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Systematic Review
Craniofacial Anomalies

Hearing impairment and ear anomalies in craniofacial microsomia: a systematic review

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Abstract. The aim of this systematic review was to review the literature on hearing impairment and ear anomalies in patients with craniofacial microsomia and to determine their prevalence. Sixty-two records including 5122 patients were included. Ear anomalies were present in 52–100% of patients. The most reported external ear malformations were microtia, pre-auricular tags, and atresia of the external auditory canal. Ossicular anomalies were the most reported middle ear malformations, whereas the most reported inner ear malformations included oval window anomalies, cochlear anomalies, and anomalies of the semicircular canals. Hearing loss in general was reported in 29–100% of patients, which comprised conductive hearing loss, mixed hearing loss, and sensorineural hearing loss. Between 21% and 51% of patients used hearing aids, and 58% underwent a surgical intervention to improve hearing. The relationship between different phenotypes of craniofacial microsomia and the type and severity of hearing loss is mostly unclear. In conclusion, the high prevalence of ear and hearing anomalies in patients with craniofacial microsomia underlines the importance of audiological screening in order to facilitate individual treatment.

Keywords: craniofacial microsomia; Goldenhar syndrome; hearing loss; ear; congenital microtia.

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Craniofacial microsomia (CFM) is one of the most common congenital malformations resulting in facial asymmetry, with an incidence of 1:3000 to 1:26,000 live births[1–5]. It affects structures derived from the first and second pharyngeal arches, manifesting as unilateral or bilateral underdevelopment of the ear, mandible, orbit, facial nerve, and/or soft tissues. In addition, extracraniofacial malformations, including cardiac and urogenital anomalies, and functional anomalies, including obstructive sleep apnoea and feeding difficulties, may be present[6–10].

A common characteristic of CFM is underdevelopment of the outer ear, ranging from microtia to anotia and aural atresia[11–13]. In addition, malformations of the middle and inner ear have been described, including the absence of oval and round windows, malformed auditory

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ossicles, hypoplasia of the cochlea, and a short or wide internal auditory canal[13,14]. Consequently, these ear anomalies may cause varying types and degrees of hearing loss[12,15].

Early recognition of ear and hearing anomalies is important, as a timely diagnosis of treatable anomalies may lead to early intervention, thereby preventing secondary developmental disorders regarding speech and language development[12,15–18]. In order to develop screening and treatment protocols for these patients, in addition to existing newborn hearing screening in most countries, it is important to determine the prevalence of ear and hearing anomalies, and to investigate if patients at risk of hearing impairment can be identified. Therefore, the aim of this review was to describe the types of auricular anomalies and hearing impairment in patients with CFM, and to determine their prevalence.

Methods

Search strategy

This systematic research was guided by the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement[19]. A systematic search of the literature was conducted to identify records on auricular anomalies and/or hearing impairment in CFM. The search was conducted within the databases Embase, PubMed, MEDLINE, Ovid, CINAHL EBSCO, Web of Science, Cochrane, and Google Scholar. The databases were searched from inception until May 26, 2021. The search strings were constructed using a combination of keywords and Boolean operators for all databases. For the same query, different index terms (e.g. MeSH terms, Emtree terms) and syntax may be used between databases in order to yield similar results. The full search string of each database is included in **Supplementary Material** Appendix A. A list of synonyms for CFM used in the search is included in **Supplementary Material** Appendix B. Additionally, a manual reference list search of the included records was performed to identify additional records relevant to the study.

Two researchers (P.A.E.T. and W.R.) independently screened the titles and abstracts of all records for relevance. Records without an abstract underwent a full-text review. Irrelevant records were excluded. After initial selection on title and abstract, the full texts of relevant records were retrieved. Full-text records were assessed for eligibility based on predetermined inclusion and exclusion criteria. The inclusion criteria were (1) original research reporting auricular anomalies and/or hearing tests in CFM patients, and (2) written in the English or Dutch language. Exclusion criteria were (1) case reports, conference abstracts, letters, notes, and editorials, (2) animal studies, and (3) studies including <10 CFM patients. Studies only describing patients with isolated microtia (i.e. microtia without any other manifestation signifying CFM) were excluded.

Data extraction

The following data were extracted from the included records: type of study; number of included patients; types of auricular anomaly; prevalence of auricular anomalies; prevalence and type of hearing impairment; treatment modalities used for auricular anomalies and/or hearing impairment; correlations or associations between auricular anomalies and hearing impairment, and between clinical manifestations of CFM and auricular anomalies or hearing impairment. Where possible, patients with isolated microtia were left out of the analysis. The Oxford Centre for Evidence-Based Medicine (CEBM) criteria were used to grade all studies on quality of evidence.

Results

Study selection

In total, 3977 records were identified through the initial literature search and after the manual reference list search. Following the removal of duplicates, 2509 records were screened for relevance. After initial screening, 2372 records were excluded, resulting in 137 records for full-text assessment against the eligibility criteria. Finally, 62 records remained and were included for analysis. All 14 records identified through reference list searching were included after full-text assessment (Fig. 1).

Study characteristics

Of the 62 records included, 21 were case series[13,14,20–38], 30 were retrospective studies[5,12,15,39–65], 10 were prospective studies[16–18,66–72], and one was both retrospective and prospective[73] (**Supplementary Material** Table S1). Seventeen studies included patients with isolated microtia[16–18,28,30,36,41,44,45,64,66–72]; these patients were excluded from further analysis where possible. In total, 5122 patients with CFM were analysed in this literature review. However, 14 patients were analysed twice, once by Bisdas et al.[13] and once by Hennersdorf et al.[43]. In addition, Werler et al.[71], Collet et al. (2011)[16], Heike et al.[18], Speltz et al. (2017)[69], Wallace et al.[70], and Collett et al. (2019)[17] analysed patients who were part of a multicentre longitudinal study from craniofacial centres across the United States and Canada. Since these records related to different phases of that study, they were all included. Siebold et al.[67], Luquetti et al.[66], and Speltz et al. (2018)[68] investigated the same cohort of 108 patients referred to as the 'Craniofacial microsomia: Longitudinal Outcomes in Children pre-Kindergarten' (CLOCK) study. Johns et al.[72] analysed 89 patients from the same cohort at a later time; 19 patients from the original cohort were lost to follow-up. Several authors described external ear anomalies in patients from the same retrospective multicentre cohort study[6,63,74–76]. However, to avoid a repetition of the same results, only the most recent record describing this cohort was used for further analysis[63].

External ear malformations

External ear malformations were described in 50 records (**Supplementary Material** Table S2)[5,12–15,18,20–22,24,26–30,32–45,48–50,52–63,65–67,69,71,72]. Microtia, reported in 52–100% of patients, was described in 31 records[5,12,14,15,18,20–22,27–30,32–34,38,39,41,42,45,50,52,53,55,56,60,65–67,69,73], and anotia, reported in 2.8–6.7% of patients, was described in five records[18,28,33,39,66]; five records did not distinguish between microtia and anotia[24,37,40,59,71], reporting a prevalence of 52–100%. Two records did not distinguish between the presence of microtia, anotia, and/or pre-auricular tags, which were reported in 98–100% of patients[35,36].

Pre-auricular tags (14.4–90%) were described in 16 records[15,21,24,26–28,33,39,40,44,49,50,52,55,59,66] and pre-auricular pits (2.3–20%) in seven records[15,21,26,28,39,59,66]. In four records, pre-auricular tags and pits were reported as one entity, being present in 42–70.6%[14,20,42,53].

Fifteen records reported atresia of the external auditory canal in 12.2–86% of patients[13–15,18,24,28,30,40,48–50,53,54,58,66] and six records reported stenosis of the external auditory canal in 10–36%[14,24,48–50,54]. Nine records did not distinguish between external auditory canal atresia and stenosis, reporting this anomaly in 25.1–98.6% of

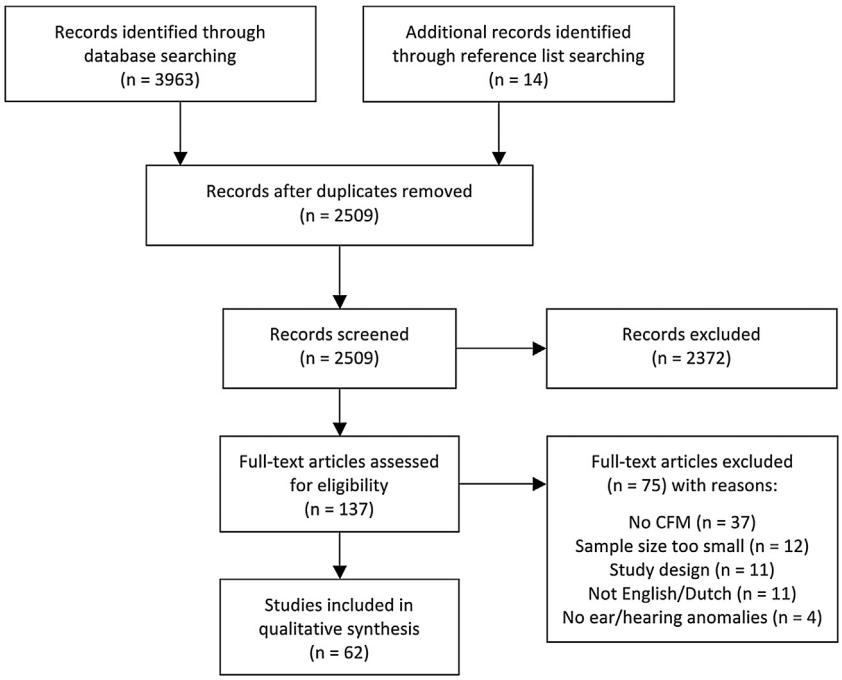

Fig. 1. PRISMA diagram of the record selection process.

patients[12,14,22,32,39,43,44,52,65]. One record did not distinguish between the presence of pre-auricular pits, external auditory canal atresia/stenosis, lobule hypoplasia, and dysmorphic ears, which were reported in 23% of patients[35].

'Ear' scores from the Orbit, Mandible, Ear, Nerve, and Soft tissue (OMENS) classification were described in nine records[24,26,48,54,57,58,61–63]. E0 (i.e. a normal external ear) was reported in 0–34%, E1 (i.e. mild deformity, all parts present) in 8–34%, E2 (i.e. moderate deformity; auricle significantly smaller, not all parts of the external ear present) in 13–41%, and E3 (i.e. severely malformed ear) in 20–58%. Only Rooijers et al.[63] used a version of the OMENS classification that includes the score E4 (i.e. anotia; reported in 2.5% of patients). All other records used OMENS classification 'Ear' scores limited to E3. These records did not describe the prevalence of anotia separately.

Middle ear malformations

Middle ear malformations, including anomalies of the ossicles and tympanic membrane, described in 10 records, were reported in 30–93% of patients (Table 1)[13,14,22,43,48,51,54,64,65,73]. Eight records further specified the types of middle ear anomaly[13,14,22,43,48,54,64,65]. Ossicle anomalies (e.g. fused, dysplastic, or absent ossicles) were reported in 42–93% of patients in eight records[13,14,22,43,48,54,64,65]. Tympanic anomalies, consisting of narrowed or opacified tympanic membranes, were reported by Bisdas et al.[13] in 79% of patients.

Inner ear malformations

Inner ear malformations were reported in 7–36% of patients in nine records (Table 2)[13,14,43,48,51,54,64,65,73]. The anomalies were specified in five records[13,14,43,48,64], consisting of hypoplasia of the oval window, vestibular anomalies, cochlear anomalies, and anomalies of the semicircular canals. In addition, Bisdas et al.[13] described additional inner ear malformations including common cavity in one patient and an enlarged internal auditory canal in two patients.

Hearing loss

Hearing loss was reported in 29–100% of patients in 40 records (Table 3)[12,14–18,20,21,23–30,32,33,35,40,42–46,48,49,51–55,59,60,66,68–70,72,73]. The type of hearing loss was specified in 21 records[12,15,20,21,23,26–28,30,32,33,43–46,48,51,52,54,55,73]: conductive hearing loss (CHL) was described in 11.1–97% of patients, sensorineural hearing loss (SNHL) in 1–40%, and mixed hearing loss (MHL) in 4.5–44.4%.

The degree of hearing loss was consequently reported in seven records[12,15,45,48,51,52,54]. Of these, five described the definitions for the degrees of hearing loss that were used[12,15,45,51,52] and four described the type of audiometry that was performed for audiological assessment[12,15,45,51].

Goetze et al.[15] reported the audiological assessments of 10 patients (20 ears) with a median age of 4.5 years (range 1 year and 9 months to 27 years and 4 months). In eight patients (16 ears), the degree of hearing loss was described based on the definition of Davis and Silverman[77], using pure-tone averages calculated from the results at 500, 1000, and 2000 Hertz (Hz), or using brainstem evoked response audiometry (BERA). Of these, three patients had normal hearing (0–20 decibels; dB); two patients had unilateral hearing loss, of which one had mild (21–40 dB) CHL and one had moderate (41–70 dB) CHL. Additionally, three patients had bilateral hearing loss, one with profound (>95 dB) SNHL in both ears, one with moderate CHL in both ears, and one with mild SNHL on the left and moderate SNHL on the right. The side affected by CFM was not reported.

Keogh et al.[45] reported the audiological assessments of 40 patients with a mean age 9.2 years (range 6 weeks–41 years). An age-appropriate audiological assessment was performed (i.e. pure-tone audiometry; behavioural methods, BERA), and pure-tone averages were calculated from the results at 500, 1000, and 2000 Hz. Two patients had normal hearing (0–20 dB), 17 patients had unilateral mild (21–40 dB) CHL, and 10 patients had unilateral moderate (41–60 dB) CHL. CHL occurred on the ipsilateral side of the microtia in all individuals. Ten patients had bilateral CHL and one patient had bilateral SNHL up to 60–80 dB hearing level.

Mitchell et al.[12] described the age-appropriate audiological assessments of 79 patients with a mean age of 9 years (range 1–23 years) at the time of the most recent audiological assessment. Masked pure-tone averages were used where possible, and pure-tone averages were calculated from the results at 500, 1000, 2000, and 4000 Hz. Sixty-five patients were reported to have hearing loss: 12 patients had bilateral and 53 unilateral hearing loss, resulting in a total of 77 affected ears. Hearing loss was mild (21–40 dB) in 10 ears, moderate (41–55 dB) in five, moderately severe (56–70 dB) in 44, severe (71–85 dB) in 15, and profound (>85 dB) in three. Hearing loss was reported in 70 of 96

Table 1. Middle ear malformations.

Author	Middle ear malformations	Ossicle anomaly (malleus/incus conglomerate)	Tympanic anomalies
Bisdas et al.[13]	93% (13/14)	93% (13/14) absent/dysplastic ossicles	79% (11/14) narrowed/ opacified tympanic membrane
Brotto et al.[64]		87% (26/30)[a]	
Caldarelli et al.[22]		48% (66/137)	
Hennersdorf et al.[43]		90% (19/21) dysplasia of the ossicular chain	
Jacobsson and Granström[65]		54% (28/52 ears)	
Rahbar et al.[48]	Unilateral HFM middle ear hypoplastic 70% (23/33) Unilateral HFM middle ear atretic 18% (6/33) Bilateral HFM middle ear hypoplastic 100% (7/7) Bilateral HFM middle ear atretic 0% (0/7)	Unilateral HFM 73% (24/33)[b] Bilateral HFM 86% (6/7)	
Rosa et al.[14]	67% (8/12)	42% (5/12)	
Sleifer et al.[51]	30% (6/20 ears)		
Strömland et al.[73]	67% (12/18)		
Wan et al.[54]	74% (64/87 ears) hypoplastic middle ear or atretic	82% (71/87 ears) fused or absent	

HFM, hemifacial microsomia.
[a] Not evaluable in five patients.
[b] Not identifiable in eight (24%) patients because of severe atresia of the middle ear.

Table 2. Inner ear malformations.

Author	Inner ear malformations	Hypoplastic oval window	Cochlear anomaly	Altered vestibule	Cochlear–vestibular malformations	Altered semicircular canals
Bisdas et al.[13]	36% (5/14)			29% (4/14)		29% (4/14)
Brotto et al.[64]	31% (11/35)	74% (23/31)[a]			31% (11/35)	20% (7/35)
Hennersdorf et al.[43]	33% (7/21)		29% (2/7)	57% (4/7)		71% (5/7)
Jacobsson and Granström[65]	4% (2/52 ears)					
Rahbar et al.[48]		Unilateral HFM 36% (12/33) Bilateral HFM 29% (2/7)	Unilateral HFM 3% (1/33) Bilateral HFM 14% (1/7)	Unilateral HFM 6% (2/33)		Unilateral HFM 3% (1/33) hypoplastic semicircular canals
Rosa et al.[14]	25% (3/12)		8% (1/12)			8% (1/12)
Sleifer et al.[51]	7% (2/30 ears)					
Strömland et al.[73]	17% (3/18)					
Wan et al.[54]	8% (7/87 ears)					

HFM, hemifacial microsomia.
[a] Oval window atresia. Not evaluable in four patients.

hypoplastic ears and in seven of 62 ears unaffected by CFM.

Sleifer et al.[51] assessed hearing in 10 patients (20 ears) with an average age of 10.1 years (range 3–27 years), using pure-tone and speech audiometry tests. Pure-tone averages were calculated from the results at 500, 1000, and 2000 Hz. Hearing loss was defined using the Davis and Silverman criteria[77]. Four ears had normal auditory thresholds, seven had mild hearing loss, seven moderate hearing loss, and two profound hearing loss. The relationship between the side affected by hearing loss and the side affected by CFM was not reported.

Treatment of hearing anomalies

The treatment for ear and hearing anomalies was described in eight records (Table 4)[17,18,32,43,46,54,66,68]. The use of hearing aids was reported in 21–51% of patients[17,18,46,54,66,68]. Mandelbaum et al.[46] reported the treatment for hearing impairment in 38 of 54 patients with reported hearing loss (70%), with 16 patients (42%) receiving non-surgical treatment using external hearing aids and 22 patients (58%) undergoing surgical treatment to improve their hearing loss. Surgical treatment consisted of canaloplasty in 16 patients, and bone-anchored hearing aids (BAHA) were placed in 12 patients. Skarzynski et al.[32] described cochlear implants as the treatment for hearing impairment in two patients.

Discussion

The aim of this systematic review was to describe the type and prevalence of ear

Table 3. Hearing loss.

Author	Hearing loss/impairment	Conductive hearing loss (CHL)	Sensorineural hearing loss (SNHL)	Mixed hearing loss (MHL)
Bassila and Goldberg[20]			16% (8/50)[a]	
Beleza-Meireles et al.[40]	59% (30/51)[b]			
Bergamini et al.[35]	76% (39/51)			
Bragagnolo et al.[21]		67% (48/72)[a]	42% (30/72)[a]	
Carvalho et al.[23]	75% (74/99)	85% (63/74)	1% (1/74)	14% (10/74)
Cohen et al.[24]	78% (69/89)			
Collett et al.[17]	70% (77/110)			
Collett et al.[16]	55%			
Converse et al.[25]	47% (7/15)			
Cousley[26]	66% (33/50)	91% (30/33)		9% (3/33)
D'Antonio et al.[55]	78% (32/41)	72% (23/32)	16% (5/32)	13% (4/32)
Digilio et al.[42]	59% (51/87)			
Engiz et al.[27]	71% (15/21)[c]	80% (12/15)	7% (1/15)	13% (2/15)
Ewart-Toland et al.[28]	43% (9/21)[d]	22% (2/9)	11% (1/9)	33% (3/9)
Goetze et al.[15]	63% (5/8)	60% (3/5)	40% (2/5)	
Hamilton et al.[29]	100% (11/11)			
Heike et al.[18]	64% (91/142)			
Hennersdorf et al.[43]	100% (7/7)	57% (4/7)		43% (3/7)
Jin et al.[44]	100 (103/103)[e]	47% (48/103)[e]		53% (55/103)[e]
Johns et al.[72]	82% (73/89)			
Keogh et al.[45]	95% (38/40)	97% (37/38)	3% (1/38)	
Llano-Rivas et al.[30]	92% (134/145)[e]	92% (123/134)[e]	4% (5/134)[e]	4% (6/134)[e]
Luquetti et al.[66]	91% (98/108)			
Mandelbaum et al.[46]	87% (54/62)	81% (44/54)		19% (10/54)
Mitchell et al.[12]	82% (65/79) affecting 49% (77/158) of all ears[f]	73% (56/77 ears)	1% (1/77 ears)	10% (8/77 ears)
Pegler et al.[49]	32% (13/41)			
Rahbar et al.[48]	98% (39/40)	87% (34/39)	5% (2/39)	8% (3/39)
Rooryck et al.[59]	68% (53/78)			
Rosa et al.[34]	29% (5/17)			
Skarzynski et al.[32]	82% (9/11)[g]	11% (1/9)	22% (2/9)	44% (4/9)
Sleifer et al.[51]	80% (16/20 ears)	63% (10/16 ears)	31% (5/16 ears)	6% (1/16 ears)
Speltz et al.[68]	95% (103/108)			
Speltz et al.[69]	70% (77/110)[h]			
Strömland et al.[73]	83% (15/18)	60% (9/15)	20% (3/15)	20% (3/15)
Suutarla et al.[52]		96% (73/76)[i]	9% (7/78)[i]	
Tasse et al.[60]	85% (29/34)			
Touliatou et al.[53]	76% (13/17)			
Wallace et al.[70]	70% (77/110)			
Wan et al.[54]	100% (87/87 ears)	82% (71/87 ears)	3% (3/87 ears)	15% (13/87 ears)
Wang et al.[33]	47% (14/30)[j]	21% (3/14)	7% (1/14)	21% (3/14)

[a] Among the total population.
[b] Not able to check auditory acuity in nine patients.
[c] Audiological evaluation available for 21 cases.
[d] Type of hearing loss unknown in three cases.
[e] Isolated microtia included.
[f] Type of hearing loss unknown in 12 ears.
[g] Type of hearing loss unknown in two cases.
[h] Audiological evaluation available for 110 cases.
[i] Among total available audiograms.
[j] Type of hearing loss unknown in five cases.

anomalies and hearing impairment in CFM. Ear anomalies were reported in 52–100% of patients, of which anomalies of the outer ear were most frequently described, followed by the middle and inner ear, respectively. Hearing impairment was reported in 29–100%, mostly specified as CHL.

The wide range of prevalence rates in ear anomalies and hearing impairment is primarily a consequence of differences in study objectives and methodology of the included records. First, inclusion criteria for CFM varied substantially between records. Several records only included patients with microtia in combination with additional symptoms (e.g. facial asymmetry), resulting in a 100% incidence of ear anomalies, whereas others used more elaborate criteria to include CFM patients, and some did not report the inclusion criteria at all. Internationally, there is no consensus on the minimal diagnostic criteria for CFM[78]. A recent European guideline on CFM recommended the use of the diagnostic criteria proposed by the International Consortium for Health Outcomes Measurement (ICHOM) for CFM[78,79]. The ICHOM criteria have been developed by a worldwide consortium of patients,

Table 4. Treatment of hearing impairment.

Author	Treatment described
Collett et al.[17]	23% (28/121) hearing aid use (current or past)[a]
Heike et al.[18]	21% (30/142) hearing aid use (current or past); 50% (69/137) had middle ear surgery (e.g. atresia repair or ear tubes)
Hennersdorf et al.[43]	57% (4/7) bone conduction hearing aids in patients with conductive hearing loss
Luquetti et al.[66,b]	51% (55/108) had used hearing aid; 55% (59/108) received speech, language, or hearing therapy; 6% (7/108) placement of ear tubes
Mandelbaum et al.[46]	24% (16/68) external hearing aid use; 24% (16/68) canaloplasty; 18% (12/68) received BAHA
Skarzynski et al.[32]	64% (7/11) tympanotomy; 18% (2/11) ossiculoplasty; 9% (1/11) epitympanotomy; 9% (1/11) myringoplasty; 18% (2/11) received cochlear implants; 9% (1/11) treated with a titanium fixture for BAHA
Speltz et al.[68,b]	51% (55/108) hearing aid use (current or past); 55% (59/108) received speech, hearing, or language services
Wan et al.[54]	27% (19/70) hearing aid use

BAHA, bone-anchored hearing aid.
[a] History of the use of hearing aid was self-reported.
[b] Records describe the same patient population.

healthcare professionals, and researchers active in the care and research of patients with CFM. In essence, the diagnosis of CFM requires the presence of $\geq$2 major criteria (i.e. mandibular hypoplasia, microtia, orbital/facial bone hypoplasia, asymmetric facial movement), or $\geq$1 major and $\geq$1 minor criteria (i.e. facial soft tissue deficiency, pre-auricular tags, macrostomia, clefting, epibulbar dermoids, hemivertebrae), or $\geq$3 minor criteria. We encourage the use of these diagnostic criteria; a clear definition of CFM is needed in research to guarantee comparable and reproducible outcomes.

Second, the wide range of prevalence rates for ear anomalies may be due to differences in methodology for reporting ear anomalies. In retrospective study designs, the prevalence of anomalies depends on the completeness and accuracy of the patient charts. Anomalies may not be recorded in the charts due to perceived irrelevance, thereby lowering prevalence rates. Additionally, prevalence rates may vary depending on the population in which they are calculated. For studies reporting middle and inner ear anomalies, prevalence rates may be higher in studies only including patients with computed tomography (CT) scans of the head due to selection bias, and lower in studies where relatively few patients with CT scans were included due to underdiagnosing.

Third, prevalence rates of hearing loss may vary due to the use of different diagnostic techniques, performed at different ages, and due to different definitions of hearing loss. Most records did not report the type of audiological assessment that was performed, at which age, and how hearing loss was defined. Some records reported the use of age-appropriate techniques to assess hearing, but without any further specification of their methods. Definitions of hearing loss were only sparsely given and varied between studies. Also, the circumstances in which audiometric tests were performed differed between studies, likely influencing outcomes. Settings with more background noise, for example in a home or school environment, may cause false-positives for hearing impairment. Thus, the reproducibility and generalizability of these studies is an issue.

Furthermore, many studies included (very) young patients, which hampers the interpretation of the prevalence of hearing loss. Several non-CFM-related conditions that are highly prevalent in young children in the general population, such as acute otitis media or externa, may cause (conductive) hearing loss[80,81]. However, further analyses of the cause and severity of hearing loss at different ages were generally not described in the included records. Nevertheless, the fact that most records that reported detailed hearing assessments identified patients with hearing loss on the side contralateral to that affected by CFM, suggests that conditions other than CFM may contribute to the reported hearing loss. The prevalence of hearing loss is thus likely distorted by other conditions, causing an overestimation of the actual prevalence of hearing loss caused by CFM in records including a relatively large proportion of young patients.

For targeted screening and therapy purposes, it would be useful to identify those patients most at risk of hearing impairment, preferably by non-invasive means. Several authors reported the relationship between CFM phenotypes and hearing loss. Phenotypes with hypoplastic ears were found to have significantly higher prevalence rates of CHL, SNHL, and MHL in the ipsilateral ear when compared to normal ears[12,23,44]. Greater degrees of ear hypoplasia were reported to be associated with significantly higher rates of hearing loss[12,44]. Furthermore, higher prevalence rates of hearing loss were described in patients with mandibular hypoplasia and microtia compared to those with microtia only[12,17].

However, several studies contradicted the relationship between the phenotype and hearing loss. For instance, the severity of CFM as determined by the total OMENS score was found not to be a predictor of hearing loss[48,54]. Moreover, Wan et al.[54] concluded that neither clinical nor temporal bone findings on CT imaging were associated with the type and degree of hearing loss. Furthermore, Carvalho et al.[23] found no difference in the prevalence of SNHL between bilateral CFM and unilateral CFM, or between CFM with facial nerve involvement and without facial nerve involvement. Interestingly, both CHL and MHL were also reported in a small number of patients without external ear hypoplasia, stenosis of the external auditory meatus, or ossicle anomalies, and SNHL was reported in patients without inner ear anomalies on CT imaging[43,44]. The cause for hearing loss in these patients is unknown.

There is debate among researchers regarding whether the total OMENS score, determined by adding up the individual scores for each facial feature, is a suitable measure for the severity of the CFM phenotype. The OMENS classification was constructed using ordinal values, for which the distance between the values cannot be assumed to be equal. Therefore, treating the individual scores as numerical variables by extrapolating the sum of individual scores to a total severity score is unlikely to be a valid measure of severity. There is a possibility that using different

measures for the severity of CFM in these analyses might yield different results.

It is generally expected that hearing loss in CFM will have a negative effect on speech, language, and cognitive development[12,17]. Therefore, the timely recognition and treatment of hearing loss is imperative. Logically, patients with microtia or stenosis of the external acoustic meatus should be examined for hearing loss using age-appropriate audiology, as is common practice in developed countries. Further evaluation using CT imaging might be indicated for the assessment of middle and inner ear anomalies, and for treatment planning[46]. Surgical treatment (e.g. placement of bone conduction devices) in these patients significantly improves hearing at speech threshold and psychological function[46]. The use of cochlear implants for the treatment of hearing loss primarily caused by CFM cannot generally be advised; only one record on this treatment modality for CFM was found, describing just two patients, providing very limited information to base treatment decisions on[32].

Since patients without distinct outer ear anomalies may also suffer from hearing loss, and in the absence of convincing correlations between hearing loss and other phenotypic characteristics, we cannot recommend a protocol for targeted screening. Therefore, we comply with the screening recommendations of the European guideline on CFM that all newborns with CFM undergo a neonatal hearing test[78]. When hearing anomalies are suspected, the infant should be referred to a centre specialized in craniofacial care for complete audiological evaluation, preferably before the age of 3 months. It is recommended that hearing is re-evaluated at the age of 24–30 months in all patients. Audiological interventions should preferably be initiated as early as possible, to prevent developmental delays. Future research should focus on clarifying the relationship between the CFM phenotype and hearing loss, in order to tailor screening and diagnostic protocols.

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Competing interests

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Ethical approval

Not applicable – literature research.

Patient consent

Not applicable.

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Appendix A. Supplementary data

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