Malformation and plastic surgery in childhood

Abstract

Malformations of the head and neck show a huge variety of clinical symptoms with functional and esthetic consequences. Often times its rehabilitation requires multi-staged and multi-disciplinary procedures and concepts. These must consider eating, speech, mimic expression, hearing and "esthetics" or at least "normality".
A survey of the most common head and neck malformations and their treatment options are presented here.

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1 Introduction

Malformations of the head and neck include a huge variety of different diseases, which could not only fill whole congresses, but which are the main interest of many different scientifical societies. They are characterized by various functional and esthetic deficits and require an interdisciplinary, multi-staged rehabilitation concept in many cases. The treatment of malformed children must include esthetic surgery with its psychosocial benefits as well as functional aspects of breathing in cleft noses, eating, speech and mimic expression in facial paralysis and hearing in severe ear malformation. Therefore facial plastic surgeons should perform the necessary steps himself or organize the functional rehabilitation by close cooperation with appropriate colleagues.

In these chapters we present a short survey of some of the most common malformations of the head and neck and its surgical treatment options.

2 Fistulas and cysts of the neck

2.1 Introduction

The origin of neck fistulas and cysts are ectodermal tissue remnants remaining in the mesoderm. They are differentiated into lateral fistulas and cysts which are vestiges of the branchial clefts or the cervical sinus, and median ones, which develop from parts of the thyreoglossal duct. Fistsulas develop if there is an opening to the outside through the skin. Cycts arise when they are completely closed and its fluid cannot drain. It is a common misunderstanding when one is searching for a "duct" in cysts or for a cystic formation in fistulas. Although their origin is the same clinically they differ a lot and require different surgical techniques.

Whereas histologically proven cysts – except for its tendency to enlarge and its risk of becoming infected – are completely harmless, clinically diagnosed cysts in adults have a relatively high risk of being malignant [5]. Cervical metastasis or lymphomas of the neck often times show cystic components. Therefore cysts of the neck need histological verification.

2.2 Diagnosis

Clinical and sonographic examination of the neck is necessary. It is controversy whether to dye fistulas or not preoperatively, because depending on its anatomy and the applied pressure only part of the fistula might be reached pretending a too short duct or the dye might penetrate the surrounding tissue exaggerating their real sizes and lengths.

2.3 Surgery

After making an incision in the RSTL lateral cysts are exposed and should be removed completely. Median cysts (Figure 1) which are adjacent to the hyoid should be removed including the medial part of the hyoid bone to avoid remnants of epithelium being left behind which could lead to a recurrence.

Fistulas should be removed completely too, including part of the hyoid as described above. Lateral fistulas can extend far cranially up into the tonsillar groove (Figure 2). In those cases additional, more cranial incisions as well as simultaneous tonsillectomy might be necessary.

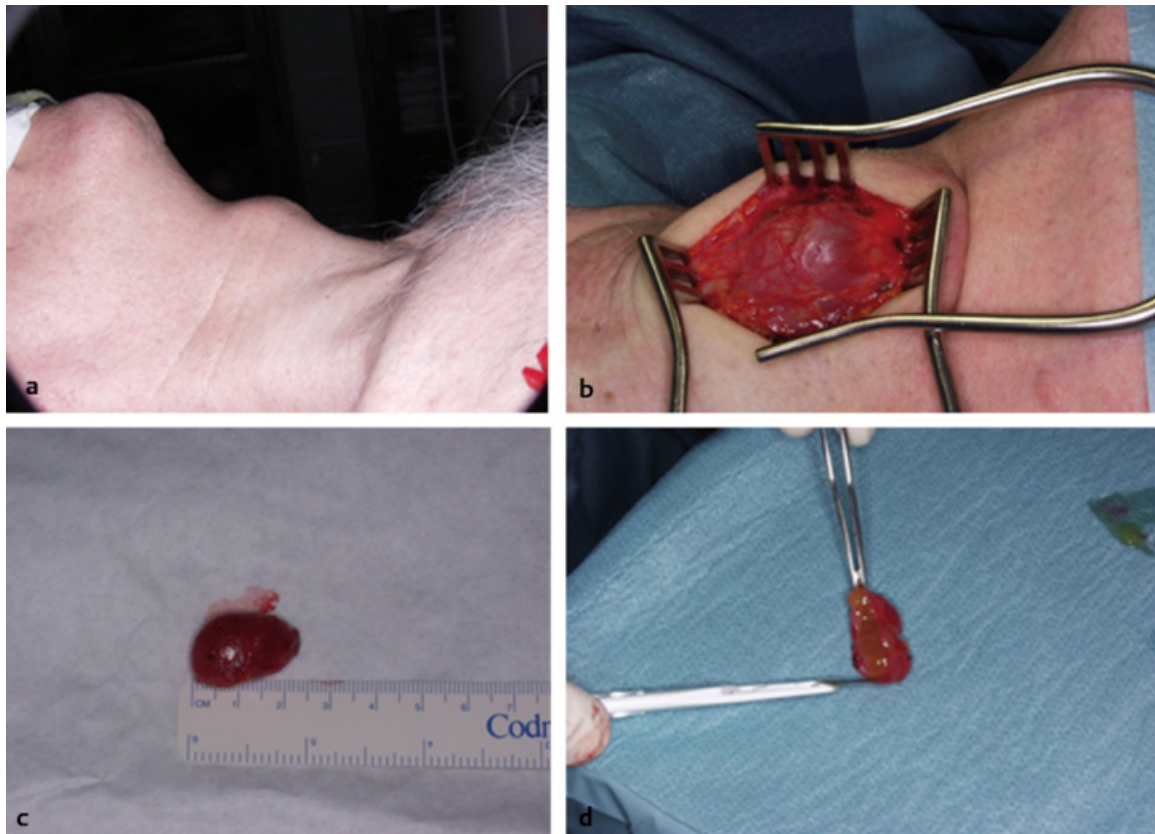We suggest operating under magnification, i.e. lupes or microscope, and without the discussed dying of the fistulas.

Figure 1: Median neck cyst
a: Smooth median neck cyst; b: Exposed cyst; c: In toto removed cyst; d: Opened cyst
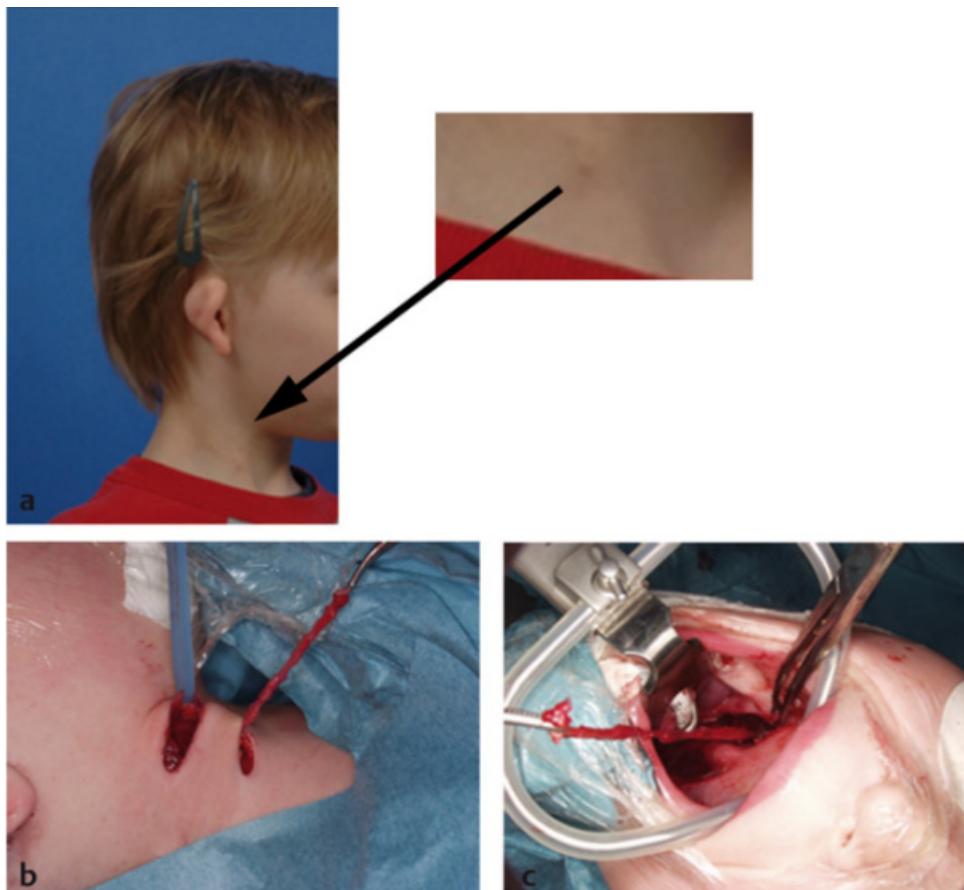

Figure 2: Lateral neck fistula
a: Hardly visible opening; b: Mobilized fistulas through two incisions in the RSTL; c: Fistulas pulled through into the mouth

3 Nose

3.1 Introduction

One of the most common malformations of the head and neck with an incidence of about 1:500 are cleft lips and palates with the so-called cleft nose. It is characterized by lateralization of the alar, stenosis of the nasal entrance, asymmetry of the columella and tip as well as septal deviation. It leads to esthetic abnormalities as well as impaired nasal breathing.

In newborns the main problems are difficulties with eating. This is improved by closure of the cleft lip and palate during the first two years of life.

The maxillary cleft can be filled with spongiosa in the first year of life or shortly before the upper thirds are erupted. The cleft nose is typically corrected after the age of about 15.

In contrast to the "cleft nose" as part of the cleft lip and palate deformity we talk about "clefts of the nose" or "nasal clefts" if the cleft is primarily located in the nose (Figure 3). Microforms of nasal clefts can be the broad nose, possibly combined with hyperteleorism. The most comprehensive classification was published by Tessier [45]. He described clefts of 15 different locations in the face and subdivided them further into clefts of soft tissue and those of bones.

Figure 3: Nasal cleft type Tessier No. 1

Rare malformations of the nose are median fistulas which have an incidence of 1:30,000. Whereas they can be inconspicuous on the nasal dorsum as an opening not being bigger than a needles head they can reach all the way through the anterior skull base into the cranium. Surgeons should always keep the possibility in mind when starting to operate on these. Nothing would be worse as leaving part of the epithelium behind which would lead to a recurrence for sure. Analogue to neck fistulas the nasal ones can also develop as cysts when they have no connection to the skin.

3.2 Diagnosis

Diagnosis of nasal malformations is primarily based on the clinical examination. Imaging, especially MRI, is indicated in nasal cysts and fistulas to evaluate their extension towards the anterior skull base. Functional examination of breathing, i.e. rhinomanometry, are a conditio sine qua non before performing functional septorhinoplasties.

3.3 Surgery

Correction of the cleft nose is considered to be difficult since quasi all structures of the nose are malformed and therefore complex reconstruction is necessary. Gubisch [15] presented a comprehensive technique, which basic points are extra corporal septal reconstruction, reliable support of the columella and nasal tip, symmetric repositioning of the alars and tension free epithelial lining of the reshaped domes. Like other septorhinoplasties these corrections should be postponed after about age 15 because it should not affect nasal growth (Figure 4).

Figure 4: Alio loco preoperated cleft lip and palate with "typical cleft nose"
a + b: Pre, c + d: post septorhinoplasty

Nasal clefts are corrected like broad noses. Specifics may be far caudally reaching proportions of the skull base with smooth transitions in cranial celes (meningo-, meningoencephalo-, meningoencephalocystoceles) that need to be examined by CT preoperatively.

Similar to the nasal clefts also nasal fistulas may extend to or even through the skull base. Despite optimal preoperative imaging, each surgeon must be prepared for unexpected intraoperative situations, since the fine, mostly collapsed fistulas cannot always be detected preoperatively. In cases of suspected intracranial pathologies a combined extranasal access with a bicoronal incision should be considered.

4 Ear

4.1 Introduction

The malformations of the ear have an almost infinite variability of morphological changes. Therefore, the correction or re-construction or, more precisely, construction of an auricle requires an enormous methodological range of different surgical techniques.

In particular, in the malformation surgery, we are often confronted both with functional disorders and aesthetic impairments. The rehabilitation of the affected children must take into account both aspects, and combine each other conceptually. This applies to the cleft lip and palates (see above), the facial paralysis (see below) and in particular for severe ear malformations with their externally visible disfigurement and conductive deafness. For them, the plastic head and neck surgeon must also ensure the hearing rehabilitation or perform the necessary steps himself.

4.1.1 Embryology

The auricle begins its development between 21 and 22 days post conceptionem (p.c.) with an ectodermal thickening in the ear region, in which a dimple develops around the 28th day p.c. [47].

By day 38 p.c. six mesenchymal hillocks evolve, arranged around the first branchial cleft, each 3 on the first (mandibular) and second branchial arch (hyoid arch). The first cranial, later ventral hillocks of the mandibular arch, form the anterior part of the external ear (tragus and crus). The caudal and dorsal later 3 hillocks of the hyoid arch form the posterior auricular shares. The concha and the external auditory canal develop from the first branchial cleft.

In addition to an increasing formation and enlargement, the ear moves of an initially antero-caudal into a dorso-cranial position.

During the embryonic and fetal development the relative amount of the mesenchyme from the mandibular arch decreases significantly. It is estimated that about 85% of mature pinna structure is formed solely by the hyoid arch [47].

Disturbances of this complex development and differentiation process at different temporal and structural levels lead to the almost infinite variety of form variants and dysplasias of variable severity.

4.1.2 Anatomy of the external ear

The external ear includes the pinna and the external auditory canal. The pinna has a characteristic relief facing front and a less distinguished posterior relief. With the exception of cartilage free lobule, the static basis of the auricle is formed by an elastic cartilage, which has a thickness 1–3 mm [11], [37], [47].

The anterior skin is fixed to the perichondrium without subcutaneous tissue in-between being almost immobile.

It is 0.8–1.2 mm thick. In contrast the posterior side has a subcutaneous layer of 1.2–3 mm which leads to a certain mobility [37].

The blood supply of the auricle is variable and dominated by branches of the superficial temporal and posterior auricular vessels.

Innervation of the auricle is due to branches of N. auricularis magnus, N. auriculotemporalis and N. occipitalis minor.

The auricular muscles are relatively unimportant and small. Two muscles might be detected intraoperatively: M. auricularis posterior and M. auricularis superior. In addition, the auricle is stabilized by one posterior and two anterior ligaments [47].

The lymphatic fluid of the auricle is drained into the superficial and deep neck nodes, into the parotid gland and towards the submandibular nodes and in addition to the postauricular and mastoid lymph nodes.

4.1.3 Esthetic units of the auricle

The esthetic units of the auricle are shown in Figure 5. In all reconstructive procedures they should be respected. Incisions should be placed along their borders and not through them as along as possible.
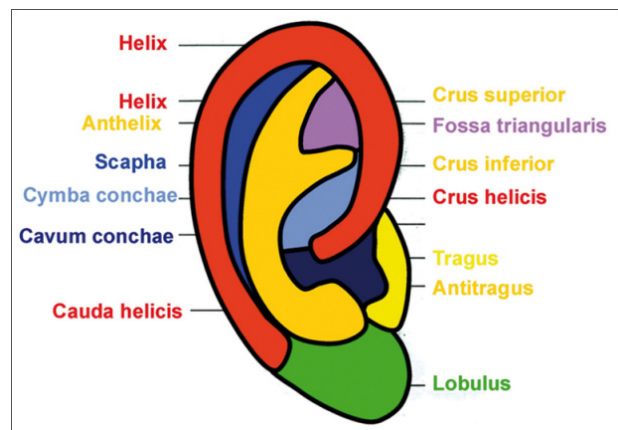

Figure 5: Scheme of the esthetic units of the auricle

4.1.4 Anthropometry of the auricle

The size and position of the auricle as well as its relief is of utmost importance for corrective and reconstructive auricular surgery. The knowledge of these data in relation age, sex and body stature are necessary for the individual surgical planning [37].

The length of the auricle depends on body stature as well as age. On average the auricle reaches 85% of its final length by the age of 6 and 90% with 9. Later during life the length of the auricle increases only slowly – mainly due to changes of the soft tissue of the lobule which is more or less a lobule chalasis instead or real growth.

The width of the auricle also depends on body stature and age, but reaches 95% of its final value already by the age of 6.

In contrast the auricular projection, i.e. its width seen from a strictly anterior perspective, is almost constant

Table 1: Synopsis of the surgical options for correction of protruding ears

Malformation	Characteristics	Technique
Antihelix-hypoplasia	Cartilage: very soft	Suture (Mustarde)
	average	Suture and posterior scoring (Converse)
	hick	Suture and posterior + anterior scoring (Crikelair modif.)
Cavum-hyperplasia	High antihelix	Cavum rotation
Protruding lobule	Prominent helical rim Soft tisue malpositioning	Scoring Suture

throughout life. On average the ear projection is 20 ± 4 mm. Its normal range is between 12 and 28 mm. These data are important for the indication and planning of otoplasty.

4.2 Classification and surgery of auricular dysplasia

4.2.1 History

The history of the treatment of external ear malformations is closely linked to the reconstruction of traumatic or oncologic defects. Celsus was probably the first who described the reconstruction of partial auricular defects (Zeis, 1863, quoted from Weerda [47]). Known, however, are the statements of Susruta (about 4th century AD), who described a technique for reconstruction of the lobule from skin of the cheek. In medieval Europe (15th century) the Sicilian Branca family was well-known for their techniques to reconstruct noses and ears with flaps of skin from the upper arm. The famous Italian surgeon Gaspare Tagliacozzi (1545–1599) also described techniques for the partial replacement of an ear. However, he deserves the major credit for having his methods for the first time also illustrates pictorially.

Until well into the 19th century reconstructions for complete replacement of an ear were considered impossible – they were frowned upon. Also, one of the forefathers of the German plastic surgery, the famous Berlin surgeon Dieffenbach, described only techniques for reconstruction of partial auricular defects. At the same time he resolutely rejected methods for complete auricular reconstruction. From about the mid-20th century, there are detailed reports of total auricular reconstruction. First and foremost must be mentioned here the pioneering work of Tanzer [44], [42], Converse [7], [6], Brent [3], [2], Nagata [25], [26], [27], [28] and Weerda [47].

According to the increasing deformity severity and therefrom in the same direction resulting increase in the reconstructive effort, the auricular dysplasias can be divided into 3 levels of severity according to recommendations of Weerda [47].

4.2.2 1st degree dysplasia

These dysplasias are only minor malformations. All structures of the auricle are present. Surgical correction means realignment of the given tissue without the necessity for grafts.

4.2.2.1 Protruding ears

It is characterized by a flat to complete absence of the antihelix, including the crura. Usually there is also a pseudo-concha hyperplasia and an enlarged helical-mastoid distance of more than 20 mm.

Despite the relatively mild malformation, since the first description of an otoplasty by Ely, 1881 (quoted from Weerda [47]) until today approximately 100 additional possibilities for the correction of prominent ears have been described. For which of the correction options you choose in each individual case depends on the individual underlying pathology, from your "surgical school" and not the least of experiences and preferences of the attending surgeon. Our recommendations on the use of different techniques depending on the underlying pathology are summarized in Table 1.

Only in very thin and soft cartilage, we use the Mustarde technique [24]. Starting from a posterior skin incision, approximately 1 cm below and parallel to the helix, in this case, first the entire cartilaginous auricular surface is exposed while sparing the perichondrium. With thin needles the scapha and the junction of the concha and antihelix is marked. With multiple mattress sutures – we use Ethibond 4-0 for this – the ear is then folded in the previously needle marks. If necessary, a conchal setback and a lobule plasty can also be performed (see below).

For thicker cartilage, we use the Converse technique [8], [6] (Figure 6). Access, preparation of the cartilaginous auricular surface and markings are equivalent to the above outlined technique of Mustarde. The posterior surface of the ear cartilage is then incised where the markings were made, but under strict preservation of ventral perichondrium. The incisions are at the base of the newly defined anthelix respectively. Shaping a harmonious running antihelix is again with several mattress sutures. In addition, if necessary, a conchal setback and lobule plasty might be performed (see below).
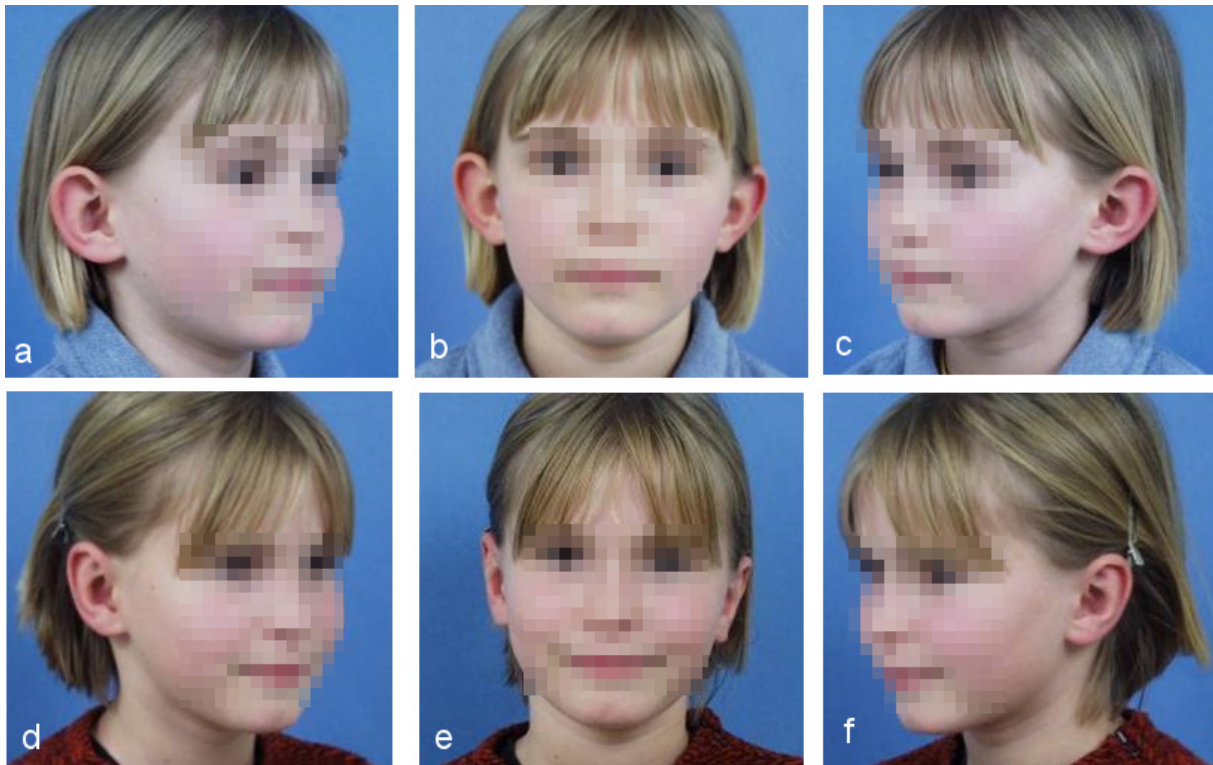
Figure 6: Protruding ears
a–c: Pre, d–f: post ostoplasty

In cases of very rigid and solid cartilage, but also in revisions, we apply a modified Chongchet or Crikelair technique [4], [10]. Unlike the techniques described above, here the ear cartilage is severed in the region scapha and then prepared anteriorly. By anterior scoring the cartilage (not deeper than 2/3 of cartilage thickness) in the running direction of the marked antihelix, a harmonious shape is formed. If the effect should not be sufficient, the Mustarde technique can be introduced with some supportive mattress sutures appropriately. The chondro-cutaneous flap is moved back and the cartilage reattached in the area of the severed scapha with some slowly absorbable sutures. Conchal setback and lobule plasty can be performed when needed.

This technique follows the same principle as the Stenström technique [41] with the difference that in the latter the skin over the antihelix is only tunneled and anterior surface of the cartilage scored with small special instruments (e.g. after Drommer, Fa Robomed, Kolbingen).

A conchal setback [14], [13] is indicated when the helix-mastoid distance is even after following one of the featured antihelix plasties more than 20 mm. Its operational technical basis consists of the resection of retroauricular fat, muscle and connective tissue and the fixation of the concha to the periosteum of the mastoid plane with stitches. Usually we put 1–3 slowly absorbable suture (e.g. Polysorb 4-0).

A real hyperplasia of the conchal cavum (usually pseudohyperplasia) shall be corrected by a sickle- or crescent-shaped cartilage excision from the concha [12] via the usual retroauricular access.

The lobule plasty stands at the end of nearly all otoplasties for protuding ears. Only in a few cases can be dispensed with. In addition to reversed Y-shaped skin excisions of the posterior lobule-concha area and scoring of the cauda helixis, we would like first and foremost a very simple yet highly effective technique for changing the position of the lobule [32], [34]. Here, the lobule is mobilized subcutaneously, starting from the retroauricular incision. The predefined highest point of the lobule is taken by a subcutaneous suture and anchored to the cavum conchae in the anterior region. With the so defined mattress suture the earlobe can be very finely dosed moved towards the head. We are using a slowly absorbable suture (PDS®) 5-0 for this.

For all protruding ears the retroauricular wound closure is usually performed in simply continuously technique. As suture material we use, in the sense of the often still small and partially uncooperative children, quickly absorbable suture (Vicryl rapid®) of 5-0. A bandage (Ototect®, from Spiggle&Theiss) remains for about 1 week. At night we recommend wearing a headband for an additional 6 weeks.

4.2.2.2 Macrotia

Macrotia denotes an auricle that is too large with regard to the patient's body stature. Often it is combined with abnormal protrusion. Since its predominant feature is the hypertrophic scapha the method of choice is the modified Gersuny technique (Figure 7).
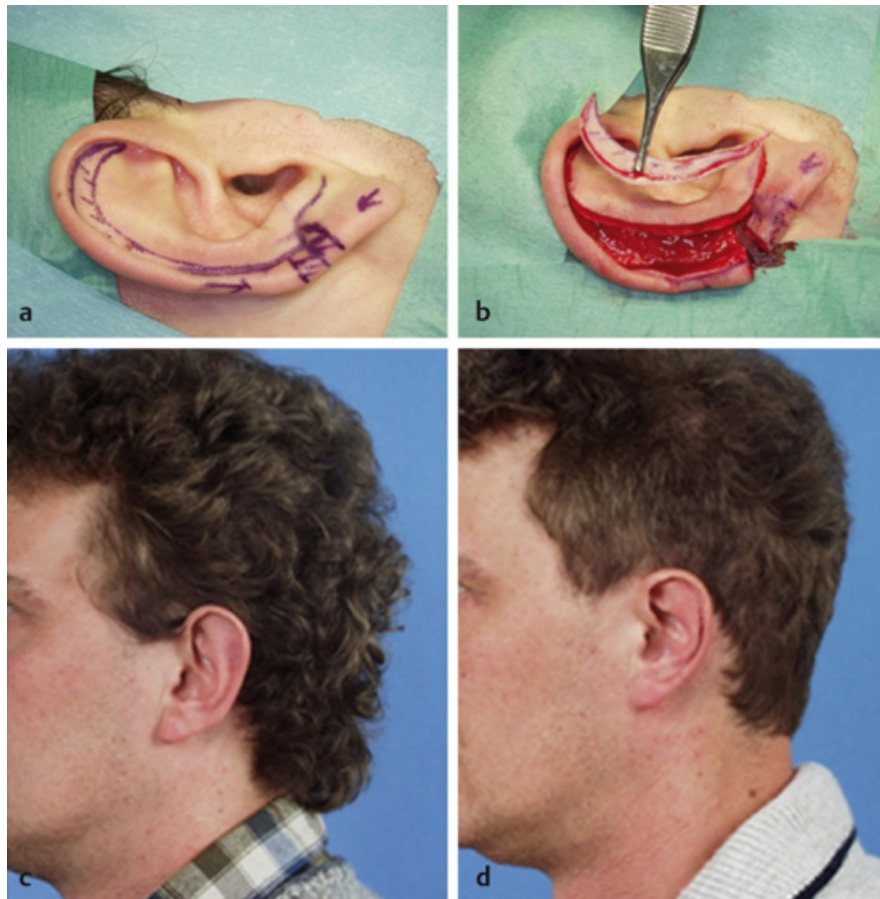
Figure 7: Macrotia
a: Markings for the modified Gersuny technique; b: Intraoperative situation with removed skin and cartilage; c: Situation pre and d: post OP

4.2.2.3 Cryptotia

Cryptotia (Figure 8) is relatively rare in Europe, but more common in Japan. It is characterized by the upper helical rim being present, but hidden under the scalp. The goal of its surgical correction is to increase projection of the upper part of the auricle and to construct the upper sulcus. This technique is similar to the 2nd step of total auricular reconstruction. The incision is done 1 cm above the hidden, but palpable heical rim and the skin mobilized above the level of the hair follicles. Then the hidden upper part of the auricular cartilage is mobilized leaving it cover with connective tissue for its nutritive blood supply. The retroauricular sulcus is covered with the prepared thin skin. An interesting alternative to using a full thickness skin graft is the application of a cranially pedicled island flap transposed into the new sulcus (Figure 8).

4.2.2.4 Mild cup ear deformity

The name cup ear deformity is used for a helical rim that "hangs over" the scaphoid fold. The superior crus might be hypoplastic whereas the inferior crus is normal in most cases.
If the helical overhang is very minor it can simply be resected, but this is rare.

The technique we typically choose for its correction is the one described by Tanzer [43]. It is an open approach exposing the whole malformed cartilage via a so-called "auricular degloving procedure". The malformed cartilage is then corrected by a Z-plasty constructing a harmonious helical rim (Figure 9). This procedure might be combined with an anthelixplasty.
Another technique was suggested by Musgrave [23]. He incised the helical rim several times in a radial direction, opens up the cartilage and stabilizes it with a cartilage graft from the conchal cavum (Figure 10).

4.2.2.5 Coloboma

Clefts or colobomas of the auricle are rare. They can be found in the area between the helical rim and the lobule (Figure 11). It can be corrected with a Z-plasty Weerda [47].

4.2.2.6 Appendages

Auricular appendages are relatively frequent. They are due to excess tissue derived from the first and second branchial arch. They are small in most cases, but they can be as large as a second auricle (Figure 12). They are excised and the wounds are closed meticulously.

Figure 8: Cryptotia
a: Markings of the subcutaneous island flap (1) and of the pedicled thin skin flap (2a) and deeper below the hairline (2b);
b: Intraoperative situation with mobilized flap (1) and (2); c: Situation pre and d: post OP
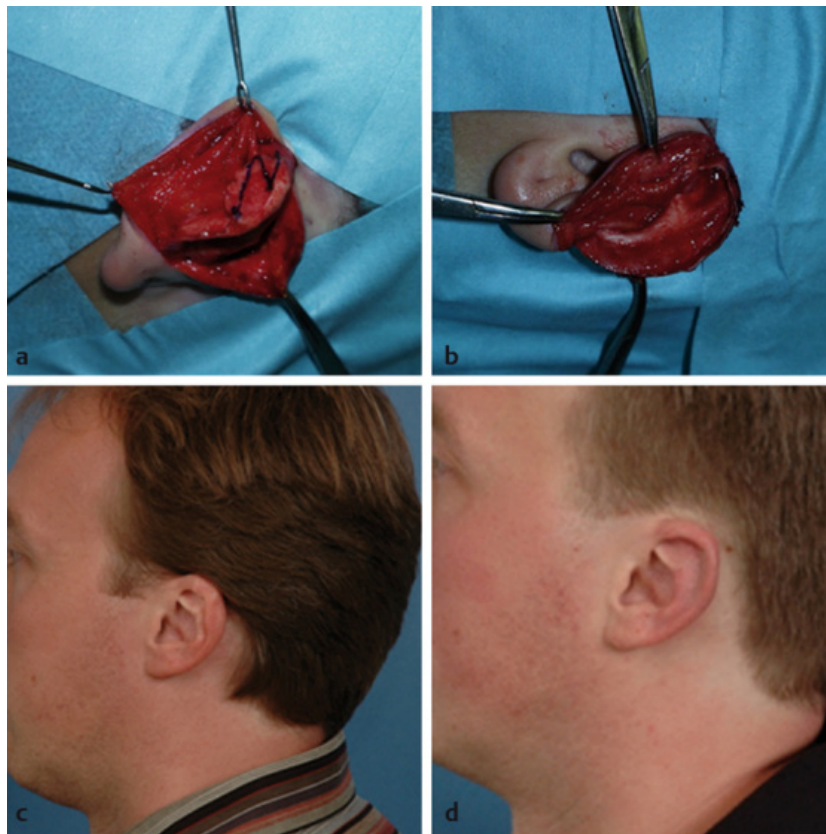

Figure 9: Cup ear deformity grade 1
a: Intraoperative situation after "auricular degloving" and markings of the cartilage plasty due to Müsebeck and Tanner;
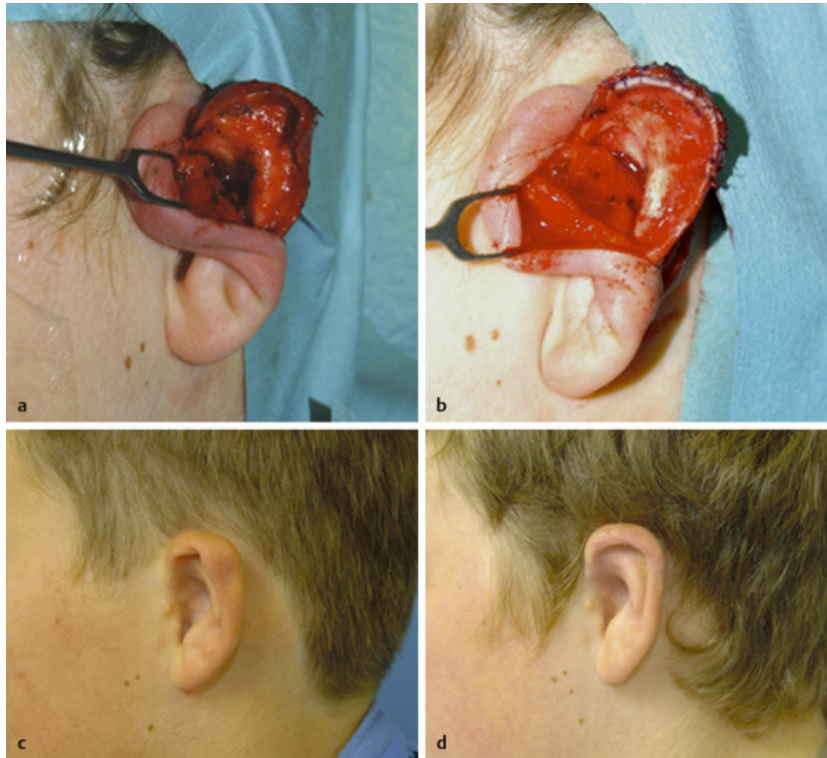b: Intraoperative situation; c: Situation pre and d: post OP

Figure 10: Cup ear deformity grade 1
a: Intraoperative situation after "auricular degloving"; b: Intraoperative situation after forming the helix with the Musgrave technique;
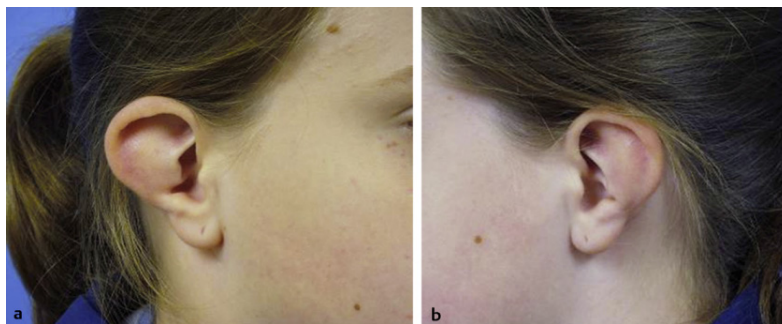c: Situation pre and d: post OP

Figure 11: Auricular colobomas

Figure 12: Huge ear appendages

4.2.2.7 Preauricular cysts and fistulas

Preauricular cysts and fistulas are mostly superficial and short, but can be very long going all the way to the skull base or run adjacent to the facial nerve. Meticulous excision taking care to avoid injury to the facial nerve is the method of choice. Intraoperative nerv monitoring might be helpful.

4.2.3 2nd degree dysplasia

These dysplasias are moderate malformation. Some parts of the auricle are normal, others have to be reconstructed. Often times they are combined with congenital atresia. Their reconstruction requires additional tissue, i.e. grafts of cartilage and skin. Although some parts of the auricle are present, in most cases we prefer a total auricular reconstruction (see below) to avoid steps in the esthetic units of the auricle. The existing structures, especially the skin, is used and integrated into the reconstructive concept. Typical examples are:

4.2.3.1 Severe cup ear deformity

In severe cup ear deformity the auricle is smaller than normal. In addition many auricles show a certain amount of dystopia mostly into ventro-caudal direction.

4.2.3.2 Mini-ear

The mini-ear is smaller than normal (Figure 13). Its structures can be almost normal or its relief might also be malformed.

4.2.4 3rd degree dysplasia

In 3rd degree dysplasia hardly any normal structures of the auricle are present. They can be subclassified into lobule-type, concha-type (Figure 14) and anotia. In most cases they are combined with congenital auricular atresia and in about 10% they are part of a syndrome, e.g. OAV-dysplasia or Franceschetti-Syndrome. In addition 18% of our patients have partial or total congenital facial paralysis (see below). Therefore our rehabilitation concept goes beyond the reconstruction of the auricle and includes treatment of hearing disorder, facial paralysis and skeletal anomalies.
We start reconstruction by the age of about 10 for two reasons:

1. For a complete framework we need sufficient and plenty of rib cartilage. By this age most children have a thoracic circumference of at least 63 cm. This is necessary to get enough cartilage of rib 6–9.
2. By this age they should be mature enough to understand the procedure and appreciate its benefits as well weight them against its morbidity. In this sense the doctor-patient relationship gets a different quality compared to treating little children.

Our surgical treatment is very standardized for the majority of cases. It includes main two operative steps mainly based on the technique described by Nagata [25], [26], [27], [28] (Figure 14), which we have further modified over the years, sometimes with some little refinements in addition.
On the day before surgery we make a template according to the contralateral healthy side in unilateral cases and according to general anthropometric data in bilateral cases. The planed position of the new auricle is than marked onto the skin.

4.2.4.1 1st step

This step includes the removal of rib cartilage normally from the ipsilateral side, the construction of the framework from the harvested rib cartilage in the technique described by Nagata, the implantation the framework with the help of the template and eventually the prefabricated external ear canal and tympanic membrane, depending on the CT-scan (CT-score after Siegert [40]).
Generally we work with 2 teams. One removes cartilage from the ipsilateral ribs 6–9 (Figure 14). In male patient the incision is above rib 7. In female patients we have described a special T-shaped incision 2 cm below the submammarian fold, that is excised in the second step and the skin transposed cranially, so that the final scar lies almost invisible exactly in the submammarian fold [20].
After removal of the cartilage the ribs are reconstructed by suturing resorbable nets (Vicryl®) onto the left in place inner perichondrium. These are filled with the otherwise unusable left over, little pieces of rib cartilage that are collected throughout the carving procedure [38]. We have been using this technique regularly since more than 7 years and could show, that stable regeneration of the ribs develop. On palpation the regenerated ribs are similar to the original or contralateral ribs.
The 2nd team creates a skin pocket in the auricular region and removes the malformed auricular cartilage. This team also carves and constructs the framework according to Nagata's suggestions. It is then transplanted to the planned position. The skin is draped over it and pulled into the relief with 2 suction drains.
If middle ear reconstruction is feasible an ear canal is fabricated made out of little pieces of rib cartilage around a Silastic cylinder. In addition the tympanic membrane is built out of the elastic malformed cartilage positioned into a special mold [31].

4.2.4.2 2nd step

Approximately 6 months after the first step auricular reconstruction is continued with the second step. It includes the elevation of the auricle and in appropriate cases also the so-called atresia operation, which is the creation of an external ear canal, tympanic membrane and creation of the sound conducting apparatus.

Figure 13: Dysplasia 2nd degree
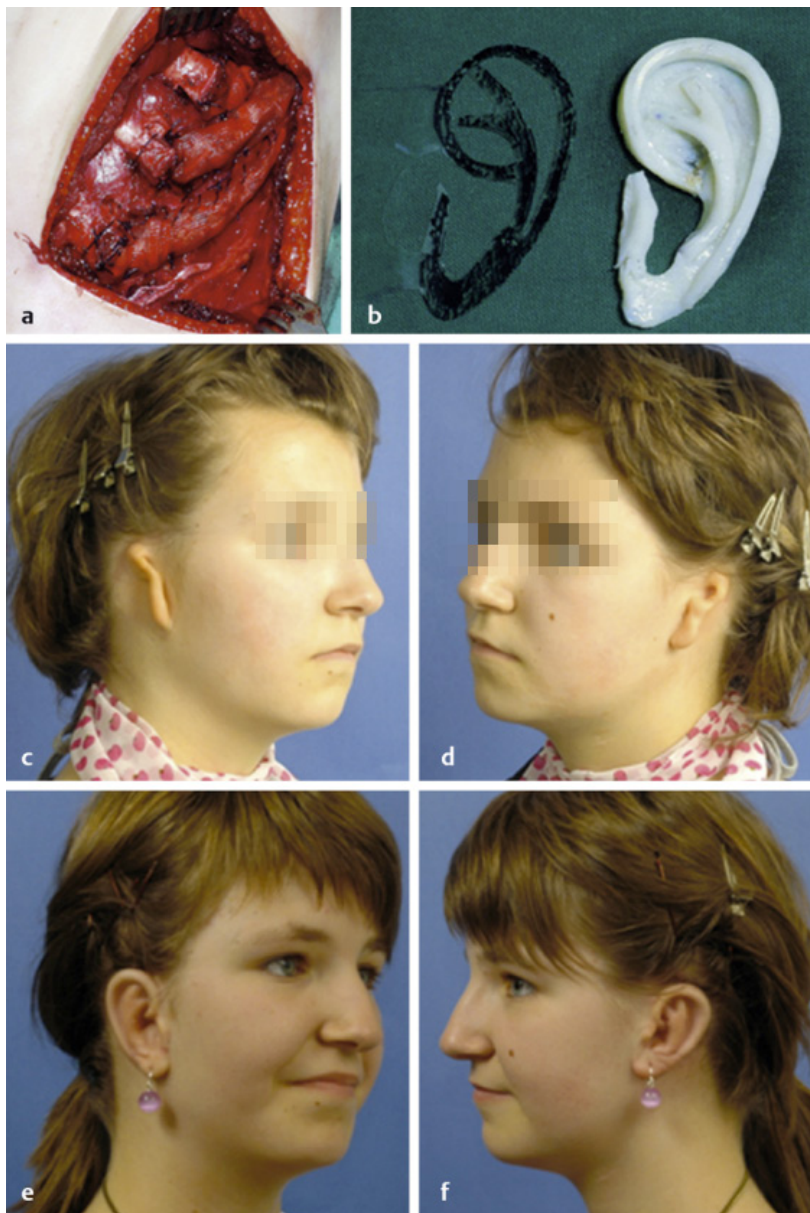a: Mini ear; b–d: Cup ear deformity 2nd degree

Figure 14: Microtia 3rd degree of the lobule type with congenital aural atresia
a: Intraoperative situation after harvesting the cartilage of ribs 6–9 and reconstruction of ribs 6–8; b: Preoperative made tremplate and auricular framework before implantation; c + d: Pat. with bilateral microtia 3rd degree pre and e + f: post OP

The incision is made 1 cm distant to the helical rim. The skin is elevated as a very thin flap leaving the hair follicles in place. Care is taking not to expose the cartilage. The whole framework remains covered with connective tissue. A highly vascularized, posteriorly pedicled flap of superficial temporalis fascia or SMAS is mobilized. After carving a crescent-shaped piece of cartilage used as a buttress for the elevation and sutured to the back of the base plate it covers the cartilage and is the base for the full thickness skin graft that covers the created retroauricular fold. The skin graft is harvested from the rib region. It includes the scar from rib harvesting, so that no additional scar is created.

To decrease the size of the retroauricular wound area a large transposition-rotation-flap from the neck is mobilized and transposed behind the framework. Then the remaining defect is covered with the skin graft. The wound is covered with a stable bandage using multiple tapes which are not removed before one week postoperatively.

4.2.4.3 3rd step

In the 3rd step refinements are performed. Remnants of the malformed pinna might have to be worked into the relief in a very meticulous way. Unwanted hair on the helix in patients with a low hair line might have to be removed. For epilation we predominantly use electroepilation, which still is the gold standard in epilation. Alternatively laser or intense pulsed light (IPL) might be used.

If the atresia operation has been performed in the 2nd step the implants are removed and the constructed external ear canal is covered with another full thickness skin graft.

In most cases these 3 operations result in good esthetic results. They can mimic normality, but due to different biophysical properties of rib cartilage they can never be completely normal. In some cases additional minor refinements can be suggested and further improve the esthetic outcome.

4.3 Future and alternatives to autogenous ear reconstruction

Tissue engineering is removed despite many laboratory tests still far from practical clinical use.

Alloplastic auricle frameworks have a long tradition. Many materials have been tried and abandoned again over the years due to poor long-term results. For several years, Medpor is specified as an alternative to costal cartilage. It is a porous polyethylene (Medpor®), into which tissue growth in [1]. The size of the pores determines thereby the ingrowth of tissue and its surface roughness. Here, large pores, into which tissue can grow well is associated with high roughness and vice versa.

The observation times of individual surgeons are now over 10 years [30]. Comparative clinical studies do not yet exist but they would also be difficult to achieve, according to scientific criteria. The extrusion rate of Medpor frameworks at experienced users are small, but long-term

considerations of the mostly young patients are not negligible. Important for trouble healing and resistance to microtrauma is a good coverage of the Medpor with superficial temporal fascia. But if it should come to the exposure of the implant due to injury or local infection, the risk of persistent infection and ultimately extrusion of the implant, in contrast to autogenous cartilage, is high. Although a final judgment based on the current data is not yet possible, we offer this option in special cases such as situation after or rejection of the rib cartilage harvesting. However, due to our experience alloplasts cannot replace the "gold standard" of autogenous reconstruction.

4.4 Hearing rehabilitation in congenital aural atresia

Due to their identical embryological roots severe external ear malformations are typically associated with congenital aural atresia with a sound conduction block (50–60 dB), while the inner ear function is usually normal. In bilateral atresia hearing rehabilitation is mandatory in the first few months of life. In unilateral malformation children have constraints of bilateral auditory function, i.e. the directional hearing and the hearing in noise. An adequate supply should be done from the first year of life.

In children up to about the age of two, first the supply of conventional hearing aids is indicated, which are worn on the head bands (Figure 15).

Figure 15: Pat. with conventional bone conduction device worn at a softband

From about the age of two partially implantable hearing aids are available. While the systems with open implants (BAHA, Ponto) are usually implanted not before preschool, we have developed a closed system (Sophono) that is used clinically since 2006 and has now been implanted in over 2,500 patients worldwide [33]. With the new minimally invasive implantation technique [36], the system can be used in virtually any bone thickness and
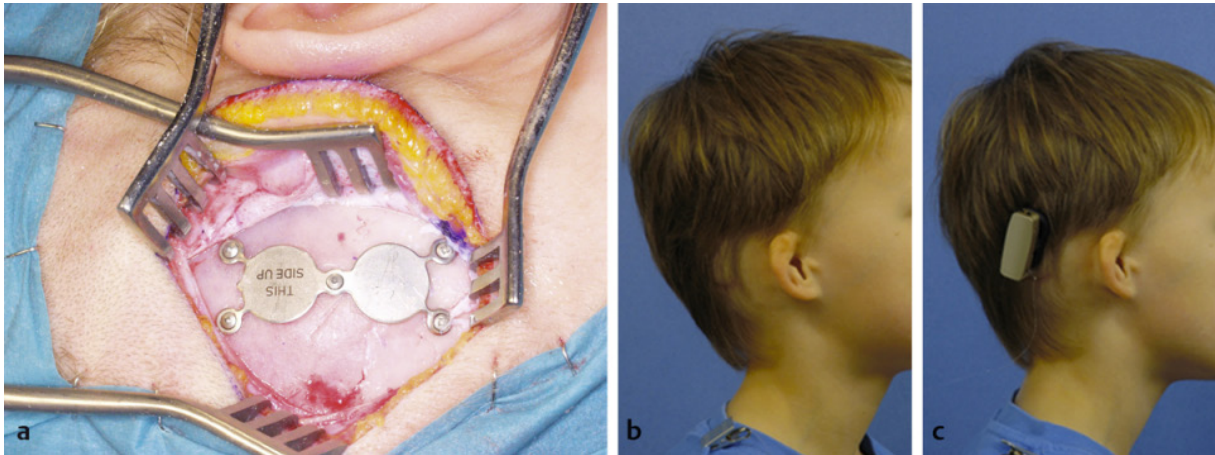
Figure 16: Partially implantable bone conduction device Sophone
a: Implanted, titanium welded double-magnet; b: Pat. after implantation of the magnets without open abutment; c: dito, with magnetically anchored device
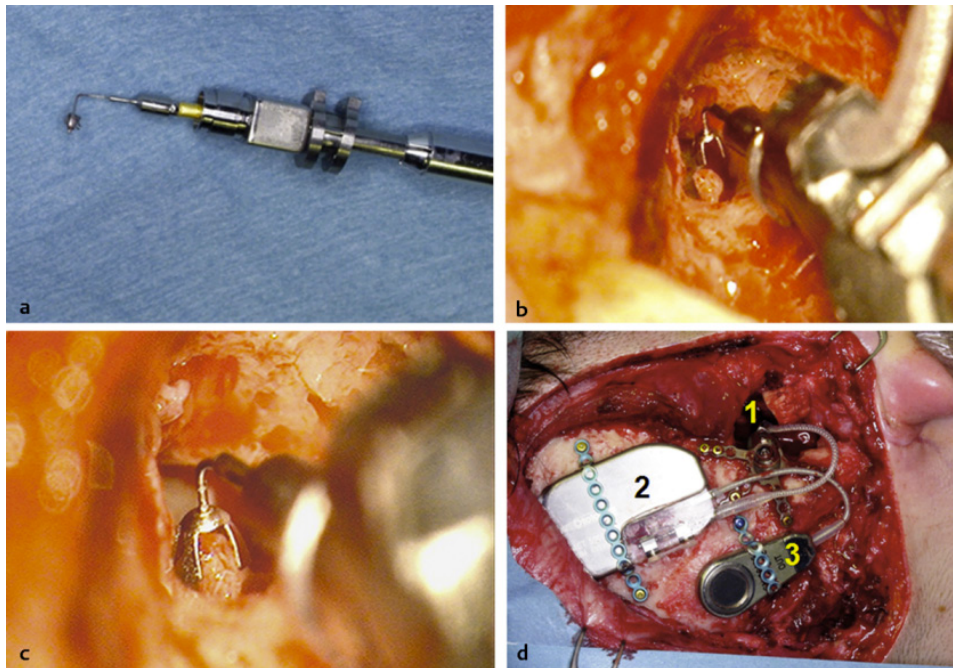

Figure 17: Fully implantable active device Carina
a: Modified transducer with prothesis; b: Intraoperative situation shortly before ...; c: ... and after positioning onto the stapes; d: System with transducer (1), electronics (2) and microphone (3)

therefore at any age. We have previously used it in children from the age of two [35]. The principle is that a titanium-welded double magnet is implanted into or onto the bone, which fixes a specific, externally worn bone conduction hearing aid on the scalp (Figure 16). With this transcutaneous coupling all biological and psychological problems of open implants in percutaneous systems can be bypassed.

When we carry out the complete ear reconstruction described above, from about the age of 10 on we estimate the chances of a middle ear reconstruction based on a temporal bone CT. Together with radiologists we have developed a grading system, with which we semi-quantitatively evaluate the relevance of the middle ear structures [40]. In good anatomical conditions we then combine the middle ear reconstruction with the staged ear reconstruction [31]. To avoid re-stenosis of the ear canal, we construct the middle ear structures also in three stages. In this case, the neo tympanic membrane and the external ear canal may be preformed as constructs and stored in a subcutaneous pocket in the first step. In the second step the actual tympanoplasty is performed and in the third step of the ear canal is opened and epithelialized.

From about the age of 15 active hearing devices may be implanted, where individual authors also report earlier implantations [29]. We had 2006 active systems modified for the first time that they were employed in atresia. Initially, only the partially implantable systems were available, then later we could also for the first time implant the fully implantable systems (Carina) (Figure 17, [39]). Their exceptional advantage is the full implantability and

thus its use in any life situation. Their disadvantages are the need of the regular battery charging, battery changing after a few years and the retroauricular implanted microphone with the lack of directional characteristics. Moreover, there was at least in the past, many technical problems which required premature implant replacement.

5 Facial musculature

5.1 Introduction

The congenital facial palsy is a severe disability with significant aesthetic, functional, and developmental effects. Even in the newborn it may lead to impairment of vision due to reduced closing of the eyes and disorders of the tear flow and of eating and drinking by restricted mouth closure. Later, it has an impact on the language and the facial expressions and so on to the non-verbal communication.

The incidence of congenital facial palsy is estimated to be about 2%. Their predominant cause is viewed with more than 80% as a birth trauma.

5.2 Diagnosis

The diagnosis is first and foremost a clinical one. A distinction is made between partial or complete facial palsy, unilateral or bilateral, developmental or traumatic birth, isolated facial paresis or paralysis as part of a syndrome possibly with other cranial nerves failures.

If there are no accompanying symptoms to elicit and if it is a "heavy" birth a birth traumatic etiology is assumed. It is postulated that the facial nerve is compressed in particular in its vertical course in the bone by pressure on the head of the newborn in the pelvis of the mother during the birth process and it could lead to a neurapraxia. In these cases, initially be awaited. If there is no spontaneous remission in the first months of life as it is at 90% of congenital facial palsy, so – but only later – further investigations such as EMG, ENOG, MRI and CT are indicated.

Syndromic facial paralysis usually occur unilaterally as part of branchial arch malformations usually in the context of hemifacialen mikrosomia together with auricular, middle ear and lower jaw malformations. Bilateral congenital facial palsy can be found most often in the Moebius sequence with an the incidence of 0.0002–0.002% [18]. In his first description Möbius [22] named only the combination of paresis of the VI. and seventh cranial nerves. Today we no longer speak of a syndrome, but of a sequence and count combinations of functional disorders of the nervs mentioned with those of Nn. trigeminal, glossopharyngeal, vagus, hypoglossal and accessorius. A genetic cause is believed only in about 2% of the patients and individual genetic loci for this sequence have now been found. Both toxic and vascular causes are assumed for the non-genetic Möbius forms.

Other rare syndromes associated with facial paralysis are CHARGE, DiGeorge, Poland, Albers-Schoenberg syndrome, trisomy 18 and 13. Infections such as polio, mononucleosis, varicella, otitis media, or meningitis can also lead to early childhood facial palsy, but can also occur later in life as well.

5.3 Surgery

Since in congenital facial palsy not only the function of the nerve is restricted, but also the target organ, the muscles of facial expression, could not develop, both muscles and a functional neural stimulation is needed for an optimal reconstruction. Harii described in 1985 a two-stage technique that still today is the standard procedure in unilateral congenital or long-standing facial paralysis with atrophic muscles (Figure 18).

In the first step, a nerve, preferably the sural nerve is removed and anastomosed to a buccal branch of the facial nerve on the unaffected side. It is important to identify at least two branches, which target regions overlap each other, so that one can be sacrificed without inducing paresis on the unaffected side. The transplanted nerve is then passed through the upper lip to the pretrageal area of the contralateral side and labeled with a non-absorbable suture.

The reinnervation of the transplanted nerve to the opposite side may well take up to 9 months. This is clinically tested by the "Tinel's sign" [46], in which one lightly taps on the transplanted nerves to the skin. If the patient then notices paresthesias in the end region of the transplanted nerve, so on the affected side, the "Tinel's sign" is positive and indicates that the axons have grown through the graft and the second step can take place.

Here, a microvascular and neuro pedicled muscle is removed. Harii recommended the use of the gracilis muscle, which we prefer too. Other authors such as Harrison [17] use the pectoralis minor muscle, but its vascular and neural stem is significantly shorter and according to our own experiences therefore less well suited. After the end of the transplanted nerve has been exposed, the muscle is partially digested and fixed in the upper and lower lip of the paralyzed side of the face (Figure 18). In addition, narrow portions of the muscle can be sutured into the cheek and eyelid. The origin of the muscle is fixed to the zygomatic body and arch or at the preauricular fascia under slight tension. Then the microvascular re-anastomosis of the muscle preferably takes place at the facial artery and vein and the neural re-anastomosis between the grafted sural nerve and the anterior branch of the obturator nerve, the motor branch to the gracilis muscle. By this technique, a relatively natural facial feature can be achieved (Figure 19).

In bilateral facial palsy is no contralateral branch available, so that only the connection on a different cranial nerve comes into question. For this purpose Zuker and Manktelow recommended the masseteric nerve of the trigeminal nerve [48], [21]. Since in these cases no cross-nerve technique is required or even possible at all, these
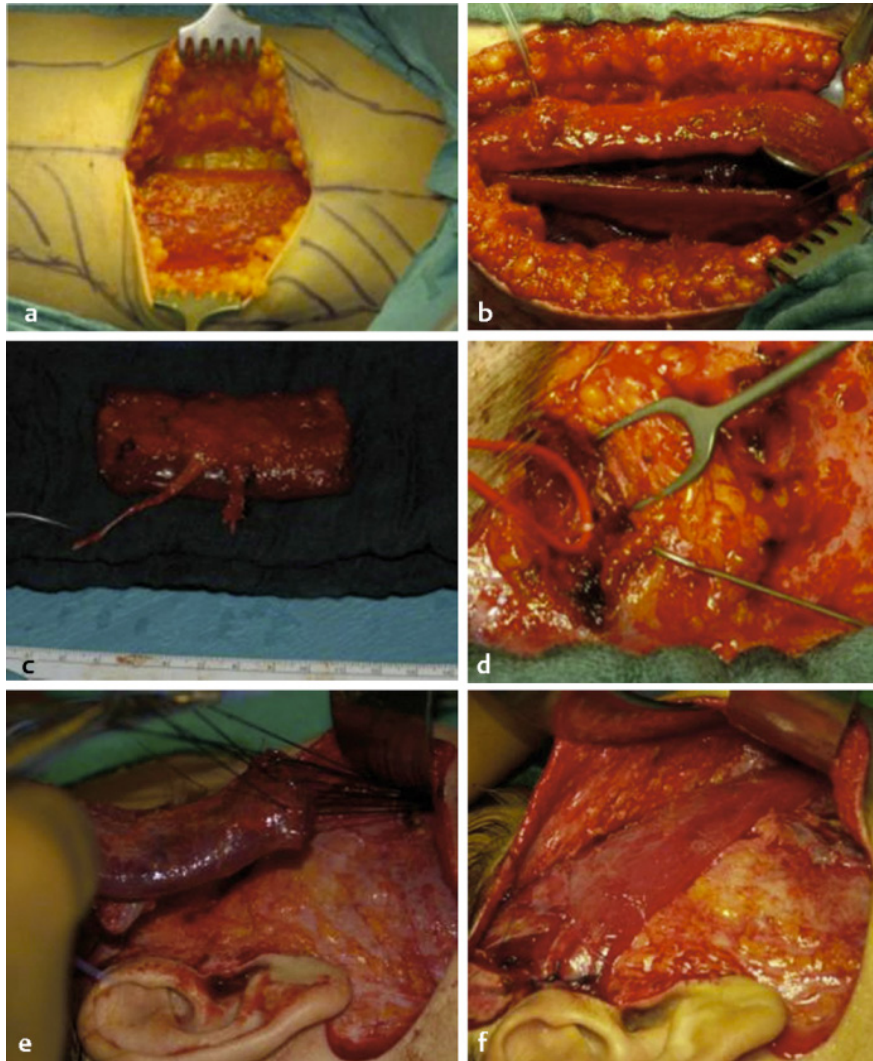
Figure 18: Microsurgical transfer of a segment of the M. and neurotisation with the R. massetericus of N.V.
a: M. gracilis; b: Separation of the muscle; c: Free muscle graft; d: R. massetericus, loosely thightened with a vessel loop;
e: Transport of the muscle graft into the face; f: Situation after reanastomosis of the microvascular (*) and microneural (arrow) anastomosis

Figure 19: Pat. with Möbius sequence
a: Preop. relaxed; b: Preop. with maximum mimic; c: 6 months after bilateral M. gracilis transplantation, relaxed; d: Symmetric smile

procedures are performed done in one stage, but separately for each side.

Although in these patients, the stimulation of the graft for facial muscle rehabilitation is not done by nerves designed for facial movements, but rather by a branch of the fifth cranial nerve for chewing, an amazingly precise function of facial expressions can be achieved due to the enormous plasticity of the brain in children and adolescents.

Notes

Competing interests

The corresponding author is inventor, shareholder and medical director of Sophono Inc., Boulder, USA.

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