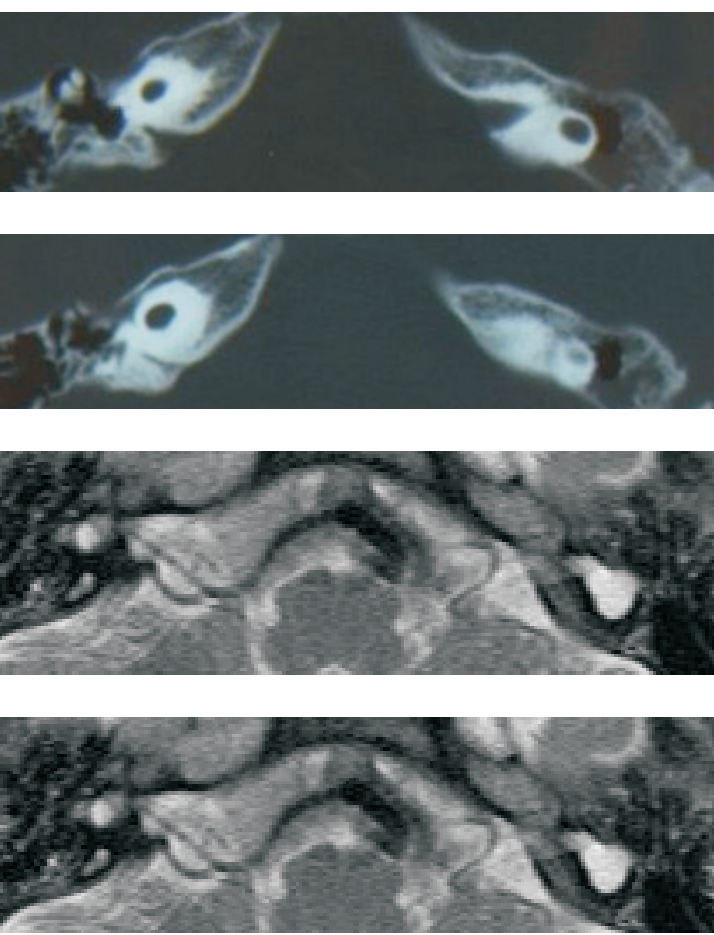


COCHLEAR IMPLANTATION IN MALFORMED INNER EAR

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Introduction

According to the functional results cochlear implantation (CI) nowadays is a method of choice in the management of congenital or acquired deafness. There are many factors that may influence the outcome of CI (Profant, 2004). One of the most influential factors is the anatomical integrity of the inner ear. Differentiation of bony labyrinth, shape of the internal acoustic meatus (IAM), size and position of the 8th nerve are the information to be evaluated by imaging in each CI candidate. In the majority of CI candidates the deafness is caused by the malfunction of the organ of Corti in anatomically differentiated inner ear with preserved ganglion cells and nerve fibers. In these patients good results can be expected if implanted in the appropriate age. In children with malformed inner ear the preoperative situation is different. Only the shape of malformed inner ear is to our disposal, there is no information on the number of hair cells, ganglion cells or nerve fibers. The number of patients with inner ear malformations is limited and all the centers should bring information on these patients to reach global experience for recommendations and counseling parents of patients with malformed inner ear. In this paper we present our experience with cochlear implantation in patients with malformed inner ear.



Inner ear malformations

Classification of the inner ear malformation is based on the developmental anatomy of the inner ear as described by Jackler (1987). This classification is focused on the particular parts of inner ear and it recognizes terms as hypoplasia, dysplasia and aplasia. In the developmental disorders of membranous labyrinth this classification differs from among the histopathological changes, while in the bony labyrinth developmental disorders this classification is based on the radiologic image of malformation. The stage of malformation corresponds with the stage of the embryological development of the inner ear.

Classification of inner ear malformation

- Malformations of membranous labyrinth
 - Complete dysplasia of membranous labyrinth (Siebenmann-Bing)
 - Partial dysplasia of membranous labyrinth
 - Cochleo-saccular dysplasia (Scheibe)
 - Dysplasia of the basal turn of cochlea (Alexander)
- Malformations of bony and membranous labyrinth
 - Complete labyrinthine aplasia (Michel)
 - Cochlear abnormality
 - Cochlear aplasia
 - Cochlear hypoplasia
 - Incomplete partition (Mondini)
 - Common cavity
 - Labyrinthine anomaly
 - Semicircular canal dysplasia
 - Semicircular canal aplasia
 - Abnormality of aqueducts
 - Dilated vestibular aqueduct
 - Dilated cochlear aqueduct
 - Abnormality of internal meatus (IAM)



Material and Methods

In the period 1994 – 2007 there have been 187 patients implanted in our department. There were 123 children (111 children with prelingual deafness, 5 children with perilingual deafness and 5 children with postlingual deafness). In 4 children with congenital deafness malformation of the inner ear was found (3,6%): **Bilateral common cavity in one child (0,9%)**
Narrow internal auditory meatus in two children (1,8%)
Bilateral aplasia of lateral semicircular canal (0,9%)

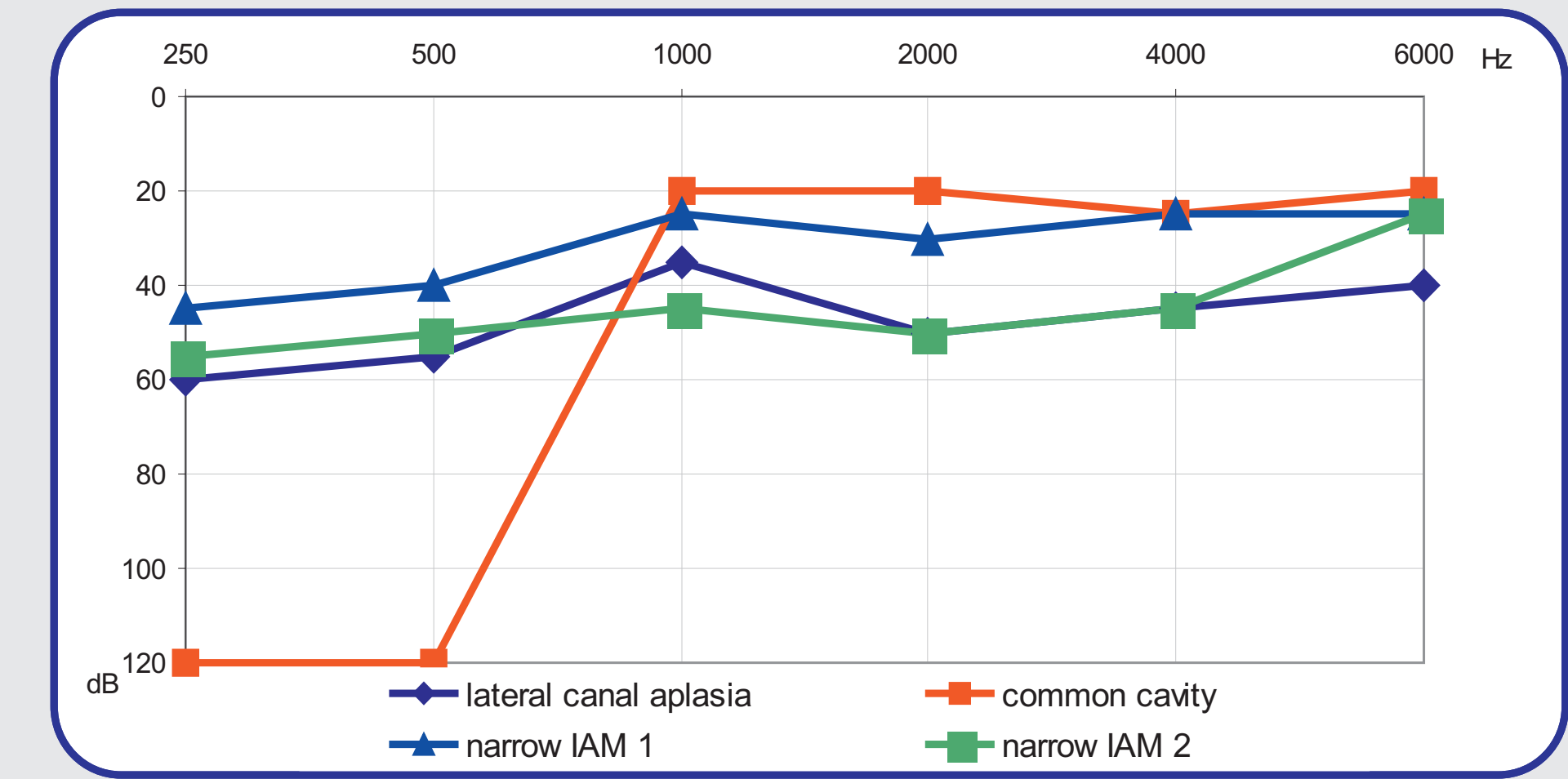
The youngest child was 2,2 years old at the time of implantation, the oldest child was 4 at the time of implantation. Three children were implanted with Nucleus Freedom standard electrode device, the child with common cavity was implanted with Medel C40+S device with short electrode array. All children were operated by one surgeon with standard approach via mastoidectomy, posterior tympanotomy and cochleostomy.

Table 1 Structure of patients and inner ear malformations

	Number of children	Gender	Reason of deafness	Age at the time of CI (Years)	Device	Implant user in time of evaluation (years)
Common cavity	1	F	Unknown	2,5	Medel C40+S	2
Narrow acoustic meatus	2	F	Unknown	2,2 3,3	Nucleus Freedom CI24RE (CA)	3 6
Lateral semicircular canal aplasia	1	M	Unknown	4	Nucleus Freedom CI24RE (CA)	0,5

The following tests have been done to evaluate the functional outcome of cochlear implantation:

PTA in free field (warble tone) with CI
Speech audiometry in free field
Category of auditory performances (CAP)
Evaluation of spontaneous speech production
Evaluation of implantee's speech understanding



Results

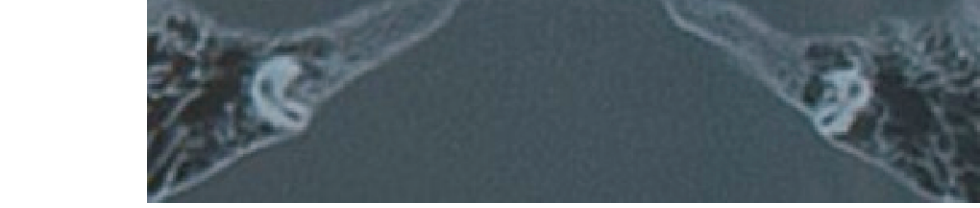
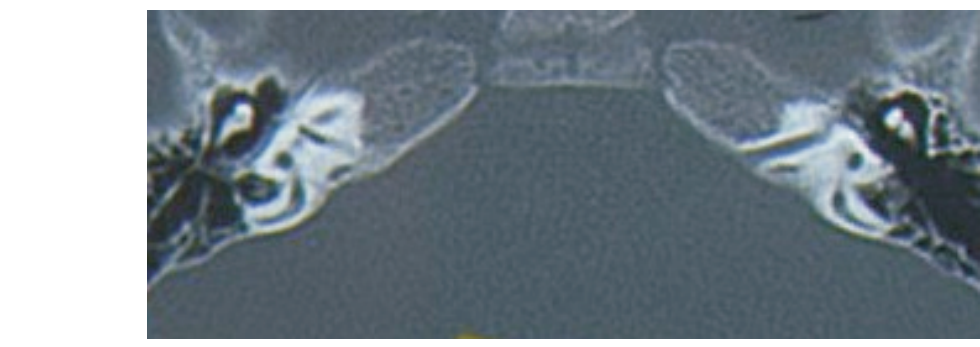
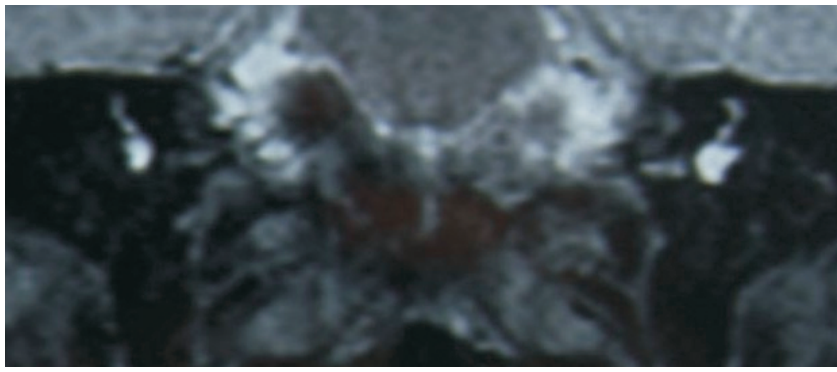
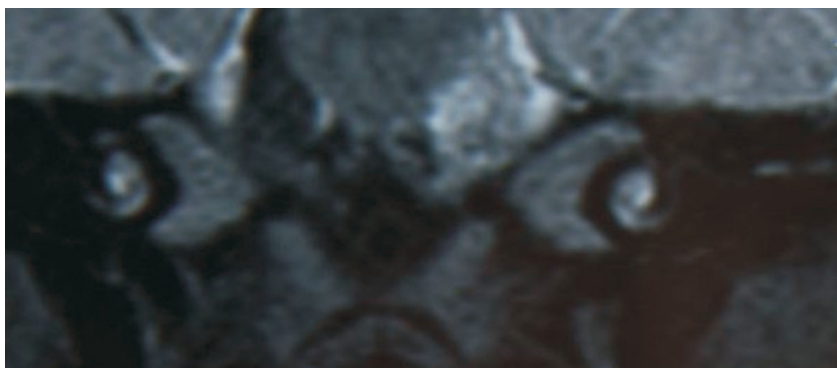
Hearing tests

Before CI there was some residual hearing for warble tone in low frequencies in a child with lateral semicircular canal aplasia (normal radiologic image of cochlea). In all other children there was no hearing before implantation. After CI in all children reaction to sound was detected. Child with common cavity has no perception in low frequencies while the PTA in middle and high frequencies can be detected at 20-25 dB. In all other children the PTA threshold in all frequencies is better than 50-60 dB. Free field PTA tests with warble tone are shown in fig. 1

Malformations of membranous labyrinth are usually histo-pathological findings in the targeted study of the temporal bone. Malformation of bony labyrinth can be detected by imaging. High resolution CT and MRI give detailed information sufficient for clinical decisions. The hearing in these patients can vary from deafness to sensorineural hearing loss (usable residual hearing). Progression of hearing loss may develop spontaneously or it may be triggered by trauma, virosis etc. Some papers stress the domination of MRI over the CT (Parry et al., 2005). Sennaroglu et al. (2002) and Zheng et al. (2002) suggested even more detailed classification which was not generally accepted.

Most frequently the cochlear malformation is diagnosed. Majority of the cochlear malformation is caused by interruption of cochlear development during embryogenesis. This opinion is supported by the shape of the malformed cochlea that reminds the cochlea in different embryological stages (Jackler et al., 1987). One can assume earlier the development is interrupted more severe developmental and functional changes will appear. Jackler estimated inner ear malformation incidence:

Incomplete partition (Mondini)	55%
Common cavity	26%
Cochlear hypoplasia	15%
Cochlear aplasia	3%



In speech audiometry tests (pediatric speech audiometry) no child can repeat a single word without lip-reading.

In evaluation of auditory performances (CAP) all children show perception of environmental sounds. Child with lateral canal aplasia 6 months after CI reacts to the speech sounds and 2 children with narrow internal acoustic meatus 3 years after CI can repeat some single words without lip-reading (Table 2)

Table 2 Category of Auditory Performances (CAP) in children with

Category of Auditory Performances		Malformations and results
0	No environmental sound detection	0
1	Detection of environmental sounds	1 Child with common cavity
2	Reaction to speech sound	1 Child with lateral canal aplasia
3	Identification of environmental sounds	0
4	Differentiation of sound speech without lip-reading	2 Children with narrow IAM
5	Understanding of everyday sentences without lip-reading	0
6	Understanding of speech without lip-reading	0
7	Telephoning	0

Speech evaluation

There are significant differences in spontaneous communication among the children of this group from meaning vocalization (child with common cavity) to creating simple 3-word sentences (children with narrow IAM) (Table 3)

Table 3 Spontaneous speech evaluation in children with inner ear

Evaluation of spontaneous speech		Malformations
0	No speech	0
1	Meaning vocalization	1 Child with common cavity
2	Using several simple words	1 Child with lateral canal aplasia
3	One-word sentences	0
4	Two-word sentences	1 Children with narrow IAM
5	Three-words sentences (subject-predicate-object)	1 Children with narrow IAM
6	Multword sentences with solecism	0
7	Multword sentences and simple compound sentences with Sporadic solecism	0
8	Full spontaneous communication	0

The understandability of speech of 2 children with narrow IAM, (active vocabulary, creating also simple sentences) is very limited for standard listener. This kind of speech is understandable for those who are experienced with communication with deaf people (parents, relatives, teachers of the deaf). The speech of other children from our series is not understandable (Table 4).

Table 4 Evaluation of speech understandability in children after CI in malformed inner ear

Understandability of speech	Malformation
0 Impossible to evaluate speech, vocabulary is missing	0
1 Non understandable speech	1 Child with common cavity 1 Child with lateral canal aplasia
2 Partially understandable speech, speaker must complete information also nonverbally	2 Children with narrow IAM
3 Understandable speech if the listener knows the context	0
4 Understandable speech if the listener is experienced in communication with deaf	0
5 Speech understandable to majority of listeners	0
6 Speech understandable to all listeners	0

In all children after the first fitting there was a progress in sound detection.

Surgical results

The electrode array was implanted in a child with lateral semicircular canal aplasia and children with narrow IAM by standard approach via mastoidectomy, posterior tympanotomy and cochleostomy with full insertion. In one of these children the stapedial reflex was missing (with normal impedances). In a child with common cavity the incus and bony bridge and finally posterior ear canal bony wall were removed temporarily to improve access to the cochleostomy site. There was deep drilling 4-5mm to open the common cavity followed by perilymphatic gusher. 2 mm were missing to the full insertion of the short version of electrode array. At the end of the procedure the posterior canal wall was fixed to its original position and cochleostomy was packed with fat. Because of liquorrhea the revision surgery was done 9 days after the surgery to pack completely middle ear cavity with fascia, muscle tissue and fibrin glue. There was no liquorrhea after revision surgery.

Discussion

Inner ear malformation is one of the reasons of congenital deafness. Jansen (1969) found inner ear malformation in about 20% of children with congenital deafness. This seems to be too high incidence. In the series of deaf children in our center we found inner ear malformation in about 5% of our deaf children. Wu et al. (2005) found 38% of malformation in the series of 160 deaf children. London et al. (2005) found 6,9% inner ear malformation in the series of 260 implanted children.

The high resolution CT gives a perfect image of bony inner ear structures. Experienced otologist and radiologist can make diagnosis of inner ear malformation very easily. MRI imaging is regularly done in all patients with inner ear malformation (some centers do MRI imaging in all CI candidates). Especially in cases with narrow IAM the information on presence and size of the acoustic nerve is of utmost importance.

Recently an information on differentiation among the stages of hypoplasia – aplasia of the cochlear nerve appeared in literature. (Zanetti et al. 2006, Govaerts et al. 2003, Casselman et al. 1997). For children with cochlear nerve aplasia brainstem implantation is the option (Colletti et al., 2002).

Special category of inner ear malformation is lateral semicircular canal aplasia. Johnson et al. (2000) found in 15 patients (2 with unilateral aplasia) all kinds of hearing loss from the conductive to the sensorineural deafness.

Cochlear malformation is a predisposition for meningitis development either spontaneous or after cochlear implantation. Spontaneous communication of inner ear liquids with CSF and communication of perilymphatic space with middle ear cavity through cochleostomy are clear etiopathogenetic factors. Immunization against meningitis is generally recommended before cochlear implantation, especially in children with inner ear malformation.

Cochlear implantation in malformed inner ear requires experienced CI surgeon who knows all available techniques to insert electrode array into the malformed inner ear. The position of the facial nerve should be identified on the CT scan. Since there might be a higher incidence of facial canal abnormalities the facial nerve monitoring is recommended (Graham et al. 2000). Mylanus et al. (2004) found aberrant facial nerve in 17% of children with inner ear malformation. There was no anatomical variation of the facial canal in our series, there was no facial nerve impairment after CI in our patients.

There was a massive perilymphatic gusher in patient with common cavity malformation. Packing of cochleostomy with electrode array and fat stopped the gusher at the end of the surgery. The liquorrhea has appeared next day with fluctuating intensity. After 9 days the ear has been revised with packing middle ear cavity with muscle and fibrin glue. Luntz et al (1997) found controlled perilymphatic gusher in several patients of her series.

All kinds of electrodes (standard, short, split electrode) should be available when the CI in malformed inner ear is planned. Electrodes with preformed shape are not suitable for malformed inner ear. Fishman et al. (2003) recommend fluoroscopically assisted insertion for accurate placement of electrodes in the malformed cochlea.

After CI in all our children with inner ear malformation the reaction to sound stimulation was recorded. Nevertheless, no child reaches results similar to average results of implantees with normal bony inner ear. The worst result was found in a child with common cavity (evaluated 2 years after CI). This child can detect the environmental sounds while average children of the same age can understand everyday sentences without lip-reading. Better improvement in speech perception than in speech production can be expected in this child in the future.

The best results can be expected in a boy with lateral semicircular canal aplasia. Now he is using the implant for several months only, he adequately reacts to sounds, to the speech stimuli and he starts to repeat single words.

Two children with narrow IAM use their implants for 3 and 6 years. They do not reach the level as other implanted children of the same age. While children without malformation understand the speech without lip-reading and speak in complex sentences, the children with narrow IAM speak in 1-2 words sentences, they can repeat only single words without lip-reading and their speech is similar to the speech of deaf children without CI.

The functional results depend also on the stage of the malformation. More severe the malformation is, poorer results can be expected. Malformed cochlea and narrow IAM (hypoplasia-aplasia of the cochlear nerve) do not allow transferring sufficient amount of information to the CNS. This fact must be thoroughly discussed with parents to have realistic expectation of functional outcome of cochlear implantation in children with malformed inner ear.

Arnoldner et al. (2004) referred on 6 children with moderate malformations (incomplete partition, hypoplasia, and unexpected gusher during surgery) with good results similar to children with normal bony cochlea. Despite this fact they recommend thorough counseling. Luntz et al. (2007) present doubtful results in children with common cavity while in children with incomplete partition they reach good results comparable to children with normal bony cochlea (identification of sound, scoring in open sets).

Papsin (2005) in the series of 298 implanted children found 38% of children with malformed inner ear. With lower dynamic range and prolonged stimulation time they reached stimulating results in children with common cavity and cochlear hypoplasia. The indication and results in children with narrow IAM they consider being problematic. Stimulating results in children with incomplete partition are presented also by Wermeskerken et al. (2007).

Conclusion

Malformation of the inner ear with present cochlear nerve is not a contraindication to cochlear implantation nowadays. On the other hand the parents of children with malformed inner ear should be clearly informed that even if implanted in the ideal age (1-3y) the results will not reach the level of the deaf children of similar age with normal bony inner ear. For CI candidate selection in case of malformed inner ear each center should keep following principles:

- To perform CT and MRI in all cases
- Absence of cochlear nerve (confirmed by MRI) is contraindication to cochlear implantation
- Counseling with parents must be open with all information on results, complications as well as on other possibilities to educate the deaf child.
- Child should be immunized against meningitis before cochlear implantation
- Surgery must be done by experienced surgeon who has all kinds of electrodes to his/her disposal
- Implanted child with inner ear malformation requires more attention in the postoperative period with intensive rehabilitation

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