery more difficult. On the other hand, the plastic surgeon should pay attention not to cause changes of the external ear anatomy that may interfere with atresiaplasty: for example, modyfiing the auditory canal course in attempting to restore symmetry to the auricles.

Modern techniques of total auricle reconstruction are derived from the original Brent technique[19], based on the harvesting of rib cartilage. Generally, it is not undertaken before age 10 and includes a minimum three steps:

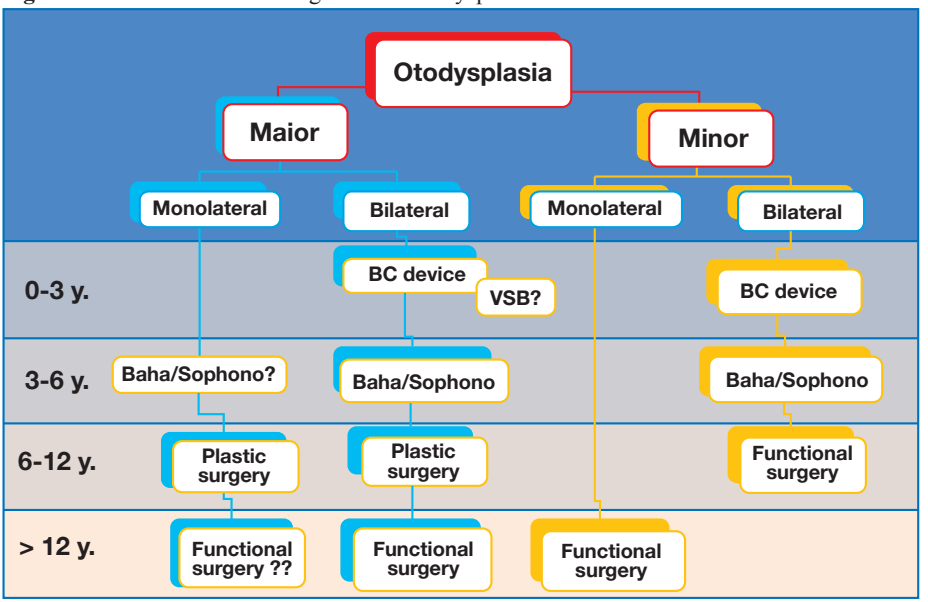
1. Harvesting of rib cartilage, assembling of an auricle and banking of the reconstructed auricle under the skin where the malformed auricle used to be.
2. Elevation of the reconstructed auricle by creating a skin flap.
3. Refinement of the reconstruction (better shaping of tragus, helix, etc).

Steps are distanced by a minimum three months off each other, so as to allow scar formation. Siegert[20] has proposed adding atresiaplasty in the same session as elevation of the auricle.

A valuable alternative to auricle reconstruction is represented by **auricular episthesis,** consisting of auricle-shaped silicone prosthesis that are fixated on the temporal bone by means of titanium Baha fixtures. The advantages of this option include its being time-sparing (only one surgery is needed) and the greater probability of a good cosmetic outcome as compared to total auricular reconstruction. This, it finds an indication in cases of unsuccessful auricular reconstruction.

The treatment protocol of otodysplasia used in our Institution is illustrated by **Figure 5**. From **Tables 4** to **6** we compare published data related to surgical technique, complications and functional results to those collected from our experience. Overall, functional otomicrosurgery as a treatment option has been progressively abandoned over the years, in favor of the increasing application of semi-implantable bone conduction hearing aids such as Baha (**Figure 6**). This is easily explained by considering the unquestionable functional advantages and the relatively low risks, together with the possibility of an early auditory and language rehabilitation in children as young as 2-3 years.

Figure 5. Protocol for the management of otodysplasia.



VSB = Vibrant Soundbridge
y. = years

Table 4. Intraoperative findings in aural atresia

Author	Post Approach	Ant Approach	Aberrant VII	Cholesteatoma of the external/ middle ear	Ossicular chair preservation	Surgeon gave up functional surgery
Pinelli '80	47 (58%)	34 (42%)				
Mattox-Fisch '86	15 (57%)	11 (43%)				
Manach '87	0	125 (100%)		0	65.5%	21.6%
Schuknecht '89	27 (42%)	37 (58%)		13% - 4.3%	42%	8%
Crabtree '93	30 (100%)	0		5%		17.9%
Chang '94	1 (5%)	19 (95%)	64%	0	24%	32%
De la Cruz '95 [17]	28 (30.5%)	64 (69.5%)	49.2%	8.7%	72%	1.4%
Lambert '98	0	55 (100%)		4.8%	84%	
De la Cruz '03			23.7%	12%	71%	
Div.ORL - OPBG	0	37 (100%)	31%	8%-3%	38%	11%

Table 5. Functional results of otodysplasia surgery.

Author	POST-OP AC-PTA	RESIDUAL POST-OP AC-BC GAP	PRE/POST OP AC GAIN
Pinelli '80			mean = 12 dB (post appr.) mean = 33 dB (ant appr)
Mattox-Fisch '86		< 30dB = 6.6% (post. appr.) < 30dB = 27.2% (ant. appr.)	>30dB = 6.6% (post. appr.) >30dB = 45.4% (ant. appr.)
Manach '87		< 25dB = 28% (early) < 25dB = 18% (from a distance)	
Schuknecht '89	< 20dB = 30% (ant) 8% (post) < 30dB = 50% (ant) 15% (post)		mean = 19.3 dB (post. appr.) mean = 22.6 dB (ant. appr.)
Crabtree '93	< 30dB = 16.6% < 40dB = 56.6%		
Chang '94 [21]		< 20dB = 19% < 30dB = 52%	
De la Cruz '95 [17]		< 20dB = 26.2% (mean follow-up = 2.6 ys < 30dB = 59.5%.)	
Lambert '98	<20dB = 36% (short term) 32% (long term) <30 dB =70% (short term.) 50% (long term)		
De la Cruz '03	<30 dB = 51%		
ENT Dept. – B.G.	<30 dB = 18%, <40 dB = 44%		

Table 6. Post-operative complications.

AUTHOR	STENOSIS OF THE NEO-CANAL	OTORRHEA	NEOTYMPANIC MEMBRANE LATERALIZATION	HIGH-FREQUENCY SENSORINEURAL LOSS	TRANSIENT/PERMANENT VII NERVE PARALYSIS	REVISION SURGERY
Pinelli ‘80	14%	29%			0%	
Mattox-Fisch ‘86	27%	23%			0%	
Manach ‘87	17.3%	34.6%	22.4%		9.6% - 2.4%	
Schuknecht ‘89	26%	4.3%	4.3%	4.3%	7.2% - 0%	17.3%
Crabtree ‘93	33%	31%	10.2%		0%	33%
Chang ‘94	48%	4%	8%	0%	0%	32%
De la Cruz ‘95	10.3% - 12.1%	10.4%	8.6%	5.2%	1.7%	29.6%
Lambert ‘98	18% - 4%		8%	3%	1.5%	34%
De la Cruz, ’03 (19)	8%		3.4%	10.3%	0%	17.2%
Div.ORL - OBG	33%	8%	15%	2%	0%	27%

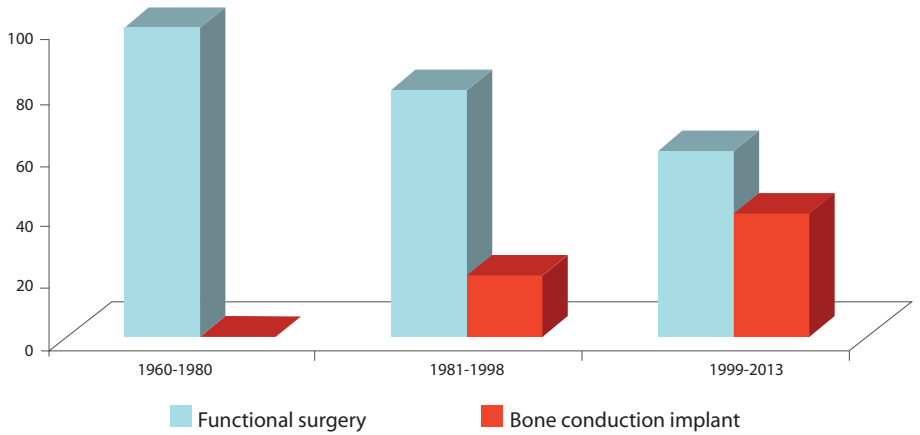


Figure 6. Prevalence (%) of functional otologic surgery vs bone conduction implants over the years.

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