REVIEW

Congenital aural atresia reconstruction: a surgical procedure with a long history

Leonidas Manolopoulos a, George X. Papacharalampous a, Ioannis Yiotakis a, Dimosthenis Protopappas c, Petros V. Vlastarakos a, Thomas P. Nikolopoulos b,*

a 1st ENT Department, University of Athens Medical School, Hippocrateion General Hospital, Athens, Greece
b 2nd ENT Department, University of Athens Medical School, Attikon General Hospital, 1 Rimini st., Chaidari, Athens, Greece
c ENT Department, Naval Hospital, 70 Deinocratous st, Athens, Greece

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Summary  Background: Pinna deformities, combined with congenital aural atresia, have been a matter of serious debate in the literature as they are associated with major aesthetic and functional problems that are difficult to manage. These problems have been described as early as 2000 BC. The aim of the present article is to approach the whole problem as one (pinna malformation and aural atresia) and present the history as well as the current approaches in reconstruction.

Methods: Extensive literature search and medical history books were used as scientific sources.

Results: For many centuries, the prevalent view was that any surgical attempts to reconstruct the pinna and the ear canal were of little value. In addition, the aesthetic result of these early surgical procedures was mostly unacceptable. Over time, new surgical techniques and synthetic materials were used, leading to satisfactory and lasting aesthetic and functional results in selected patients, improving their quality of life, while reducing the complication rate. However, many cases are still challenging for plastic surgeons and ENT surgeons alike.

Conclusions: Despite significant progress in the field, surgery for pinna deformities combined with congenital aural atresia still remains one of the most challenging and risky procedures. Accurate audiological evaluation of newborns as well as assessment of their craniofacial development is necessary and can help the plastic surgeons and otologists choose proper candidates for surgical repair and a suitable and age-appropriate therapeutic plan. History and repeated failures have taught us that close multidisciplinary approach is of paramount importance.

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* Corresponding author. Thomas P. Nikolopoulos, 114 Vas. Sophias Av, Athens 11527, Greece. Tel./fax: +302107778095.
E-mail address: thomas.nikolopoulos@nottingham.ac.uk (T.P. Nikolopoulos).

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Prehistory

Pinna deformities or aplasia combined with congenital aural atresia have been important malformations affecting the quality of life (both aesthetical and functional) of patients since ancient times. Such deformities have been described in the teratological tablets written by the Chaldeans of Mesopotamia (Figure 1) about 2000 years BC and also in the tablets of Assyrio-Babylonians found in the library of the Assyrian king Assurbanibal (669–625 BC) (Figure 2). Moreover, complete absence of the external auditory meatus has also been found in prehistoric skulls. In 1990, Hodges et al. reported a prehistoric case with congenital atresia of the external auditory meatus. Wells too presented a probable case of aural atresia in 1962, dating from Anglo-Saxon times in England.

Thus, many societies through history have attempted to reconstruct these malformations with various, usually very poor, results aesthetically and functionally. The oldest known written account of surgical reconstruction of ear lobes is found in the Sushruta Samhita, a Sanskrit text, written by an Indian physician named Sushruta who lived between 600 BC and 400 BC (Figure 3). Sushruta Samhita (samhita is Sanskrit for 'encyclopedia') is based on the Hindu text Ayurveda (knowledge of life) and provides thorough descriptions of examinations, diagnoses, treatments and procedures involving a variety of ailments. Sushruta outlined some procedures for reconstructing an earlobe using skin grafted from the cheek. He also devised a classification of ear-lobe defects and set out 15 different techniques for pinna reconstruction.

Failed surgical procedures led to disappointing results initially and, for many centuries, most surgeons were against any type of surgical intervention in such cases.

In a previous study, we described the long history of various pinna reconstruction procedures until the widely accepted techniques designed by Satoru Nagata and Burt Brent came into being. However, pinna deformities are often combined with congenital aural atresia and plastic surgeons need to closely cooperate with otologists while designing the appropriate reconstructive procedures. Therefore, this article approaches the whole problem as one (pinna malformation and aural atresia) and presents the history and current approaches in reconstruction.

The era until 1900

The first surgical attempts to repair congenital aural atresia were made in the middle of the 19th century. Thomson (1845) published an article reporting his experience of three patients operated upon by other surgeons for congenital aural atresia. In the first two cases, the operation was terminated after an incision had been made through the skin and soft tissue, when a wall of dense bone was encountered. In the third case, although the surgeon managed to maintain a recognisable external ear canal, this was soon re-closed. Thomson concluded that the complete closure of the bony part of the ear canal poses an insuperable obstacle to be relieved by surgical operation and that the malformation, which is externally visible, indicates the existence of other defects in the deeper parts of the ear. In 1882, Kiesselbach performed the first deep operation on a 6-month-old child with congenital aural atresia. Unfortunately the operation resulted in facial paralysis.

The most popular operation at the start of the 19th century was the opening of the antrum and aditus and laying in a skin graft (often 10 days later during a second-stage operation). The middle ear cavity was rarely entered into and the hearing improvement was minimal.

The first decades after 1900

Bezold and Siebenmann, in 1908, described a surgical procedure in which the antrum was opened from the mastoid and a wide canal was made by lining the mastoid cavity with grafts. In the same year, Alexander presented a case in which a mastoidectomy was performed and the skin flaps were made from the posterior part of the wound. He concluded that in cases of unilateral atresia, operation is recommended only if otitis media or mastoiditis is present on the affected side. In the case of a bilateral problem, Marx suggested surgery only if the deafness was marked and the inner parts of the ear (especially the labyrinth) were healthy.

In 1914, Page reviewed eight cases including one of his own. The usual surgical procedure included opening of the mastoid and cleaning out the cells down to the tympanum. An opening was made in the auricle, then the mastoid was closed and a skin graft placed in the mastoid wound, through the artificial meatus. Five out of eight cases showed hearing improvement with this procedure. In the 1930s the usual surgical procedure was to make a small opening on the atretic plate and cover this area with a skin flap.
graft. Surgeons usually avoided large openings because they thought this may expose the middle ear cavity to the risks of scar tissue and fibrosis.

In 1925, Beck examined the psychological part of the problem. He found that the attitude and reaction of the patients to their deformities was better after an operation which provided them with a constructed meatus and auditory canal, even though their hearing was not considerably improved.

In the 1940s, Patee in USA and Ombredanne in France presented new surgical techniques trying to improve the appearance of the ear by forming a patent meatus and providing better hearing (even by fenestrating the lateral semicircular canal).

The era from 1950 to 1970

The second important step for surgery of congenital aural atresia after the Patee and Ombredanne's procedures was the development of the modern tympanoplasty techniques which were presented in the 1950s. The methods described by Wullstein and Zollner were adopted by many surgeons in their attempts to correct aural atresia and the ossicles finally became structures to be saved rather than discarded. Howard House in 1953 and Ruedi in 1954 presented their experiences in using Patee's surgical technique. However, the latter author admitted that the long-term hearing results were disappointing. In 1957, Meurman presented his series of 74 operated patients, most of them...
with unilateral atresia. It was the largest series ever presented until then.

In 1960, Bellucci used the techniques of Wullstein and Zollner in trying to preserve the middle ear architecture. However, the result seemed to be questionable in cases of unilateral atresia.

Derlacki in 1968 was one of the first surgeons who supported the role of polytomographic radiology in the preoperative assessment of patients with congenital aural atresia. He usually used a thin full-thickness skin graft as a substitute tympanic membrane and a split-thickness skin graft for the meatus. However, he managed to provide a very good hearing result in only 25% of his cases; he also had discharge problems in about one-third of the operated ears. These were probably the reasons that made him suggest surgical intervention in bilateral cases only. In 1967 Scheer, followed by Crabtree in 1968, also attempted various ossiculoplasties.

It is clear that the aesthetic result and the opening of a closed ear canal eventually became one of the two surgical objectives as an acceptable functional (hearing) outcome was added to the surgical planning.

In 1969, Gill published his work on 83 operated ears which is still considered to be one of the landmark articles on congenital aural atresia. According to Gill, bilateral cases should have one ear operated upon as early as possible, if surgical correction is indicated for the specific type of the deformity. Certain criteria should be met before the operation is performed, including radiological demonstration of inner ear presence and audiometric demonstration of good bone conduction. As far as the optimum age for the surgery is concerned, Gill suggested that this depended on whether the deformity was unilateral or bilateral. He preferred to operate when the child was between 12 and 18 months of age in bilateral cases. In unilateral cases, he advocated surgery ideally after puberty but as this interfered with schooling and employment training he concluded that it was better for the patient to be operated upon between the ages of 4 and 6 years. In the analysis of his results Gill reported acceptable results in a fair proportion of his patients. By 1971, his series had increased to 113 operated ears on 95 patients.

The era from 1970 to 1980

Colman (1971, 1974) presented his surgical series of 184 cases during this decade. Half of these patients had bilateral atresia. However, postoperative results appeared to be excellent in patients with narrow external auditory canal, fixed ossicles and deformed stapes. In more serious cases, the results seemed to be much poorer.

Jahrsdoerfer published an article in 1978 presenting his experience of surgical correction of congenital aural atresia. He classified the malformations as 'minor' when limited to the middle ear and 'major' referring to all cases of atresia and stenosis of the external auditory canal. Surgical correction was attempted on 20 ears in 18 patients. Jahrsdoerfer usually preferred anterior approaches and his techniques eventually became very popular. He often used a fascia graft overlay in conjunction with a centre-hole skin graft. The results eventually became satisfactory. However, he stated that surgery in unilateral cases should take place only in properly selected patients. With regard to bilateral cases, he suggested that functional criteria should be applied, hearing being very important. Regarding the optimum age for the operation, Jahrsdoerfer stated that in cases of bilateral atresia a bone conduction hearing aid should be fitted as early as the third month of life, regardless of the surgeon's age preference for the procedure. On the contrary, in unilateral cases he suggested that the operation should not take place until the child is old enough to decide for himself.

1980 till the present

Modern imaging techniques, especially high-resolution computed tomography (CT) scanning, developed in the 1980s, provided surgeons with accurate anatomical details regarding the middle and the inner ear and the mastoid cavity. This fact improved pre-operative planning and led to better functional and aesthetic results in the majority of the operated patients. Several refinements in canalplasty, tympanoplasty and ossiculoplasty for congenital aural atresia have been made during the years since 1980 and large surgical series have been reported by various surgeons. Both, aesthetic and functional, results are considered important and feasible, artificial pinnae are now close to excellence and plastic surgery has evolved significantly with acceptable outcomes. However, difficulties and complications are still a matter of concern.

In 1989, Schuknecht, one of the leading modern otologists, presented his experience in surgical correction of 69 ears with congenital aural atresia. In five out of the 62 ears the operation resulted in temporary facial nerve palsy and in another five ears, the surgery was terminated due to significant anatomical malformations encountered during the procedure. However, 30% of the patients who underwent canalplasty and 8% who underwent mastoidectomy with stapediopexy obtained an excellent hearing result postoperatively.

In 1992, Jahrsdoerfer created a classification system for congenital aural atresia, based on high-resolution CT scan findings of the temporal bone. The parameters assessed were the presence of stapes, the condition of the oval and round window and middle ear cavity, the facial nerve, the malleus-incus complex, the pneumatisation of the mastoid, the incudostapedial connection and the appearance of the external ear. One point was given to every parameter that was within normal limits, excluding the presence of stapes for which two points were given. According to Jahrsdoerfer, the obtained score (out of a maximum 10 available points) expresses the possibility of satisfactory postoperative results. However, his classification is mainly functional, as he gives only one point to the appearance of the external ear. It is now evident that plastic surgeons as well as ENT surgeons have different and, sometimes, conflicting priorities.

In 1993, Shih and Crabtree reviewed 39 surgical cases of congenital aural atresia for complications and long-term results. They proposed widening of the narrow external meatus through systematic drilling, the elevation of canal skin in order to ensure adequate examination of the
conducting mechanism and the ossicular reconstruction with autogenous tissues or prosthetic materials, if needed. In more serious cases, they proposed a more extensive post-auricular approach, removal of the atretic plate and ossicular reconstruction with suitable materials (autogenous grafts or prosthetic materials). The two most common complications were external ear canal stenosis and chronic infections with recurrent otorrhea. The incidence of stenosis was 33% in primary cases, whilst infection appeared in 31%. The use of split-thickness instead of full-thickness skin graft was associated with fewer complications.

In 1994, Chang et al. introduced a modified anterior approach and repositioning of the pinna by a z-plasty incision. The atretic plate was removed and tympanoplasty was carried out. The posterio-inferior part of the new external auditory canal was covered with inferiorly based periosteal flap. The most common postoperative complication encountered was external ear canal stenosis.

In 1998, Lambert presented a retrospective study of 55 patients (59 ears) who underwent surgery for congenital aural atresia during an 11-year period. He used anterior surgical approach under continuous facial nerve monitoring. An external canal was created and the surrounding bone removed from the ossicular chain, so the latter was centred on the new external auditory meatus. A fascia graft was used to construct a tympanic membrane. The external canal was finally lined with a split-thickness skin graft overlapping the fascia graft. Revision surgery was necessary in approximately one-third of the patients. The main complications that occurred, as reported by Lambert, were facial paralysis (1.5% of the patients) and significant hearing loss (3%).

In 1995, De la Cruz et al. reported their results in a series of 92 ears, operated for congenital aural atresia using a mastoid approach. All primary cases underwent...
atresiaplasty with split-thickness skin graft. In case of revision surgery, atresiaplasty was performed and the use of skin graft was decided depending on the particular situation encountered. The most common complications reported were external auditory canal stenosis (10% of primary cases and 4% of revisions), and lateralisation of the tympanic membrane (9% of primary cases and 15% of revisions).

De la Cruz and Teufert presented their increased surgical series in 2003.40 The total number of the operated atretic ears in this series was 116. The main complications were soft tissue stenosis of the external auditory canal (in 8% of primary cases and 3.4% of revisions) and re-fixation of the ossicular chain (in 11.5% of primary cases and 6.9% of revisions).

As the surgical techniques employed were gradually improved and various modifications and materials adopted, De la Cruz and Teufert presented an article in 2004 in which they attempted to compare the results and complication rates in cases of aural atresia operated upon before 1994 (36 cases) and within after 1994 (80 cases).41 The reason for this, according to the authors, was that all the modifications used in practice, such as the use of argon laser, thinner split-thickness skin grafts and the use of Silastic sheets and Merocel wicks in the external auditory meatus, had been performed as a routine procedure at their institute by 1994. With regard to the main complications, they reported that the most common were soft-tissue stenosis and bony growth of the external auditory canal (in 3.8% of the new and 13.9% of the old cases) and re-fixation of the ossicular chain (in 3.8% of the new and 25% of the old cases).

Ralf Siegert42,43 also offered a significant contribution to combined reconstruction of congenital aural atresia and severe microtia, as he proposed a combination of plastic surgery for the auricle and functional surgery for the middle ear. This surgical procedure consists of three basic stages. In the first operation, autogenous cartilage is harvested and the auricular framework fabricated and implanted (Figure 4). The tympanic membrane and the external auditory canal are also prefabricated and put in a subcutaneous pocket (Figure 5). In the second operation, the new auricular framework is elevated and this procedure is combined with the operation for atresia, using the prefabricated tympanic membrane and the external auditory canal (Figure 6). Finally, in the last operation, the cavum conchae are deepened and the external auditory meatus is opened and covered with a skin graft. No re-stenosis of the ear canal was observed in the Siegert’s series.42,43 The aesthetic result of the reconstructed pinna was also very satisfactory (Figure 7).

The use of bone-anchored hearing aids and middle ear transducers

Bone-anchored hearing aids (BAHAs), one of the significant achievements in modern otology, have almost replaced conventional bone-conducting hearing aids and are considered to be a suitable alternative to surgical treatment for congenital aural atresia.44 Such hearing aids are also used prior to surgery in order to ensure speech and language development. Baha Softband (fitted with a headband without surgery) is also a valid but temporary method of management in children with congenital aural atresia, especially in children who are too young for percutaneous BAHA surgical insertion (Figure 8).45 Both, classic BAHA and Baha Softband, provide children with adequate hearing, so
that language development is in accordance with their cognitive development.\textsuperscript{44,45} Fully implantable hearing aids may also be involved in the treatment of congenital aural atresia, although they have not yet been as widely accepted as BAHA.

Despite the progress and the various modifications of the surgical techniques for atresia reconstruction, a hearing loss may still remain after all these procedures. Therefore, various types of hearing aids may still be needed. Thus, Siegert et al. proposed in 2007 the use of a specially modified fully implantable hearing aid in patients with congenital aural atresia and severe co-existing malformation of the middle ear.\textsuperscript{46} The surgical instruments, the transducer and the operative technique were modified to meet the specific needs of these patients thus providing a completely new dimension to their rehabilitation.\textsuperscript{46}

Despite the increasing surgical experience, the modern surgical techniques and the new synthetic materials which are used, surgery for pinna deformities combined with congenital aural atresia still remains one of the most challenging and risky procedures. Accurate audiological evaluation of newborns as well as assessment of their craniofacial development is necessary and can help the plastic surgeons and otologists to choose the proper candidates for surgical repair and proceed with a suitable and age-appropriate therapeutic plan. The aesthetic result, along with the postoperative hearing gain, seems to be satisfactory and lasting in selected patients, improving their quality of life, while complication rates appear to be significantly reduced.

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None.

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None.

**Ethics approval**

Not needed because of the nature of the study.

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