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ORIGINAL ARTICLE

Unilateral hearing loss in children: a retrospective study and a review of the current literature

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Abstract Despite the introduction of universal newborn hearing screening (UNHS), unilateral hearing loss (UHL) is sometimes recognized late. This diagnostic delay has adverse repercussions, given the importance of binaural hearing for the development of normal auditory processing. It is incorrect to maintain that unilateral hearing is the minimum requirement for adequate speech development and that hearing aid provision is consequently unnecessary. In our retrospective study, hearing aid provision resulted in improved directional and selective hearing (quiet *and* noisy environments) and, compared with their chronically ill counterparts, the children in our study displayed superior health-related quality of life (HRQoL) scores in all areas. On the basis of the results, the authors conclude that even mild hearing losses (from an auditory threshold of 30 to 40 dB) should have the opportunity for hearing aid provision. A selective literature review was conducted in PubMed and textbooks and with reference to national and international guidelines. Early diagnosis and treatment of UHL have a positive effect on verbal-cognitive, linguistic, communicative, and socio-emotional development, as demonstrated by neurophysiological studies. Among the treatment modalities with differing effects on the quality of binaural hearing, cochlear implants are now used increasingly in children with hearing loss bordering on deafness.

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Conclusion: Published evidence and clinical experience support early diagnosis and treatment. Wherever feasible, hearing aid provision before or at the end of the first year of life is recommended for children with UHL.

What is Known:
- *Almost 30 years ago, poor academic performance was reported in children with unilateral hearing loss (UHL).*
- *Despite improvements in treatment options, it is traditionally held that unilateral hearing is the minimum requirement for adequate speech development and hearing aid provision is unnecessary.*

What is New:
- *Academic and behavioral deficits in children with UHL may be mediated by deficiencies in the default mode network.*
- *Published evidence supports the recommendation for hearing aid provision before or at the end of the first year of life in children with UHL.*

Keywords Unilateral hearing loss (UHL) · Binaural hearing · Auditory processing · Speech development · Hearing aid provision · Health-related quality of life

Abbreviations

BAHA	Bone-anchored hearing aids
BERA	Brainstem evoked response audiometry
BTE	Behind-the-ear (hearing aids)
CI	Cochlear implant(s)
CMV	Cytomegalovirus
CROS	Contralateral routing of signal
CT	Computed tomography
DGPP	German Society of Phoniatrics and Pediatric Audiology
DMN	Default mode network
FAST	Frequency Animal Sound Test
FM	Frequency modulation (systems)
HRQoL	Health-related quality of life
mFAST	Multi-Frequency Animal Sound Test
OAE	Otoacoustic emission(s)
SNHL	Sensorineural hearing loss
SNR	Signal-to-noise ratio
TEOAE	Transient evoked otoacoustic emission(s)
UNHS	Universal newborn hearing screening

Introduction

Binaural hearing is important for hearing, speech, and general child development, as international studies have demonstrated for decades. In routine clinical practice, however, these insights are not reflected in the depth of the diagnostic workup or in the breadth of treatments offered.

Hearing has to be "learnt." While the cochlea is mature by week 23 of gestation, the development of auditory processing and perception requires binaural hearing ability and takes more than a decade to emerge [30]. This is the only way that elements such as redundancy and the head shadow, squelch, and cocktail party effects can come into play to permit sound localization, speech perception in background noise, and spatial hearing (see Table 1).

By contrast, adults and children with unilateral hearing loss (UHL) experience difficulties with sound localization [55]. Even a proposed compensatory mechanism is not a true substitute for the sound localization achieved with normal binaural hearing [28]. In addition, UHL and lack of head shadow mean that the speech signal cannot be separated from background noise [31]. Speech perception is reduced in background noise [13, 55] but also in quiet conditions [13].

Inadequate reception of acoustic stimuli and irregular stimulation of the central auditory system can have repercussions for the development of hearing ability. In early-onset hearing impairment, in particular, moderate and severe UHL can have adverse effects on the child's verbal-cognitive, linguistic, communicative, and socio-emotional development [9–11, 14, 18, 69].

Positive developments have been recorded in the field of UHL in terms of age at diagnosis, available treatment modalities, and treatment recommendations.

Whereas UHL used to be diagnosed in preschoolers or early school-aged children [10, 22, 34, 67, 69], initial diagnosis can now be made in the third month of life thanks to the high quality of universal newborn hearing screening (UNHS) [56].

Technical advances have made it possible to provide behind-the-ear (BTE) hearing aids and bone-anchored hearing aids (BAHA) instead of a contralateral routing of signal (CROS) system. Frequency modulation (FM) systems are used in schools, where clearer auditory information and elimination of background noise can improve concentration. Cochlear implants (CIs) are increasingly being used in children, and treatment recommendations have been issued at the international level [3, 20].

Despite these developments, a scientific basis for standardized diagnostic workup, counseling, and treatment is still wanting. Thirty years ago, Bess and Tharpe observed academic deficits in children with UHL [10], and yet decades later we still lack any large prospective studies or an international database with results from validated tests or from gold-standard pediatric audiological and radiological diagnostics.

Materials and methods

To evaluate the impact of hearing aid provision versus non-provision on quality of hearing and quality of life, the medical records of a total of 152 children with

Table 1 Effects and advantages of binaural hearing

Effect	Definition	Advantage
Dichotic hearing	Reception and understanding of two different pieces of speech information presented simultaneously •Right ear advantage. Signal processing in the left hemisphere (e.g. speech) • Left ear advantage. Signal processing in the right hemisphere (tonal musical stimuli)	Improved speech perception, e.g., in a noisy environment
Redundancy	Binaural neuronal processing of two identical signals by the brain	Improved speech perception due to information overlap in the brain [40] • Frequency differences due to the pinna, leading to a change in sound signal • Loudness improved by 1.7 sone (unit of measurement) Clearer and more easily comprehensible auditory impression [40] Speech perception in background noise [33]
Head shadow effect	Decrease in volume level when the acoustic signal (speech) and background noise arrive from two different directions • Reduces speech and noise on the side turned away from the sound as they are blocked by the acoustic shadow of the head • Switches the brain to the side with the better signal-to-noise ratio and signal analysis • Degree of reduction is determined by frequency and direction of incidence of the acoustic signal [40] • Especially for high tones • Unit of measurement in the case of lateral sound incidence for speech signal approx. 7 dB	Speech perception in background noise [33]
Squelch effect	Makes the desired acoustic signal more audible by suppressing background noise • If binaural processing is intact, possibility of binaural noise suppression • Binaural masking level difference (BMLD) (for simple tasks above 25 dB)	Speech perception in background noise [33] Spatial hearing
Cocktail party effect [12, 17]	Binaural neuronal processing of two different signals by the brain • Filters out a single speaker from the babble of voices at a party • Noise reduction and spatial orientation by means of signal differences in terms of time, spectral range, and loudness • Comparison of these signals: brain can amplify the desired signal from one direction and suppress background noise and reverberation from the other direction up to 15 dB	Speech perception in background noise Separation between several auditory objects in the room Spatial hearing

hearing loss were reviewed. Within this sample, data were analyzed retrospectively for 107 children with UHL treated in the Department of Voice, Speech and Hearing Disorders, University Medical Center, Hamburg-Eppendorf (UKE), or in the Werner Otto Institut (WOI), Department of Phoniatrics and Pediatric Audiology. Both centers are staffed by multi-disciplinary expert teams experienced in the prevention, diagnosis, and treatment of pediatric hearing loss and speech disorders. The patients who provided retrospective data for the present study comprised all children who had been diagnosed with and routinely monitored and/or treated for UHL in these two units.

The children taking part were born in the period between February 1990 and September 2004 and at the time of the study were between 4 and 18 years old, giving an observation period of 162 months (13.5 years). The mean age of all 87 children observed was 10.2 years. Twenty children were excluded from the study because they had a global physical and mental disability or acute inflammation of the outer, middle, and inner ear or because important audiometric data were missing.

The study was approved by the ethics committee of the Hamburg regional medical board.

The diagnosis of UHL was made by careful pediatric audiological testing. Hearing loss was interpreted as an average hearing loss greater than 21 dB at frequencies of 0.5, 1, 2, and 4 kHz [8, 46, 58, 70]. Functional gain (the difference between audiometric curves recorded in the aided and unaided subject on exposure to tones and sounds) was determined, and the improvement in auditory threshold under aided conditions was described in terms of levels (1, 2, or 3, as defined in Table 2).

Table 2 Degree of hearing loss

Degree of hearing loss on tone audiometry	Mean hearing loss at 0.5, 1, 2, and 4 kHz
Mild hearing loss	21–40 dB
Moderate hearing loss	41–60 dB
Severe hearing loss	61–80 dB
Deafness	>80 dB

Three groups were formed: children (A) with and (B) without hearing aids and (C) children previously fitted with hearing aids. No child was supplied with a cochlear implant.

Various questionnaires were used (Table 3). From the pool of questions used for the "Living with Hearing Disorders" project, a 23-item questionnaire was designed and sent out. Analysis of the questionnaire was limited purely to the descriptive level, the aim being to address relevant problem areas and to identify any trends. It was not possible to compare this purely descriptive analysis of the pediatric questionnaire with the analysis of the DISABKIDS parental questionnaire.

The questionnaires were sent separately to parents/legal guardians and children, together with an information sheet and a consent form.

Results

The retrospective study covered the period from February 1990 to November 2007.

Audiological data were evaluated from 87 children (34 children (A) with and (B) 43 children without hearing aids and (C) 10 children previously fitted with hearing aids). Questionnaires were sent to 87 families, and responses were received from 74 families (85%): in detail, responses were received for 32 children currently with a hearing aid, 32 children without a hearing aid, and 10 children previously with a hearing aid.

The three groups (A, B, and C) showed virtually no differences in terms of demographic data: age, observation period, side (right/left), gender, type of school, age at initial diagnosis, age at hearing aid provision, and speech development (see Table 3).

The hearing of 48.6% of children currently or previously with a hearing aid and supplying valid audiometry data improved by one level. Two- and three-level improvements were recorded in 16.2 and 21.6% of these children, respectively. (Seven out of the total of 44 children currently or previously provided with a hearing aid had no audiometric data for improvement following hearing aid prescription.)

More than half of the children with hearing aids felt that their directional hearing and selective hearing in quiet and noisy environments had improved.

Hearing aid acceptance was high. Irrespective of the degree of hearing loss, time of hearing aid provision, family circumstances, number of household members, and the working hours of both parents, the hearing aids were worn for more than 8 h a day. Not quite 60% of parents reported a marked improvement as a result of hearing aid provision.

Compared with chronically ill children from the DISABKIDS Project, children with UHL scored better in all areas of health-related quality of life (HRQoL). The two groups scored very differently on the "Emotion" and "Physical limitations" subscales.

In the individual groups (A, B, and C), parental assessments of HRQoL of children with UHL showed hardly any differences in terms of independence, emotion, social inclusion, social exclusion, physical limitations, and treatment, with scores ranging from 71.2 to 90.1%.

General quality of life for children with UHL ($n = 32$) currently supplied with a hearing aid showed a mean score of 81.9%. Modified HRQoL (modified in the sense of extracting the hearing aid-related questions) was compared between the three groups of children currently with a hearing aid ($n = 32$), without a hearing aid ($n = 32$), and previously with a hearing aid ($n = 10$). All three groups had mean HRQoL scores in excess of 80% (83.3, 85.9, and 82.5%, respectively).

Review and discussion of the current literature

Definition, incidence, and prevalence

UHL is a mild to severe hearing impairment, bordering on deafness in one ear with normal hearing in the other ear. It includes chronic conductive, sensorineural, and combined hearing losses [3].

The most common congenital condition is hearing loss and affects 1 to 3 per 1000 live births [3].

The prevalence of UHL in US adolescents between the ages of 12 and 19 years rose from 11% in 1988–1994 to 14% in 2005–2006 [63]. Prevalence estimates for UHL may vary by as much as a factor of 2 with various applications of commonly accepted case definitions. A general consensus is needed to define the parameters (threshold levels, tympanometry status, and pure-tone audiometry frequencies) to be used as a basis for prevalence rate estimates [57].

Table 3 Overview of retrospective study participants (including demographic data) and questionnaires used

	Study participants		
	Group A: children currently with hearing aid	Group B: children without hearing aid	Group C: children previously fitted with hearing aid
Age (years)	10.7 (mean) 10.4 (median)	9.7 (mean) 9.5 (median)	10.6 (mean) 10.4 (median)
Observation period (years, maximum)	17.1	17.7	13.6
Sensorineural hearing loss (*n*)	30	40	8
Conductive hearing loss (*n*)	2	2	1
Combined hearing loss (*n*)	2	1	1
Mild hearing loss (*n*)	7	13	0
Moderate hearing loss (*n*)	12	5	4
Severe hearing loss (*n*)	3	6	2
Hearing loss bordering on deafness (*n*)	12	19	4
Right side/left side (*n*) [%]	21:13 [61.7:38.3]	23:20 [53.5:46.5]	3:7 [30:70]
Boys/girls (*n*)	17:17	22:21	6:4
Behind-the-ear hearing aid (*n*)	27		10
In-the-ear hearing aid (*n*)	3		
CROS (*n*)	1		
No specification on the hearing aid type	3		
Type of school (*n*) Mainstream school/non-mainstream school/other	26:2:1	26:1:0	7:2:0
Age at first diagnosis (years)	6.2	5.9	5.1
Age at hearing aid provision (years)	7.1		6.4
Delayed speech development (*n* = no/yes), [% = no/yes)]	27:5 [84.3:15.7]	28:5 [84.8:15.2]	8:1 [88.9:11.1]
Binaural hearing gain in directional hearing	Definitely improved: 4 Improved: 13 No change: 7 Worse: 6		
Gain in quality of selective hearing in quiet surroundings	Definitely improved: 9 Improved: 13 No change: 5 Worse: 3	Completed only for children currently with hearing aid	Completed only for children currently with hearing aid
Gain in quality of selective hearing in noisy surroundings	Definitely improved: 5 Improved: 13 No change: 7 Worse: 5	Completed only for children currently with hearing aid	Completed only for children currently with hearing aid
Gain in speech comprehension in noise	Definitely improved: 2 Improved: 14 No change: 9 Worse: 4	Completed only for children currently with hearing aid	Completed only for children currently with hearing aid
Acceptance of wearing hearing aids	Not accepted at all: 3 1–4 h: 2 >4–8 h: 5 >8 h: 21	Completed only for children currently with hearing aid	Completed only for children currently with hearing aid
Questionnaires	Content		
DISABKIDS questionnaire	Measures HRQoL in children with chronic health conditions. The chronic generic module consists of 37 Likert-scaled items assigned to six dimensions or subscales. The six subscales are additionally associated with three domains, denoted as mental, social, and physical. http://www.disabkids.org/questionnaire/disabkids-core-instruments/dcgm-37-long-version/		
"Living with Hearing Disorders" questionnaire	Questions about the quality of life of children with UHL compiled from the "Living with Hearing Disorders" project www.uzh.ch/orl/dga2008/programm/wissprog/Bruett.pdf		
"Wearing acceptance" questionnaire	Questions about wearing acceptance among children with UHL (http://www.unimedizin-mainz.de/typo3temp/secure_downloads/27121/0/4148a55d324e464ee6f34ee166e8031c84ee296b/Fragebogen_Kinder_einseitige__Schwerhoerigkeit.pdf)		
"Sociodemographic data" questionnaire	Captures sociodemographic data		

Etiology and prognosis

The etiology of UHL is unknown in 35% [14] to 60% [22, 35] of cases. In addition to inherited syndromic or non-syndromic hearing loss, acquired prenatal and perinatal UHL may also develop [49]: high-tone hearing losses have been reported in hydrocephalic children, occurring in the ear ipsilateral to shunt placement in 83% of cases [64]. The most common causes of

postnatal acquired UHL are neurotropic viruses (e.g., cytomegalovirus (CMV)) and head trauma [14]. Presence of progressive hearing loss is found in 14 to 32.8% of cases [7, 69] and is a predictor of abnormal computed tomography (CT) outcome [7]. However, pathological CT findings are not dependent on the degree of hearing loss, a positive family history, or consanguinity. CT scans of the petrous bone have revealed abnormalities—such as dilated vestibular aqueduct(s)—in 31 to 44% of cases, sometimes bilaterally [7, 48]. The possibility of progression from unilateral to bilateral hearing loss must also be recognized [48]. In one study, two thirds of asymptomatic CMV-infected children suffered subsequent deterioration of hearing, starting as UHL in 78% of cases [71].

Speech development/general development/school career

Some form of speech development disorder is found in 2 to 15% of 4- to 6-year-olds [6].

In our retrospective study, 14.9% of the children were "behind" in terms of speech development. No difference was found in this respect between children who were provided with a hearing aid and those who were not. As a caveat, it should be pointed out that the information on speech development was gleaned from case records, questionnaire responses, or reports from the doctor or therapist.

Few studies have specifically investigated speech using language tests in children aged between 3 and 12 years who have UHL but no other impairment [13, 43, 44, 53, 55]. These studies covered a variety of facets, such as type of hearing loss, degree of hearing loss, and test materials for investigating receptive and expressive language knowledge. There is no gold standard for language testing in children with UHL [32]. The speech problems presented do not indicate the formal and non-formal language levels where these problems are manifest. Children with UHL have poorer test results in expressive and receptive tests than their normal-hearing peers [43, 44]. Children with UHL scored better in expressive language tests than children with mild bilateral, severe, or conductive hearing loss [13]. The significant differences seen in 4- to 5-year-old children with severe UHL bordering on deafness compared with normal-hearing peers could no longer be detected by the age of 6 years [13].

In a recent prospective longitudinal study, individualized education plans and higher baseline cognitive levels were predictors of better results over time in standardized cognitive and language tests [42], but without any improvement in academic performance. The results could not be generalized to all degrees of hearing loss (severe hearing loss bordering on deafness was noted in most cases (61%)), and any continuing impact through to their working career was not ascertained.

Early-onset, perinatal, and/or postnatal complications as well as profound right-sided hearing loss bordering on deafness increase the risk of deficient academic performance (Table 4) [11, 14, 18, 43, 44, 66, 67].

Noise levels in classrooms are rising because of changes in teaching methods (working in pairs and groups instead of traditional teaching from the front). Where acoustics are poor, "normal-hearing" students at the back pick up only 60% of the information being communicated [45, 47]. The principal reasons are background noise and reverberation. In rooms with adverse acoustic conditions (reverberation time >0.55 s), communication difficulties may arise which cannot be accounted for by a poorer teacher-pupil relationship [38].

Certain aspects of auditory processing are not fully developed in childhood [52]. A normal-hearing pupil in grade 1 requires a greater signal-to-noise ratio (SNR) for 95% intelligibility than a grade 6 pupil does (Fig. 1; modified from [72]). Children with UHL need an even greater SNR [60] and face particular challenges against such a background. The behavioral problems of children with UHL are thought to be due to hearing loss and the associated deficits in attention and communication [66]. The proportion of behavioral problems and difficulties with classmates of the same age can be 2.4 times higher than in normal-hearing pupils. Children who have poorer communication skills and integrate less well into the class are affected to a greater extent. Socio-emotional development is not dependent on gender, age, or hearing status. Hintermair and Wiegand [24] conclude that teachers must ensure that communication is encouraged in children with UHL, enabling them to participate actively in lessons. Involving them in the social environment might foster better socio-emotional development, although future studies will be required to demonstrate this [24].

Audiological diagnostic workup: scope and limitations

The UNHS performance targets for the audiological diagnostic workup are not always adhered to because the importance and purpose of the particular measuring techniques are not properly understood [56].

Table 4 Overview—academic deficits

Risk
Lower level of academic attainment [18]
At least one school year repeated by 18–35% [66]
Lower verbal intelligence in those repeating a year [11]
Learning difficulties [67]
Academic resource help needed for one or more years in 12 to 60% [66]
Behavioral problems in 20 to 59% [11, 14]
Lower scores in receptive and expressive language tests [43, 44]

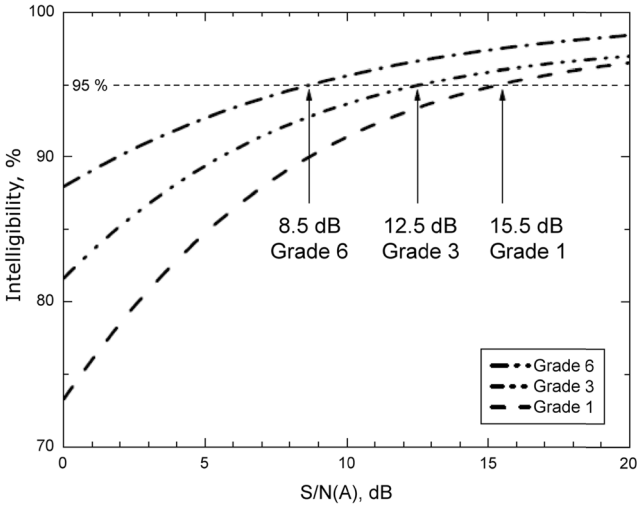

Fig. 1 Dependence of speech intelligibility on signal-to-noise ratio by school grade (modified from [72])

Transient evoked otoacoustic emissions (TEOAEs) are detected using a series of click stimuli with a broad frequency range between 1000 and 4000 Hz. Functional integrity of the outer hair cells is necessary for active and non-linear sound amplification by the inner ear [25].

A functional disorder of the inner and/or middle ear is frequently the underlying cause of abnormal OAEs. A defect of the outer hair cells and/or middle ear fluid results in failure to generate OAEs. The detected presence of OAEs does not necessarily indicate normal hearing. Where there is a functional disorder of the inner hair cell, synapse, or cochlear nerve, where efferent regulation of the outer hair cell is disrupted [1], and where hearing loss occurs between 21 and 30 dB and at very high or low frequencies, TEOAEs are detected even though auditory impairment is present. As a result, mild hearing losses are not picked up by OAE measurement during UNHS. To complete the diagnostic workup, frequency-specific brainstem evoked response audiometry (BERA) should be used when hearing loss is suspected. Free-field testing in infants does not detect UHL. Mild and unilateral hearing losses that are not diagnosed during the course of UNHS require hearing screening at a later stage. The Frequency Animal Sound Test (FAST4) or its successor, the multi-Frequency Animal Sound Test (mFAST), are validated measuring instruments.

Age at first diagnosis

At least for mild hearing losses <30 dB, it is currently unlikely that age at first diagnosis can be brought forward because even UNHS sometimes fails to detect such losses. In our study, the (mean) age at first diagnosis of 5.9 years is consistent with findings in the literature for children prior to the introduction of UNHS [35, 56].

Early intervention for hearing losses diagnosed before the age of 1 year

Early diagnosis calls for new strategies in early hearing and speech intervention.

Early diagnosis and intervention barely alter perceptions of stress in parents of infants with bilateral hearing loss compared with parents of healthy children. The empowerment concept assigns parents the decisive role in their child's development, hence the need for family-centered work to strengthen the parent-child relationship and the family's social networks, including the contribution of parents' associations [61].

It might be appropriate to transpose these findings to children with UHL, but this would need to be verified empirically [personal communication from Professor Hintermair: 60].

Treatment

A range of treatment options are available with differing effects on the qualities of binaural hearing, and the nature and degree of hearing loss must clearly play a role in treatment choice (Table 5) [20].

Intervention at the 6-month stage (onset of crawling, greater distance from person speaking, and increasing background noise) helps to improve sound reception, speech perception, and stimulation of the central auditory system. Such measures may promote development of auditory processing and perception and counteract speech problems.

The subject of CI provision for patients with UHL was first addressed in 2008 [68]. Improved sound localization [5, 23, 33] and speech comprehension in quiet conditions [29] and in noise as a result of squelch and head shadow effects [5, 23, 33, 65] as well as improved directional hearing as a result of loudness difference [29] have been demonstrated in adults and/or children.

However, the results reported in children by Hassepass et al. [23] cannot (yet) be generalized because of post-lingual deafness, the associated binaural hearing experience and brief hearing deprivation, the small group size, and learning effects in the test-retest model.

A less significant beneficial effect has been found for BAHA and CROS than for CI, but there are possibly pseudo-binaural benefits [5].

In the retrospective study reported here, unilateral hearing aid provision produced an objective and demonstrable binaural hearing gain in both directional hearing and quality of selective hearing in quiet and noisy surroundings as well as in speech comprehension in noise in more than 50% of cases.

Acceptance of wearing hearing aids among children with mild to moderate hearing loss has been reported as high in

Table 5 Unilateral hearing loss and treatment options (adapted from [20])

Type	Type of hearing loss	Possible intervention/treatment
A	Unilateral sensorineural hearing loss (SNHL) 30 to 60 dB	BTE or in-the-ear hearing aid on the impaired side
		Remote microphone systems (e.g., FM system) coupled to the hearing aid on the impaired side or as an open fitting of a receiver to the good hearing ear
		Sound field system (≥6 years for sound field system)
		Early hearing and speech intervention
		Optimizing classroom seating and classroom acoustics
B	Unilateral SNHL 60–80 dB	Attempt hearing aid fitting, including for hearing loss of 70 to 80 dB
		CROS provision in special cases from adolescence onwards
		Remote microphone systems (e.g., FM system) coupled to the hearing aid on the impaired side or as an open fitting of a receiver to the good hearing ear
		Sound field system (≥6 years for sound field system)
		Early hearing and speech intervention
		Optimizing classroom seating and classroom acoustics
B	Severe UHL and no adequate speech comprehension with hearing aid or unilateral sensorineural deafness (with auditory nerve intact)	Cochlear implant, irrespective of age
		Fitting of a conventional CROS hearing system only in special cases from adolescence onwards or as a "transcranial" CROS system with a high gain hearing aid or bone-anchored hearing aid (BAHA) (>5 years) on the impaired side
		Early hearing and speech intervention
		Optimizing classroom seating and classroom acoustics
C	Unilateral conductive hearing loss (aural atresia)	Bone conduction hearing system (trial fitting of hearing aid before end of 1st year of life)
		Later options: bone-anchored hearing aid, bone conduction implant, middle ear implant
		Early hearing and speech intervention
		Optimizing classroom seating and classroom acoustics
After hearing aid provision:		
Evaluation of hearing aid fitting by a pediatric audiologist, including feedback through structured interviews (questionnaires) from parents, teachers, early intervention specialists, and therapists and, if possible, from the child.		
When checking auditory gain, measurements with masking should be recorded wherever possible. The use of questionnaire systems is highly recommended. Hearing aid fitting and prescription should only be finalized once there is satisfactory acceptance of the hearing aid by both child and parent(s) and after a sufficiently long wearing period (aside from exceptional cases: all day).		

some cases [34, 36], and our retrospective study revealed high acceptance over all grades of hearing loss. The assessment of our study results carried out suggests that even mild hearing losses (from an auditory threshold of 30 to 40 dB) should have the opportunity for hearing aid provision. Children who have an auditory threshold between 21 and 29 dB should, like all other children with UHL, be followed up regularly by a pediatric audiologist.

Despite the considerable progress made in the technological development of hearing systems, speech intelligibility in background noise and sound localization will remain challenges until the problem of parallel amplification of desired sound and ambient noise is solved. Signal processing strategies that eliminate impairment of the cocktail party effect are currently being developed [39].

Quality of life

Health-related quality of life (HRQoL) is a multi-dimensional construct based on measurements obtained in the domains of physical and mental well-being, everyday functioning, and social integration [15]. Alongside the audiological diagnostic workup, measurement of HRQoL is relevant when deciding on a treatment and assessing its outcome.

The instruments used to measure HRQoL may be generic or disease-specific.

Changes in the HRQoL of adults with UHL reflect improved speech intelligibility in background noise and improved sound localization. The largest effect sizes have been associated with CIs, followed by bone conduction hearing aids and BTE hearing aids [37].

A validated HRQoL instrument exists only for bilaterally deaf adolescents [54]. Similar instruments are still lacking for all other hearing-impaired children and adolescents. Attempts are being made with KINDL, KIDSCREEN, and DISABKIDS to remedy this shortcoming for all other hearing-impaired children [50].

Whereas KIDSCREEN is used to survey HRQoL in healthy and ill children and adolescents, DISABKIDS surveys HRQoL in children and adolescents with chronic illnesses. The two projects—KIDSCREEN and DISABKIDS—have engendered a third ("Living with Hearing Disorders") which is setting out to develop a measure for HRQoL in hearing-impaired children and adolescents. The needs of—and the problems faced by—children provided with a hearing aid have been recognized [50].

An ideal instrument should be age-appropriate, contain brief and understandable questions, and reflect the following items: limitations in daily living due to hearing loss, benefit provided by the hearing aid in respect of those limitations, contribution to general quality of life made by the hearing aid, and general quality of life [19]. It is advisable to combine this with a questionnaire for parents and/or teachers; however, assessments offered in the parents' questionnaire merit critical scrutiny because parents are not necessarily good reporters of their child's quality of life [26].

Fundamental research

If it becomes established in the early sensitive period of childhood, aural preference is difficult to reverse [41]. Factors such as auditory deprivation and specific stimulation can lead to reorganization of the central auditory system, especially the auditory cortex [59].

The activity of the brain is increasingly being viewed from the perspective of functional integration and connectivity. The default mode network (DMN) denotes the network structures located in the temporal region that are functionally interconnected by intrinsic activity; the DMN is described as the "resting state network." Subareas of the DMN, as the baseline of the brain, show activity in the context of daydreams, empathy, and awareness of others' intentions [51]. Dysfunction of this intrinsic network plays a role in various conditions such as autism [16, 21], attention deficit hyperactivity disorder [16], and tinnitus [27]. UHL leads to reduced deactivation of the DMN during audio-visual tasks [62]. If self-awareness is not adequately suppressed during a task, the participant may become a "daydreamer." The academic and behavioral deficits seen in children with UHL may therefore be mediated by deficiencies in the DMN [62].

Outlook

Once a diagnosis of UHL has been made, the traditional view prevalent in otorhinolaryngology circles is that unilateral hearing is the minimum requirement for adequate speech development and consequently hearing aid provision is unnecessary. However, analysis of the published evidence, including the aspect of auditory deprivation and other neurophysiological facts, supports the contention that a hearing aid should be provided before or at the end of the first year of life for every child with UHL for whom this is feasible. "Feasibility" can be assessed after appropriate pediatric audiological diagnostic testing, in-depth counseling of the parents, and consideration of the resultant burden on the family [20].

In its updated 2013 guidelines, the American Academy of Audiology stipulates that children with aidable UHL should be considered candidates for amplification in the impaired ear [2]. The DGPP has also incorporated a similar statement into its consensus paper [20].

To reduce auditory deprivation and the extent of auditory cortex reorganization [41], early diagnosis and treatment of children with UHL should be an integral part of prospective studies from the outset.

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Authors' contributions A-KR and JF are joint principal authors who contributed equally to the retrospective study and to the literature review presented here. A-KR, JF, FM, and TW participated in the concept, design, and interpretation of the manuscript. JF and A-KR coordinated the study centers and patient inclusion, and JF was responsible for the practical conduct of the retrospective study. MH, TW, and MR provided the patients for the study, and ABr and AS were responsible for diagnostic testing. JF, FM, A-KR, and TW conceptualized the data analysis. JF, ABo, and TW developed the questionnaires for the retrospective study. A-KR, FM, TW, and JF drafted the manuscript, and all the listed authors have reviewed and approved the final version of the manuscript as submitted.

Compliance with ethical standards

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Ethical approval The study was approved by the ethics committee of the Hamburg regional medical board. All procedures performed in this study involving human participants were in accordance with the ethical standards of the ethics committee of the Hamburg regional medical board.

Informed consent Informed consent was obtained from all individual participants included in the study or their parents/legal guardians.

Appendix

Table 6

Agenda (modified from [4])

To-do list for children with UHL

- The cause of UHL should be investigated in the same way as in a child with bilateral hearing loss. CMV testing should not be omitted. Newborn examination by an ophthalmologist should rule out any associated sensory syndrome and should ensure that, if perception of acoustic signals is reduced, at least visual signal perception is possible. Examination of the child should exclude reduction of peripheral vision (ophthalmologist) but also any disorder of visual perception (orthoptist).
- If a vestibular aqueduct syndrome is detected by CT or MRI, situations should be avoided that carry a risk of head injury (e.g., sports such as soccer) or pressure changes (e.g., scuba diving).
- Early provision of an FM system, hearing aid, CROS hearing aid, and bone conduction hearing aid. Provision must be tailored to the individual child.
- Attendance for regular pediatric audiological checks is advisable to pick up changes in hearing ability, including any bilateral hearing loss that may be developing.
- Otitis media with effusion or blockage of the auditory canal with earwax can contribute to a temporary worsening of the existing hearing impairment.
- A more active approach should be adopted for treating recurrent and chronic otitis media with effusion. Hearing in the affected ear—as well as that in the good ear—should not be additionally made worse.
- Ensure regular information exchanges with parents, preschool/school teachers, and therapists about speech and language development as well as the child's academic performance, providing access to the latest literature and current knowledge about UHL and risks.
- Preserve the child's existing hearing: ear defenders at loud events (concerts, fireworks). Set an appropriate volume level when using an iPod or MP3 player.
- Provide good counseling to children and adolescents as they grow up. They should take "primary responsibility" for their hearing. Dangerous Decibels (virtual exhibit) is an example of how knowledge can be imparted (http://www.dangerousdecibels.org/exhibit/virtual-exhibit/).

To-do list for children with undetected UHL

- UHL should be investigated by a pediatric audiologist if there are any abnormalities in language development, attention and concentration, school development, or social behavior.

Prevention

- Enrolment in hearing and speech screening programs is advisable in early childhood through to starting school as this screening may pick up hitherto undetected UHL and delayed or impaired speech development.
- Consider including schoolchildren in screening programs designed to detect academic problems early.
- Some minor anomalies, such as delayed or absent babbling phases, are often overlooked.

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