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Spectrum of Hearing Disorders and Their Management in Children With CHARGE Syndrome

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Objective: The CHARGE syndrome is associated with ear anomalies and deafness in addition to other malformations. Deformations of the ossicles or aplasia of the semicircular canals, cochlear hypoplasia, hypoplasia or aplasia of the VIIIth cranial nerve and abnormal routing of the VIIth cranial nerve, sigmoid sinus, and emissaries are typical findings. The aim of this study is to explore the feasibility and procedure of cochlear implantation in patients with CHARGE syndrome and to assess the outcome.
Study Design: Retrospective case review.
Setting: Tertiary referral center; cochlear implant program.
Patients: Ten patients with CHARGE syndrome and 3 patients with CHARGE-like syndrome treated in our center due to hearing impairment. Eleven patients were congenitally deaf, 1 patient had progressive hearing loss, and 1 patient had mixed hearing loss.
Intervention: Computed tomography of temporal bones and magnetic resonance imaging of the brain; bone-anchored hearing aid surgery, cochlear implantation, rehabilitation results.
Main Outcome Measures: Surgical suitability and hearing rehabilitation.
Results: We illustrate the management of preoperative diagnostics, surgical planning, and hearing rehabilitation. One patient with mixed hearing loss underwent bilateral bone-anchored hearing aid surgery. Because 2 patients had bilateral aplasia of the auditory nerves, we recommended an auditory brainstem implant. The unilateral cochlear implantation was performed in 9 patients and bilateral in 1 patient. In selected cases, it was helpful to plan the operation using a simulator for temporal bone surgery. Complex malformations, such as in CHARGE syndrome, with an increased intraoperative risk for complications should be facilitated by using intraoperative digital volume tomography–assisted navigation and intraoperative digital volume tomography control of electrode position. The results after CI surgery vary due to the differing extent of additional disabilities such as developmental delay, intellectual delay, and visual impairment. Nine of our patients showed improved responsiveness with the cochlear implant. Open speech comprehension could not be observed in 8 patients, whereas the follow-up period was less than 1 year in 4 patients. The relatively high age of our patients at implantation might be an important factor.
Conclusion: Careful planning of the treatment of CHARGE syndrome patients with sensorineural hearing loss can, to a limited extent, lead to auditory benefit without increasing surgical complications. Cochlear implantation is therefore indicated after critical assessment. **Key Words:** Bone-anchored hearing aid—CHARGE syndrome—CHARGE-like syndrome—Cochlear implant—Hearing loss—Navigation.
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CHARGE syndrome is a disease with multiple organ involvement. The acronym CHARGE is based on the initials of the most frequent symptoms (C, coloboma of the eye; H, heart defect; A, atresia of the choanae; R, retardation of growth or development; G, genital hypoplasia; E, ear malformation).

The syndrome was first described by Hall (1) and Hittner et al. in 1979 (2). The acronym CHARGE was coined by Pagon et al. (3) in 1981. This definition was revised in 1998 by Blake et al. (4) in a consensus based on additional information obtained over the intervening years. The 4 major criteria (CCCC) that occur most often in patients with CHARGE syndrome are coloboma, choanal atresia, involvement of cranial nerves (often multiple, most often in the following sequence: VIII, VII, IX, and X) and characteristic ear anomalies. Less frequent minor characteristics are genital hypoplasia, delayed development, cardiovascular malformations, retarded growth, orofacial cleft, tracheoesophageal fistulas, and characteristic facial features. CHARGE syndrome is defined in patients with all 4 major characteristics or 3 major characteristics and 3 minor characteristics. In rare cases, the presence of cleft lip and palate can be taken as a major

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TABLE 1. *Prevalence of major and minor malformations in patients with CHARGE and CHARGE-like syndrome*

	CHARGE syndrome									
	Major characteristics				Minor characteristics					
Patient	Coloboma	Choanal atresia	Cranial nerve involvement	Ear anomalies	Cardiovascular malformations	Psychomotor retardation	Genital hypoplasia	Renal malformations	Malformations of the ribs	Characteristic facial features
1	+		+	+	+	+				+
2	+		+	+	+	+				+
3	+		+	+	+	+				+
4	+		+	+	+	+				+
5	+		+	+	+	+				+
6	+		+	+		+	+			+
7	+		+	+	+	+	+			+
8		+	+	+	+	+				+
9		+	+	+	+	+				+
10	+		+	+		+	+		+	+
	CHARGE-like syndrome									
	Major characteristics				Minor characteristics					
Patient	Coloboma	Choanal atresia	Cranial nerve involvement	Ear anomalies	Cardiovascular malformations	Psychomotor retardation	Genital hypoplasia	Renal malformations	Malformations of the ribs	Characteristic facial features
11			+	+	+	+		+		+
12				+	+	+				+
13	+		+			+				

characteristic in place of choanal atresia (5). Some authors consider the characteristic malformations of the middle and inner ear in the computed tomography of patients with CHARGE syndrome to be a sensitive means of diagnosis and recommend including these as additional major criterion (6). The absence of semicircular canals, atresia of the oval window, and cochlear dysplasia, in that order, are the most sensitive characteristics of patients with CHARGE syndrome.

The diagnosis in children who present clinically with the signs of CHARGE syndrome, but do not show all of the major characteristics of the syndrome, should be characterized as "CHARGE-like" syndrome. The patients should be treated like patients with CHARGE syndrome due to the multiple malformations (7,8).

Because of the complexity of the middle and inner ear malformations, most patients have moderate to profound combined or inner ear hearing impairment. For this reason, early evaluation of hearing is necessary to enable optimal hearing rehabilitation of these multiple-handicapped children.

The purpose of this study was to examine the decision-making process, the surgical procedure of cochlear implantation, and the outcomes in a group of patients with CHARGE syndrome.

MATERIAL AND METHODS

The data of 10 patients who fulfilled the criteria for the definition of CHARGE syndrome were evaluated in a retrospective analysis of patients examined at the University-ENT Clinic Freiburg (tertiary referral center) between 1998 and 2009.

In addition, we included 3 patients who presented with the clinical picture of CHARGE syndrome but did not have all major characteristics; we classified them as having a CHARGE-like syndrome according to the definition by Shah et al. (7). The audiometric examinations were adapted to age and developmental stage of the patients. All patients underwent brainstem evoked

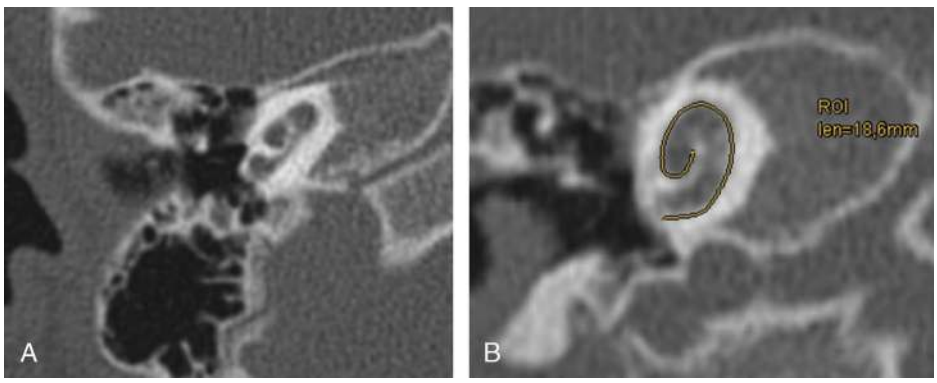

FIG. 1. Axial HR-CT scans of the cochlea. Hypoplasia of the cochlea (*A*). Length of the cochlea 18.6 mm (*B*).

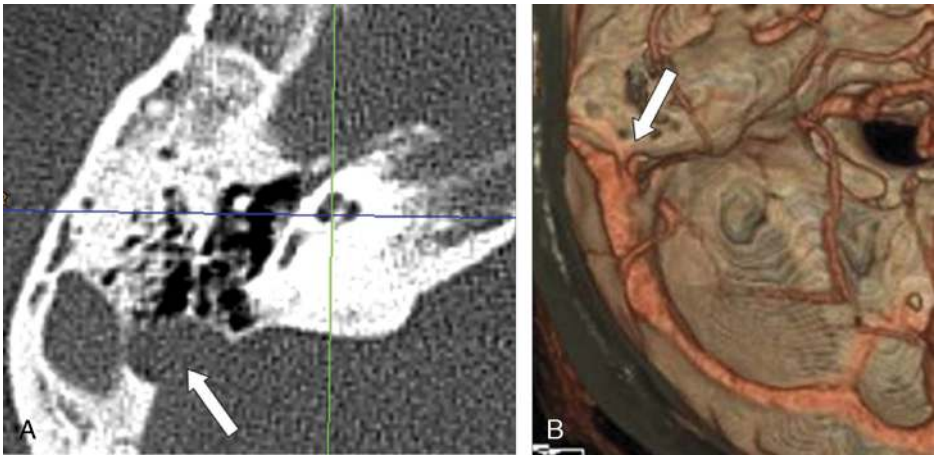

FIG. 2. *A*, Axial angio-CT scans: *arrow* shows atypical route of the sigmoid sinus and emissary veins. *B*, Three-dimensional reconstruction of the vessel malformations.

response audiometry. Pure-tone-audiometry was possible in 11 patients. Computed tomography (CT) and magnetic resonance imaging (MRI) data sets were evaluated with respect to the malformations present. We performed cochlear implantation in 10 patients. The intraoperative findings and the surgical procedure were assessed. A bone-anchored hearing aid surgery was performed in 1 case. Audiometry after cochlear implantation was performed with Freiburg Open Speech Test and Oldenburg Sentence Test at 70 dB. Speech therapy rehabilitation reports were also evaluated.

RESULTS

The analysis of patients at our clinic identified 10 patients (6 women, 4 men) with CHARGE syndrome and 3 patients (2 women, 1 man) with CHARGE-like syndrome. The individual distribution of major and minor characteristics of the patients is shown in Table 1. Of 10 patients with CHARGE syndrome, 9 were congenitally deaf, and 1 girl presented at age 3 with combined hearing loss. Congenitally deaf were 2 of the patients with CHARGE-like syndrome, whereas 1 patient had progressive hearing loss.

Air-conduction hearing aids had been fitted to 9 children at the age of 6 months to 2 years and to 2 children at age 2 with bone-conduction devices. Pure-tone audiometry was performed in all patients. A vibration curve was only available in 5 cases. We could determine bone-conduction thresholds between 70 and 100 dB in the remaining patients. The child with mixed hearing loss had a hearing threshold in the free acoustic field up to 1,000 Hz of 35 dB and greater than 2,000 Hz of 50 dB. No potentials in brainstem evoked response audiometry were found in 12 of the 13 patients.

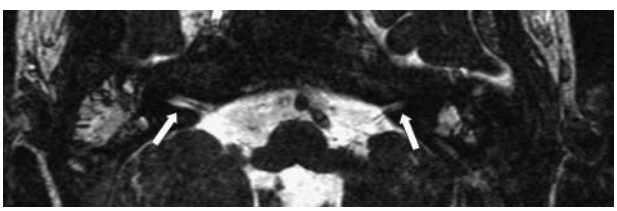

FIG. 3. Axial MRI (T2, ciss–3-dimensional sequence) of the IAC. Bilateral aplasia of the cochlear nerve (*arrows*).

CT and MRT Findings

All patients underwent high-resolution CT (HR-CT) supplemented in 12 cases by MRI. The most frequent malformations were dysplastic or missing ossicles and cochlear hypoplasia (all patients, Fig. 1). Aplasia of the semicircular canals was found in 12 of the 13 patients. An atypical routing of the facial nerve and of emissary veins was present in 6 patients and an atypical anatomy of the sigmoid sinus in 5 patients (Fig. 2). We observed atresia of the oval window in 10 patients and of the round window in 9 patients. Computed tomographic scans revealed dysplasia of the internal auditory canals in 7 cases (12 ears). Magnetic resonance imaging scans identified an aplasia of the vestibulocochlear nerve in 4 patients (2 patients bilateral) and a missing cochlear nerve in 1 patient (Fig. 3). Vestibular hypoplasia was less frequent, and an enlarged vestibular aqueduct was present in only 1 patient (Table 2).

Therapy of CHARGE and CHARGE-Like Patients

The 7-year-old CHARGE syndrome girl with combined hearing loss had been fitted at age 3 with a transcutaneous

TABLE 2. *Computed tomographic and MRI findings of the middle and inner ear malformations*

No. patients (%)	Malformation
13 (100)	Dysplastic or missing ossicles
13 (100)	Cochlea hypoplasia
12 (92)	Semicircular canal aplasia
10 (77)	Oval window atresia
9 (69)	Round window atresia
7 (54)	CT scans: dysplasia of the internal auditory canal
5 (38)	MRI scans: aplasia of the VIIIth nerve (2 patients bilateral); 1 patient aplasia of the cochlear nerve
6 (46)	Aberrant course of facial nerve/emissary veins
5 (38)	Atypical localization of sigmoid sinus

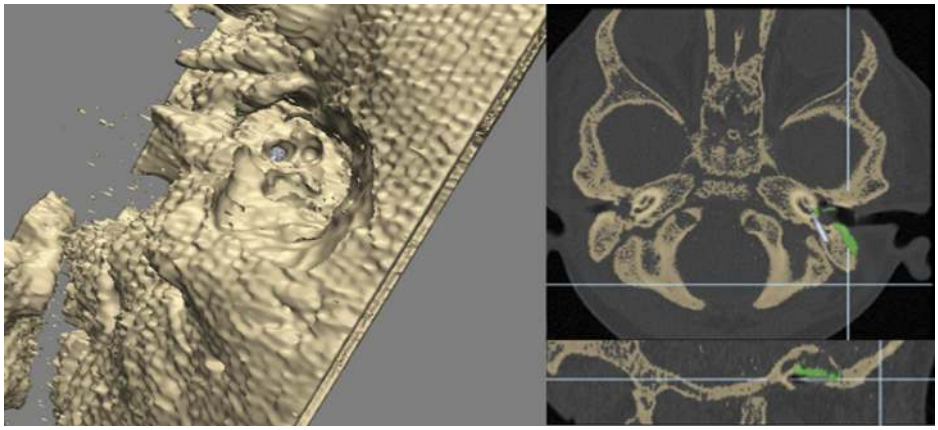

FIG. 4. Preoperative 3-dimensional simulation of the surgical approach using the VOXEL-MAN TempoSurg in a patient with CHARGE syndrome; the drill is positioned at the cochleostomy.

bone-conduction hearing aid, and bone-anchored hearing aid surgery was performed at age 6. The indication for cochlear implantation (CI) was given in 7 patients with CHARGE syndrome and all 3 patients with CHARGE-like syndrome. Because 2 patients had an aplasia of the auditory nerves on both sides, cochlear implantation could not be recommended. The parents were intensively informed regarding the possibility of auditory brainstem implantation. To date, the parents have refused this option because of the surgical risks.

Operative Procedure During CI

The mean implantation age of the patients with CHARGE syndrome was 5.31 years (1.5–14 yr). Two patients with a CHARGE-like syndrome underwent implantation at the age of 3.5 years and 1 patient at the age of 31 years. Because of atypical anatomy, preoperative planning of the surgical access and cochleostomy was made in 2 cases with a simulator for temporal bone surgery (VOXEL-MAN TempoSurg; University Medical Center Hamburg-Eppendorf, Germany; Spiggle

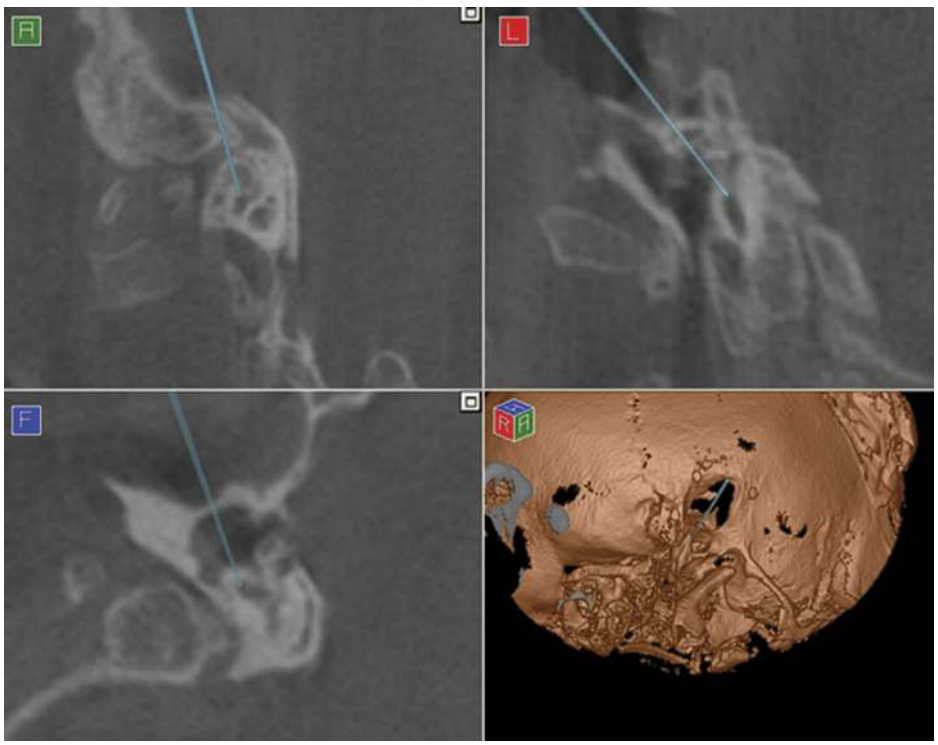

FIG. 5. Intraoperative DVT with navigation due to complex malformation of the mastoid, sigmoid sinus, and atypical course of emissary veins; *blue spot* shows the localization of the cochleostomy.

TABLE 3. *Patient demographic, results after surgery*

Patient	Age at surgery, yr	Ear	Onset of deafness	Surgery	Electrode	Insertion	ECAPS	Intraoperative stapedius reflexes	Follow up, yr	Communication mode	General remarks
CHARGE syndrome											
1	3.2	Left	Congenital		CI 24 RST	22 + 8	Negative	Missing stapedius tendon	2 (9)	Sign language	Refused the CI after 2 yr
2	4.2	Left	Congenital		CI 24 RST	22 + 5	Positive	Missing stapedius tendon	8.5	Sign language	More receptiveness and responsiveness
3	6	Left	Congenital	Navigated	CI 24 RCA	Complete	Positive	Missing stapedius tendon	0.75	Begins to use oral language	More receptiveness and responsiveness
4	3.75	Right	Congenital		CI 24 RCA	Complete	Positive	Positive	3	Sign language	More receptiveness and responsiveness
5	1.5	Left	Congenital		CI 24 RCA	Complete	Positive	Positive	0.6	Begins to use oral language	More receptiveness and responsiveness
6	4.5	Right	Congenital	Navigated	CI 24 RCA	Complete	Positive	Positive	0.3	Begins to use oral language	More receptiveness and responsiveness
7/8							Auditory brainstem implantation recommended				
9	6						Bilateral bone-anchored hearing aid surgery				
10	14	Left	Congenital		CI 24 RST	22 + 2	Negative	Missing stapedius tendon	10.2	Sign language	More receptiveness and responsiveness
CHARGE-like syndrome											
11	31	Left	Progredient	Navigated	CI 24 RST	Complete	Positive	Missing stapedius tendon	3.75	Oral language	Good understanding even in noise
12	3.5	Right	Congenital		CI 24 RCA	Complete	Negative	Positive	0.75	Sign language, begins to use oral language	Reaction to quiet sounds
13	3.5	Bilateral	Congenital	1. re 2. li	1. Surgery CI24 RST 2. Surgery CI24 RCA	1. Surgery: 22 + 6 2. Surgery, complete	1. Surgery, negative 2. Surgery, positive	1. Surgery, negative 2. Surgery, positive	7.25	Oral language	College preparatory school

& Theis, Overath, Germany). The Digital Imaging and Communications in Medicine–CT data set of each patient was uploaded into the system, and the surgical approach was simulated in 3-dimensional images (Fig. 4). In 3 cases, digital volume tomography (DVT) was used intraoperatively under navigation conditions due to the complex additional malformation of the mastoid, sigmoid sinus, and the atypical course of the emissary veins and/or facial nerves (Fig. 5). Intraoperative radiologic control was performed to determine the correct position of the electrode in the hypoplastic cochlea. There was a slight gusher in 1 case, which was controlled by sealing the cochleostomy. Six patients received a Nucleus CI 24RCA and 3 patients a CI 24RST. A girl with bilateral implantation received a CI 24RST on the first side and a CI 24RCA on the second side. Complete insertion of the electrode array was possible in all cases. Intraoperative neural response telemetry could be recorded in 7 patients. Intraoperative stapedius reflex measurement was not possible due to a missing stapedius tendon in 5 cases (Table 3).

Audiologic Results

The mean follow-up time in the CHARGE syndrome patients after cochlear implantation was 3.6 years (0.3–10.2 yr). The CHARGE-like syndrome patients were followed up for 0.75 to 7.25 years (Table 3). Even after a longer follow-up period, 4 patients with CHARGE syndrome were only able to use sign language and could not develop any speech understanding or oral language as a means of communication. One patient showed no benefit after cochlear implantation and refused the cochlear implant 2 years after surgery. Three patients with CHARGE syndrome who hade been using the CI less than 1 year were beginning to use oral language and to recognize speech as a means of communication. Of 3 patients with CHARGE-like syndrome, 2 patients with an implantation time of 3.75 and 7.25 years used oral language as sole instrument for communication. One patient after bilateral implantation was even attending college preparatory school. Another child who has been using the CI for only 9 months is beginning to make sounds in addition to sign language. Speech tests such as Freiburg Monosyllables and Numbers Test or Oldenburg Sentence Test were not possible in any patient with CHARGE syndrome. In 2 patients with CHARGE-like syndrome, open speech comprehension could be demonstrated in Oldenburg Sentence Test and in Freiburg Monosyllables at 70 dB in each case (Table 3). These 2 patients have good speech comprehension even in distracting noise. The families reported on improved response and reaction capability of the patients in all cases.

DISCUSSION

In newborns with choanal atresia and/or coloboma, it is required to look for additional malformations to identify a CHARGE syndrome. In such a case, despite frequent primary intensive care monitoring and treatment due to cardiac malformations and involvement of cranial

nerves, it is mandatory to diagnose impaired hearing as early as possible. Late onset of therapy could lead to delayed development of the child. In profound hearing loss, cochlear implantation should be considered early because communication with the environment is limited in children with CHARGE syndrome because of an additional visual disability due to coloboma, which is often present. Feasibility and successful implantation in patients with CHARGE syndrome have been described by several authors (9–15).

An HR-CT scan is mandatory before cochlear implantation to detect anomalies in the middle and inner ear. According to Amiel et al. (6), the most frequent inner ear anomalies in patients with CHARGE syndrome are aplasia of the semicircular canal, atresia of the oval window, and cochlear dysplasia. The most frequent malformation in our patients was cochlear hypoplasia. Semicircular canal aplasia and atresia of the oval window were found with equal frequency. Atresia of the oval window, which occurs frequently, was also described by Lemmerling et al. (16) and Morimoto et al. (17). Information on atypical routing of the facial nerve is important when planning surgery. Abnormal courses of the facial nerve in CHARGE syndrome have been described by several authors (16–18). In 3 cases, either the entire facial nerve or another branch of the nerve crossed the promontorium in our patients and partially obscured the round window. In these cases, the cochleostomy was performed more inferior than during the routine procedure. Today, using intraoperative facial nerve monitoring is a matter of common knowledge. Important information obtained by CT scans before cochlear implantation is the value of the internal auditory canal (IAC) because this may provide evidence of aplasia of the VIIIth nerve. Morimoto et al. (17) assume hypoplasia of the IAC when the diameter is less than 2 mm. Bamiou et al. (19) reported on narrow IACs in 5 of 6 ears with aplasia of the VIIIth. We could demonstrate a hypoplasia of the IAC in 12 of 26 ears with the CT scans. Aplasia of the VIIIth nerve was one-sided in 3 patients and bilateral in 2 patients, as shown in MRI scans. For this reason, patients with CHARGE syndrome should always undergo MRI examination to rule out possible aplasia of the VIIIth nerve. In the case of one-sided aplasia, cochlear implantation could be possible on the other side. In bilateral aplasia, the alternative of auditory brainstem implantation must be discussed with the patient's parents. Because of the multitude of anomalies in the middle and inner ear, careful planning of the surgery with respect to the surgical access to the cochlea and a possibly required intraoperative navigation and imaging such as x-ray, DVT (20), or HR-CT scan (21) is mandatory. In our patients, preoperative planning with a simulator for temporal bone surgery and intraoperative-navigated DVT has proven valuable in difficult cases. This procedure is not necessary in all patients with CHARGE syndrome, but it should be available for cases of complex malformations. The selection of the electrode to be implanted is influenced by the cochlear anatomy. Shorter, perimodiolar, or compressed electrode arrays should be used for cochlear hypoplasias. In principle, cochlear implantation is possible in patients with CHARGE syndrome as long as attention is paid to the anatomic situation.

The results after CI surgery in patients with CHARGE syndrome vary due to the differing extent of additional disabilities such as developmental delay and visual impairment. Twelve of our patients showed improved responsiveness with the cochlear implant. Open speech comprehension could not be observed in the patients with CHARGE syndrome, whereas the follow-up period was less than 1 year in 3 patients. The relatively high age of our patients at implantation might be an important factor. Three children followed up for less than 1 year who were implanted at a relatively early age are beginning to make sounds, and the parents report a clear increase in attention. This confirms the assumption that implantation at an early age can be recommended for these children as well. The reason for the good speech development and comprehension in 2 patients with CHARGE-like syndrome might be the lower degree of additional diseases or malformations in these patients. Our results in patients with CHARGE syndrome are comparable with those of other authors. In a follow-up period of between 1 and 15 years, Lanson et al. (14) also observed only sign language or no form of communication in 6 of 10 children. However, they also reported clearly improved responsiveness and attention. We assume that the poor results, as found in several of our patients, are owing to neural deprivation, late implantation, and, in some cases, to poor cognitive abilities. Because of the retrospective study design, there are no IQ test results available. This would be necessary to confirm our hypothesis. It should be considered that these patients require a considerably longer rehabilitation time due to their delayed development. We anticipate improvement in communication capability after cochlear implantation in this group of patients with severe multiple handicaps. Speech development or even speech comprehension will only be possible in individual cases. This should be made clear when informing the parents.

Long-term results of 156 implanted children, including multihandicapped children, showed that congenitally deaf children or children with residual hearing implanted before the age of 2 had significantly better results in speech comprehension than children implanted late (22). The data showed highest significance in all tests; the younger the implantation age of the patient, the better the outcome with a follow-up of 5 years. This should also apply for multiple-handicapped children with CHARGE syndrome especially because these children must be helped as early as possible to guarantee optimal development and thus enable communication with the environment.

Raqbi et al. (23) examined early prognostic factors for the intellectual development results of children with CHARGE syndrome. It is interesting that severe initial medical problem such as hospitalization in the intensive care ward, serious cardiac disease, multiple surgeries, and tracheotomies do not correlate with the intellectual development result. Profound hearing impairment has

a nonstatistically significant negative influence on the intellectual result. Overall, 50% of the children with CHARGE syndrome reach a good and 25% a very poor intellectual result.

Thus, it is absolutely necessary to identify hearing impairment in children with CHARGE or CHARGE-like syndrome during neonatal hearing screenings to immediately initiate measures aimed at achieving optimal conditions for development with respect to hearing rehabilitation in this group of patients.

In conclusion, early identification of sensorineural hearing loss in children with CHARGE syndrome, an intensive counseling of the pediatrics and parents, and careful planning of the surgery can lead to auditory benefit without increased surgical complications. Therefore, after critical assessment, cochlear implantation is indicated.

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