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Hearing-related quality of life, developmental outcomes and performance in children and young adults with unilateral conductive hearing loss due to aural atresia

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ABSTRACT

Background: While research has shown that children with single sided deafness have a lower quality of life and developmental outcomes compared to normal hearing peers, little is known about these domains in children with unilateral congenital conductive hearing loss due to aural atresia.
Objectives: This study aims to investigate the hearing-related quality of life, developmental outcomes and educational performance in children and young adults with unilateral conductive hearing loss due to aural atresia.
Methods: Nineteen children and young adults with unilateral aural atresia received a set of five questionnaires. Hearing-related quality of life (SSQ), general quality of life (Kidscreen-27), speech and language development (CCC-2-Nl), educational performance and problems in social-emotional and behavioral domains (CBCL/YSR/ASR) were measured with validated questionnaires. Scores on the questionnaires were compared to their norm scores. Mann-Whitney U tests and independent t-tests were used to identify significant differences between age groups.
Results: Mean scores on the SSQ subscales were speech 6.78, spatial 5.00 and quality 6.98. Mean scores on the Kidscreen-27, CCC-2-NL, CBCL/YSR/ASR fell within normal or non-clinical range. A high number of cases needed speech therapy (60.7%) or special measures in class (79.3%) or showed grade repetition (>30%) in primary or secondary school.
Conclusion: Children and young adults with unilateral conductive hearing loss due to congenital aural atresia showed lower scores regarding hearing-related quality of life compared to normal-hearing peers. The result show similarities with children with single sided deafness. Regarding general quality of life, speech and language development and in social-emotional and behavioral domains the studied children and young adults seem to develop according to norm scores. It is important to observe these children closely as they may need guidance during education to allow them to thrive.

1. Introduction

Congenital aural atresia (CAA) is defined as a partial or complete lack of development of the external auditory canal, which results in varying degrees of middle ear malformation [1,2]. In most cases CAA is accompanied by a congenital malformed pinna known as microtia. Reported prevalence of CAA varies with ranges between 1 in 10,000–20,000 births [3]. In many cases it is part of a craniofacial microsomia or presents in association with other deficits such as in Goldenhar' s syndrome, Treacher Collins, and trisomy 21 [2].

Levels of hearing loss differ, though, in most cases a purely

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Abbreviations

ASR	Adult Self-Report
CAA	Congenital aural atresia
CBCL	Children's Behavior Checklist
CCC-2-NL	Dutch version of the Children's Communication Checklist
dB	Decibel
dB nHL	Decibel above normal hearing level
IQR	Interquartile range
SD	Standard deviation
SNHL	Sensorineural Hearing Loss
SSQ	Speech, Spatial and Quality of Hearing Scale
YSR	Youth Self-Report

conductive hearing loss is seen at the affected side by the deformities of the middle and/or outer ear. Current literature demonstrates that the impact of this loss on daily life is understudied. The vast amount of literature about consequences of unilateral hearing deficits is based on cases with unilateral sensorineural (severe to profound) hearing loss (SNHL). Patients subsequently suffer from the resulting impaired ability for speech perception in noise and sound localization abilities. In children this can result in difficulties in daily life and developmental disorders such as language delays [4]. Regarding educational performance, unilateral SNHL in children is related to increased rates of grade failure, a need for speech therapy and additional educational assistance [4–6]. In addition, children with unilateral SNHL have been found to have a lower quality of life compared to normal hearing peers [7].

The influence on daily life and social-emotional development of unilateral conductive hearing losses caused by aural atresia is unclear. These children differ from those with single sided deafness as the magnitude of hearing loss is mostly less profound. Secondly, they still hear their own voice in the affected ear resulting in stimulation of the central auditory system on the affected side, and thirdly, bone conduction devices can overcome the conductive hearing deficit in most cases. So far, only a few studies have investigated educational performance in children with aural atresia [8–10]. A higher need for individualized education plans and speech therapy was seen [8–10]. Though, evidence is lacking on hearing-related quality of life or the language, educational or social-emotional development of these children. As a result there is an ongoing debate about whether the use of hearing amplification or guidance of children with unilateral congenital aural atresia needs to be considered to overcome the deficits in young life. A better knowledge and understanding of the situation may help to provide parents with a clearer image of what they could expect of their children. Therefore, we aim to investigate the hearing-related quality of life of children with unilateral aural atresia. Additionally, general quality of life, speech and language development, educational performance, and social-emotional and behavioral problems will be investigated prospectively in a cohort of children with CAA.

2. Method

2.1. Ethical consideration

This study has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki). The ethics committee of the University Medical Center Utrecht (UMCU) declared that no formal approval of the detailed protocol was needed according to the Dutch Medical Research Involving Human Subjects Act (No.14–850/C).

2.2. Study design

This study had a cross-sectional cohort design and included children and young adults with unilateral conductive hearing loss due to congenital aural atresia. The study took place at the department of Plastic Surgery and Otorhinolaryngology, including the Center of Audiology of the Wilhelmina Children's Hospital Utrecht, the Netherlands, as a center of expertise for children with craniofacial microsomia.

2.3. Study population

To assembly the cross-sectional cohort, children and young adults with unilateral conductive hearing loss due to aural atresia were selected. Patients were recruited from the patient database of the department of Plastic Surgery of the Wilhelmina Children's Hospital Utrecht after historical consultation for reasons of microtia. In order to participate in this study, patients were required to meet all of the following criteria: 1) between six and 20 years of age at the time of the study enrolment, 2) have Dutch as primary language, 3) have single-sided congenital aural atresia (codes Q16.0 to Q16.4 in the International Classification of Diseases and Related Health Problems), 4) have a hearing loss of minimal 40 dB nHL with perceptive hearing level maximum of 20 dB nHL (1000–4000 Hz) on the atretic side measured during the latest hearing assessment in an audiological center, 5) at the same time having a hearing level of 20 dB nHL maximum (1000–4000 Hz) on the contralateral (non-atretic) side, and 6) have given informed consent, either themselves or their caregivers (depending on age), to participate in the study. This study also included children with syndromes that met the inclusion criteria. As such, there were no criteria that led to the exclusion of a participant.

2.4. Study enrolment

Out of the database of 381 patients that was used to include participants, 161 children and young adults were selected based on the eligible age criteria to participate in the study. Recruitment of participants took place from December 2017 to April 2018. Participants were contacted by the researchers by postal mail, in which participants were informed about the study and asked for consent for study participation. After consent of the child (and parents), children were included in the study after verifying to reach the inclusion criteria. Questionnaires to assess outcomes were then sent by postal mail. After three weeks without returning the questionnaires, participants were contacted by phone and reminded to fulfil the study requirements. Participants were marked as a drop-out after six weeks without a response.

2.5. Study procedures

Participants were divided into age groups at time of enrolment; 6 up to 11 years of age, 11 up to 16 years of age, and 16 up to 21 years of age. A single package of questionnaires specified per age group was used to assess demographic variables and outcomes.

2.6. Outcome measures

Demographic data consisted of participants 'age sex, multilingualism and medical conditions or syndromes and level of education of legal custodians (divided into professional education, university of applied sciences and university).

2.7. Hearing related Qol

To assess the primary outcome of this study the hearing-related quality of life (QoL) measured by the validated Speech Spatial and Qualities of Hearing Scale (SSQ) was used [11]. The SSQ consists of 24

VAS-scales, scoring 0 (not at all) to 10 (perfect), covering the domains speech, spatial and quality of hearing. The mean score of the three subdomains are then compared to norm scores [11]. For children of 6 up to 15 years, a parental version was used (adapted by Karyn Galvin and previously translated to Dutch by Liesbeth Royackers Labo Exp ORL, Leuven). Participants of 16 years old and above completed the SSQ themselves [11].

2.8. General Qol, language development, educational performance and social emotional and behavioral outcome

As secondary outcome general quality of life, language development, educational performance, and social-emotional and behavioral problems were assessed. General quality of life was measured by the Dutch version of the Kidscreen-27[12] This is a validated questionnaire on five subcategories (physical well-being, psychological well-being, autonomy & parent relation, peers & social support and school environment), covered by 27 questions with a 5-point rating scale (ranging from not at all/never to extremely/always). Scores were calculated by summing up the scores of the subscales, converting them to a Rasch score and converting those Rasch scores to a T-score. Participants of 11 years and older filled in the questionnaire themselves. For children younger than 11 years old, the Kidscreen-27 was filled in by parents. Scores were compared to the available norm scores [12].

Language development for children between 6 and 16 years old was measured with the Dutch version of the Children's Communication Checklist (CCC-2-NL) [13]. The CCC-2-NL is a parental questionnaire and divided into ten categories: speech, syntax, semantics, coherence, initiation, stereotypical language, use of context, non-verbal communication, social relations and interests. In addition, the CCC-2-NL provides three communication scales for general communication, social interaction and pragmatics. The CCC-2-NL. In total, the CCC-2-NL consists of 70 4-point frequency scales, ranging from never to always. The scores on the items are added together per subscale and are then converted to a standard score. Their totals were compared to the norm scores of the CCC-2-NL [13], for which higher scores mean lower ability on the respective subscale. Participants of 16 years of age and older were not evaluated on speech and language development by the lack of a validated questionnaire to assess this outcome.

To investigate the educational performance of the participants, questions were covering aspects of the occurrence of grade retention, as well as the need for special education, speech therapy, special measures in class, educational assistance and hearing amplification during their lifetime of the participants.

Lastly, to assess behavioral, emotional and social problems the Children's Behavior Checklist (CBCL) was used for participants between 6 and 18 years old [14]. The CBCL is a validated parental questionnaire which consists of 113 statements with 3-point rating scales (ranging from not true to often true), where higher scores denote higher risk of behavioral problems [14]. The items on the CBCL correspond with several subdomains: withdrawn, somatic complaints, anxious/depressed, social problems, attention problems, delinquent behavior and aggressive behavior. For participants between 6 and 18 years old, the CBCL was filled in by parents. Additionally, participants between the age of 11 and 18 years old filled in the Youth Self-Report (YSR), which is a self-report version of the CBCL. 18 to 20-year-old participants filled in the Adult Self Report (ASR), which is an adult version of the CBCL. The total scores of these subdomains were compared to their norm scores. These norm scores are age specific. Scores on the CBCL and YSR/ASR were calculated with a scoring program, ADM version 9.1.

2.9. Data analysis

All statistical analyses were conducted using IBM SPSS software version 24.0. Descriptive statistics were used to describe the characteristics of the participants. Differences between the means of SSQ subscales and age groups were calculated using a one-way ANOVA. In case of a significant difference, an independent *t*-test was used to identify the difference between groups. Quantitative data of the secondary outcomes of the Kidscreen-27, CCC-2-NL and the CBCL/YSR/ASR questionnaires were compared to their norm scores. Differences between age groups were calculated using a Kruskal-Wallis test. When a Kruskal-Wallis test returned a significant difference, a Mann-Whitney *U* test was used to identify which groups differed from each other. Differences between subscales were calculated with a Wilcoxon signed rank test (with not-normal distribution) or a paired *t*-test (with normal distribution).

. Data regarding educational performance, the use of speech and language therapy and the use of hearing amplification are descriptive. This study was reported according to the STROBE statement [15].

3. Results

3.1. Participants

Fig. 1 shows the flowchart of the study participants. Out of 370 patients included in the database of patients with microtia 161 participants were selected by age criteria (6–20 years at time of assessment), and invited to participate in the study. Of those, 37 patients did meet the inclusion criteria and returned the informed consent. These patients were sent questionnaires to assess outcome. In total, 29 participants returned the questionnaires of which data was used for analysis of outcomes. The demographic characteristics of these study participants are summarized in Table 1. Of the 29 participants, the majority was male (18 out of 29; 62.1%) with age ranging between 7 and 19 years old. Numbers of participants were equally divided between age groups. Twelve out of 29 children (41.4%) were diagnosed with a syndrome or medical condition, including Goldenhar syndrome (n = 2), Attention Deficit Disorder (n = 1) and Auriculo-condylar syndrome (n = 1).

3.2. Educational performance

School performance characteristics are described in Table 1. Ten out of 29 participants (34.5%) repeated a class (primary school: 3 times year 1, 1x year 2, 1x year 3, 4x year 4; secondary school: 1x year 6). Two participants (6.9%) were in special education programs. Seventeen participants (60.7%) have needed speech therapy. 23 participants (79.3%) have needed special measures in class, of which seven needed multiple measures. Measures mentioned included orientation with their best ear towards the teacher, a special position in front of the class and educational assistance. One participant was brought up multilingually. Thirteen out of 29 participants (44.8%) have made use of hearing amplification (11 patients used a bone conduction device (with (n = 3) or without (n = 8) solo equipment), 1 patient only solo-equipment, 1 patient hearing aid and solo-equipment)). Of those children repeating grades, 10 out of them used hearing amplification at a prior age (duration and age at which amplification was used was not known).

3.3. Hearing-related quality of life

Scores on the primary outcome of hearing-related QoL measured with the SSQ questionnaire were calculated in subscales (Table 2). For all children together, participants scored significantly lower on the spatial subscale (mean(SD) 5.08(1.99)) compared to the speech (mean 6.86(1.38), Z −4.206, $p < 0.001$) and quality of hearing (mean 7.21 (1.51), Z −4.314, $p < 0.001$) subscales. Moreover, scores on the speech subscale were lower than on the quality of hearing subscale (Z −2.482, $p = 0.013$). Sex, use of hearing amplification and multilingualism were not to affect SSQ scores. Within subscales, no significant differences were found between age groups.

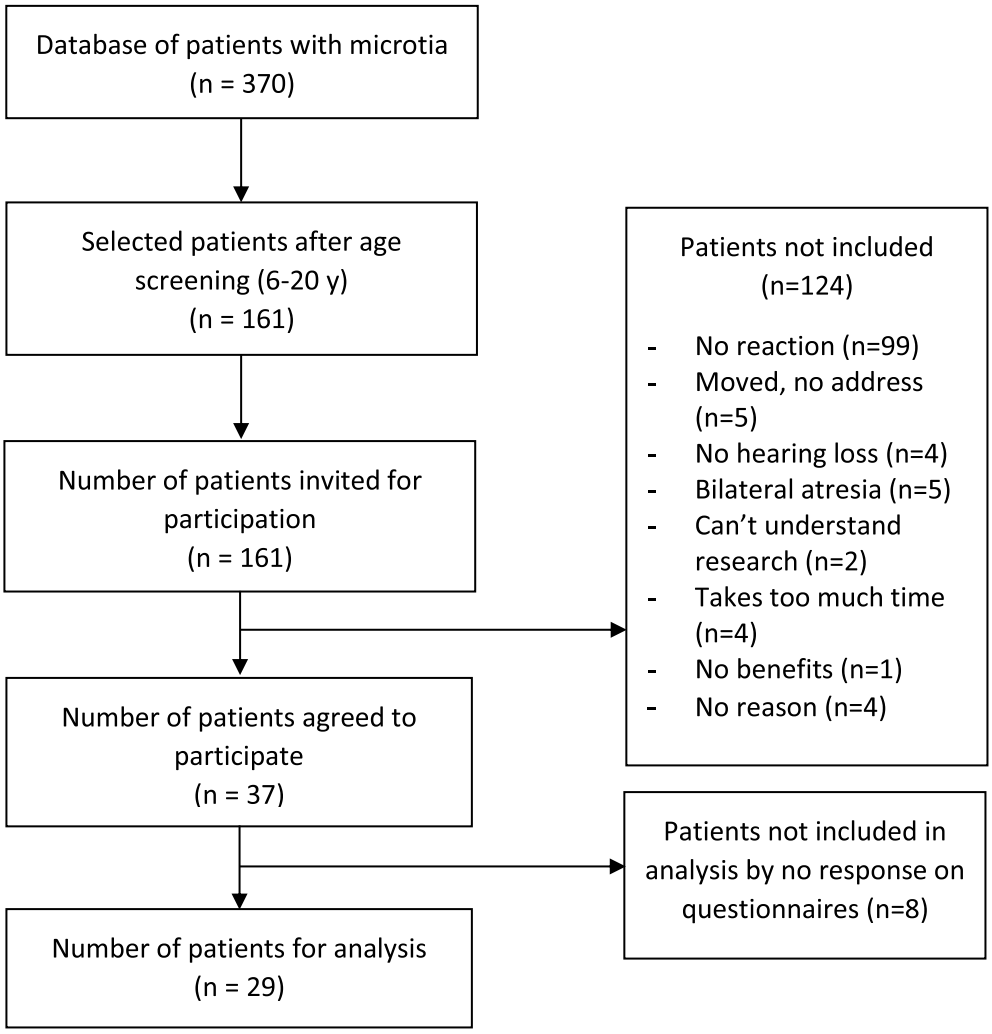

Fig. 1. Flow diagram of the study participants.

3.4. General Qol

Mean T-scores on the Kidscreen-27 assessing general QoL are summarized in Table 2. Almost all mean subscale scores were within normal ranges. Only for the physical well-being subscale, the youngest age group scored 1.2 standard deviations above the norm (mean(SD) 62.79 (8.57)). This group scored significantly higher to age group of 11–15 years of age (52.96(7.55), Z −2.428, p = 0.015) and age group 16–20 (45.49(9.50), Z −2.697, p = 0.007). The middle and highest age groups did not differ significantly in outcome (Z −1.676, p = 0.094) on this subscale.

3.5. Language- and social-emotional problems

The outcome of language development and communication scores measured by the CCC-2-NL are summarized in Table 3. Mean scores on the ten language development subscales fell within one standard deviation of the norm. Mean scores for the communication scales fell within normal range as well: general communication fell between the 70th and 75th percentile, social interaction fell between the 45th and 60th percentile, and pragmatics fell between the 70th and 75th percentile. No significant differences were found between the age groups on any subscale of the CCC-2-NL. On the semantics subscale of the CCC-2-NL, children who grew up in a highly educated environment (mean(SD) 8.40 (2.61)) scored significantly lower compared to a medium (11.73 (2.61), t 2.365, 95% CI 0.070–6.585, p = 0.046) and lower educated environment (12.80(1.10); t 3.479, 95% CI 1.215–7.585, p = 0.0016). On the general communication subscale, children who grew up in a highly educated environment (73.60(13.61)) scored significantly lower compared to a lower educated environment (103.40(13.16), t 3.519, 95% CI 10.267–49.333, p = 0.008). On the pragmatics subscale, children who grew up in a highly educated environment (37.40(7.64)) scored significantly lower compared to a low educated environment (52.20(6.22), t 3.360, 95% CI 4.570–25.030, p = 0.011).

Outcomes of social-emotional- behavioral problems of participants assessed by the CBCL, YSR and ASR questionnaires are summarized in Table 4. All mean scores fell within non-clinical range. Though, a difference was found between age groups on the attention-seeking behavior subscale of the CBCL: the youngest group showed higher scores (mean(SD) 60.45(5.32)) than the oldest group (55.00(2.65), Z −1.977, p = 0.048). Additionally, a difference was found between age groups on two subscales of the YSR/ASR. On the withdrawn/depressed subscale the older group showed higher scores (60.43(6.08)) than the middle group (52.10(2.60), H 6.625, p = 0.010). On the delinquent behavior subscale the older group showed higher scores (52.57(1.13)) than the middle group (51.10(2.42), H 5.195, p = 0.023).

4. Discussion

In this study we investigated the hearing-related quality of life, developmental outcomes, general quality of life and school performance characteristics of children and young adults with unilateral conductive

Table 1
Demographic and school performance characteristics of participants (N = 29) in frequency and (percentages) or (IQR; interquartile range) when indicated. Because one participant refused to fill in school performance characteristics, a part of the outcome was based on n = 28 as indicated by[a].

	Total	Age 6–10 (N = 11) (%)	Age 11–15 (N = 11) (%)	Age 16–20 (N = 7) (%)
Age (yrs) (median) (IQR)	12.8 (6.6)	9.1 (2.3)	13.4 (3.4)	17.8 (2.3)
Sex				
Male	18 (62.1)	8 (72.7)	5 (45.5)	5 (71.4)
Female	11 (37.9)	3 (27.3)	6 (54.5)	2 (28.6)
Repeating of classes	10 (34.5)	3 (27.3)	4 (36.4)	3 (42.9)
Use of special education	2 (6.9)	0 (0.0)	1 (9.1)	1 (14.3)
Language therapy[a]	17 (60.7)	6 (54.5)	5 (45.5)	6 (100.0)
Use of hearing amplification[a]	13 (46.4)	7 (63.6)	3 (27.3)	3 (50.0)
Use of special measures in education[a]				
No	5 (17.9)	3 (27.3)	1 (19.1)	1 (16.7)
Yes	16 (57.1)	6 (54.5)	6 (54.5)	4 (66.7)
Yes, multiple	7 (25.0)	2 (18.2)	4 (36.4)	1 (16.7)
Highest level of education[a]				
Professional education	10 (35.7)	1 (9.1)	5 (45.5)	4 (66.7)
Applied sciences	12 (42.9)	7 (63.6)	4 (36.4)	1 (16.7)
University	6 (21.4)	3 (27.3)	2 (18.2)	1 (16.7)
Multi-lingualism	1 (3.4)	1 (9.1)	0 (0.0)	0 (0.0)
Presence of syndrome	12 (41.4)	6 (54.5)	4 (36.4)	2 (28.6)

Table 2
Hearing related QoL SSQ subscale scores in means (SD) and general QoL Kidscreen-27 scores in mean T-scores (SD). Data provided per subscale in total and by age groups. Mean norm score of the Kidscreen-27 is 50 (SD = 10).

	Total	Age 6-10	Age 11-15	Age 16-20
SSQ (N = 29)				
Speech	6.86 (1.38)	7.07 (1.01)	7.06 (1.63)	6.21 (1.44)
Spatial	5.08 (1.99)	5.12 (1.52)	5.10 (2.05)	4.99 (2.75)
Quality	7.21 (1.51)	7.54 (1.36)	7.55 (1.42)	6.17 (1.57)
Kidscreen-27 (N = 28)				
Physical	55.22 (10.56)	62.79 (8.57)	52.96 (7.55)	45.49 (9.50)
Psychological	49.24 (7.47)	49.77 (5.48)	51.11 (9.19)	44.84 (6.38)
Parents	52.45 (6.33)	51.72 (8.41)	51.36 (5.24)	55.68 (3.12)
Peers	50.52 (8.31)	48.75 (8.01)	54.77 (8.74)	46.67 (5.68)
School	54.44 (7.73)	56.61 (9.72)	54.58 (6.80)	50.18 (3.09)

hearing loss due to aural atresia.

The mean scores of the hearing related QoL subdomains (speech 6.9; spatial 5.1; quality 7.2) were low compared to data of normal hearing children out of the literature (respectively 9.0; 8.5; 9.0) [16]. Though, outcome was relatively similar to reported scores of children with SSD (respectively 8.0; 3.0; 8.0) [17]. This outcome is to be expected in cases of moderate to severe hearing loss of both conductive and perceptive origin. Regarding the unilaterality of their hearing loss, both groups will experience impaired speech perception in noise and sound localization abilities [18,19].

In this study, mean scores for general quality of life (Kidscreen-27), language development (CCC-2-NL) and the social-emotional-behavioral assessment (CBCL, YSR/ASR) fell within normal ranges. This suggests that these children develop similarly to normal-hearing peers in these domains. Some differences were found in outcomes between age groups.

Table 3
Mean standard scores on the CCC-2-NL items in total and by age group (N = 21).[a] mean norm is 10.[b] norm range 44–122; 80 corresponds to 50th percentile.[c] norm range −19 to 19; 0 corresponds to 50th percentile.[d] norm range 19–63; 40 corresponds to 50th percentile.

	Total	Age 6-10	Age 11-15
Language			
Speech[a]	10.95 (3.71)	10.45 (3.24)	11.50 (4.28)
Syntax[a]	10.86 (3.09)	11.36 (3.41)	10.30 (2.75)
Semantics[a]	11.19 (2.79)	11.91 (2.74)	10.40 (2.76)
Coherence[a]	11.24 (2.79)	11.27 (2.72)	11.20 (3.01)
Initiation[a]	10.10 (2.93)	10.36 (2.87)	9.80 (3.12)
Stereotypical language[a]	11.48 (3.09)	10.73 (3.13)	12.30 (2.98)
Use of context[a]	11.71 (3.05)	11.73 (2.61)	11.70 (3.62)
Non-verbal[a]	11.48 (2.56)	11.45 (2.95)	11.50 (2.22)
Social relations[a]	10.33 (2.69)	10.18 (3.13)	10.50 (2.27)
Interests[a]	10.76 (2.43)	11.45 (1.75)	10.00 (2.91)
Communication			
General communication[b]	89.24 (18.78)	89.73 (19.09)	88.70 (19.44)
Social Interaction[c]	−1.52 (9.28)	−1.45 (8.50)	−1.60 (10.54)
Pragmatics[d]	44.76 (9.30)	44.27 (9.90)	45.30 (9.09)

Table 4
Mean T-scores and (standard deviations) on the CBCL (N = 16) and YSR/ASR (N = 13) in total and by age group. Mean norm is 50 (SD = 10). A score above 68 denotes clinical range.

		Total	Age 6-10	Age 11-15	Age 16-20
Anxiety	CBCL	54.96 (6.79)	57.00 (8.60)	54.20 (4.66)	50.00 (0.00)
	YSR/ASR	54.00 (4.65)		53.20 (4.47)	55.14 (5.01)
Withdrawn/Depressed	CBCL	56.13 (6.33)	56.73 (7.28)	54.30 (5.31)	60.00 (5.20)
	YSR/ASR	55.53 (5.96)		52.10 (2.60)	60.43 (6.08)
Somatic	CBCL	57.50 (6.88)	55.91 (7.44)	58.50 (6.47)	60.00 (7.21)
	YSR/ASR	57.12 (9.48)		58.60 (12.13)	55.00 (3.16)
Social	CBCL	54.58 (5.45)	56.45 (6.41)	53.40 (4.55)	51.67 (2.08)
	YSR/ASR	53.82 (4.85)		51.80 (2.10)	56.71 (6.29)
Thoughts	CBCL	58.04 (7.54)	59.82 (7.52)	57.00 (7.80)	55.00 (7.81)
	YSR/ASR	55.00 (5.91)		54.50 (5.52)	55.71 (6.80)
Attention seeking	CBCL	56.58 (5.66)	60.45 (5.32)	52.80 (3.74)	55.00 (2.65)
	YSR/ASR	54.06 (6.88)		53.80 (8.65)	54.43 (3.69)
Delinquent behavior	CBCL	53.17 (3.63)	54.91 (4.32)	51.30 (1.57)	53.00 (3.61)
	YSR/ASR	51.71 (2.09)		51.10 (2.42)	52.57 (1.13)
Aggressive	CBCL	54.63 (5.99)	57.55 (7.05)	51.90 (3.57)	53.00 (4.36)
	YSR/ASR	51.41 (3.43)		51.30 (4.11)	51.57 (2.44)

The group of 6–10 years old scored significantly higher compared to the older participants on the physical well-being subscale (Kidscreen-27). An explanation could be that parents of young children with aural atresia are confronted with many uncertainties about the development, appearance, and psychological outcome of their young child [20]. This might enhance being more watchful for their young atretic child's health [21]. In line with this, the youngest group showed higher scores on attention-seeking behavior. A watchful parent might give more attention to their child, rewarding them more often than they would other children, and thereby reinforces the child's need for more attention [22]. On the other hand, higher scores on the withdrawn/depressed and

delinquent behavior subscales of the CBCL/YSR/ASR were seen in the older group. If this could be explained by their awareness of being different to their peers or reluctance in communication by experienced difficulties is unclear [23]. However, all mean scores still fell within non-clinical range. This raises questions about the validity and relevance of these findings. It is possible that any actual differences do not present themselves due to the wide range of the norms and the limited size and selection of the study population. Additionally, while the differences that were found were significant, all mean scores fell within normal or non-clinical range. It could be discussed whether the effects thereby are of any clinical importance. A tendency towards certain behavior is observed at most, but due to the small number of participants any found tendencies are tentative at best. Therefore, the results of this study imply that children with unilateral conductive hearing loss due to aural atresia develop normally compared to normal-hearing peers.

While the literature on the outcome and performance of children with unilateral congenital aural atresia is limited [8–10], some comparisons can be made. Notably, while aural atresia has been suggested to have a detrimental effect on general quality of life [24], no such effect has been found in the current study. Likewise, the current study did not find an adverse effect of aural atresia on language development. Still, it must be taken into consideration that 45% of the participants of this study received hearing amplification at a prior age and the majority needed speech therapy or additional measures in class. Secondly, >30% repeated a class, of which the vast majority in the first years of primary school. For example, 4/29 children scored repetition of grade 4 (age 7–8 years of age) of primary school. This number exceeds norm values [25] (e.g. around 5% of Dutch children repeat the class they are in around age 7). Similar results in children with SNHL are explained by the encountered language problems [4–6], although scores of speech and language development in our study were comparable to norm scores.

As mentioned, our study has some limitations. A small number of participants was included. This can be expected considering the fact that microtia/aural atresia is a rare disease with limited numbers of affected people. Though, partly this will be due to the single center design of the study and selection of participants after invitation and confirmation of participation. The response rates of this study (39%) could have been improved with the use of pre-inclusion contact via telephone or e-mail [26]. Secondly, selection bias will be introduced by the recruitment of participants with microtia who historically visited the department of Plastic Surgery as a center of expertise in craniofacial microsomia. In the future, a prospective study on this group of children is advised.

5. Conclusion

Children with unilateral aural atresia are suggested to have a lower hearing-related quality of life compared to normal-hearing peers and show a higher need for speech therapy, educational assistance and repetition of classes. Regarding language development and in social-emotional and behavioral domains, however, children and young adults with aural atresia seem to develop according to norm scores. Due to the small number of participants included in this study, any significant differences that were found have to be confirmed in future studies to ascertain their veracity.

Children with unilateral conductive hearing loss due to aural atresia seem to encounter similar educational problems as children with SNHL. It is therefore prudent to pay attention to children with aural atresia regarding their educational and general development.

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Declaration of competing interest

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