REVIEW ARTICLE

Developmental Disorders of the Ear in Children and Adolescents

Conservative and Surgical Treatment Options

Thomas Braun, John Martin Hempel, Alexander Berghaus

SUMMARY

Background: Developmental disorders of the ear can impair hearing and cause cosmetic deformities. In recent years, new surgical treatments have become established, above all in audiological rehabilitation.

Methods: We selectively searched the PubMed database up to May 2013 for publications in English and German about the therapeutic options.

Results: No randomized trials have been performed, for both ethical and practical reasons (inadmissibility of placebo surgery, specialization of surgeons for individual techniques). To correct prominent ears, cartilage-sparing suture techniques are preferred, as they lead less often than scoring and incisional techniques to the formation of persistent, incompletely correctable ridges and scaffolding defects. The successful esthetic rehabilitation of severe deformities of the external ear is achievable through pinna reconstruction with costal cartilage (main risks: tissue defect at donor site, scaffolding resorption) or porous polyethylene (main risk: implant extrusion). The functional rehabilitation of conductive or mixed hearing impairment due to ear-canal atresia and major middle-ear deformities is preferably achieved with active middle-ear implants or bone-conduction hearing aids. Functional rehabilitation should be provided even when the hearing impairment is unilateral, in order to improve directional hearing and hearing with ambient noise. In cases of purely cochlear, unilateral, severe hearing impairment or deafness, a bone-conduction hearing aid can be tried, and the individual indication for a cochlear implant can be considered.

Conclusion: The treatment options described here enable the affected children to benefit from complete functional and esthetic rehabilitation before they start school.

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This review of developmental disorders of the ear which can cause hearing impairment and cosmetic problems concentrates on two new aspects. Since the introduction of universal newborn hearing screening programs, not only otorhinolaryngologists and pediatric audiologists, but also pediatricians, family physicians and other specialists are increasingly involved in the early diagnosis and therapy of hearing disorders. In addition, new audiologic technology has made possible different therapeutic approaches including active middle ear transplants for auditory canal atresias and cochlear implants for unilateral hearing impairment.

Methods

A selective PubMed search for therapeutic papers in this field was performed, analyzing both English and German publications for evidence level and recommendation grade according to the Scottish Intercollegiate Guidelines Network (SIGN) (*Table e1* and *e2*).

Developmental disorders of the external ear

Epidemiology

The most common developmental anomaly is prominent ears which affects 5% of the people (e1); the prevalence of severe anomalies is around 1:10 000–20 000 in newborns (e2). Malformations can be isolated or part of a syndrome (as in Goldenhar, Franceschetti, Nager, Apert, Moebius or Crouzon syndrome). They are often associated with a hemifacial microsomy; the incidence is around 1:5600 newborns (e3). The spectrum ranges from minimal changes such as prominent ears (the distance of the pinna from the scalp clearly exceeding the norm which is around 20 mm in adults) to severe facial asymmetry with microtia (rudimentary external ear) (e2, e4).

Embryology and pathogenesis

The complexity of malformations of the external ear is explained by incomplete development or abnormal fusion of the six mesenchymal auricular hillocks (microtia results from abnormalities of hillocks 2–5). Hemifacial microsomy also has bone and soft tissue defects with involvement of the mandible, temporomandibular joint, muscles of mastication and ear (e3, e5).

Classification of malformations and dysplasias

Accessory tragi are pre-auricular cutaneous protrusions which are considered choristomas (arising from misplaced embryological tissues which continue to develop). They should be separated from external ear malformations, although the two often occur together (e2).

The classification of Weerda is widely used for external ear malformations *(Table 1)*.

TABLE 1

Classification of external ear malformations after Weerda (modified from [e2])

Dysplasia grade	Examples
Grade I (minimal)	– prominent ear – macrotia – buried ear (cryptotia) – scaphoid – Stahl ear – satyr ear – question mark (Cosman ear) – ear lobe malformations – minor malformations of the tragus and antitragus, absent crus helicis, Darwin tubercle – cup ear malformations types I, IIa and IIb after Tanzer
Grade II (intermediate)	– cup ear malformation type III after Tanzer – microtia ("concha type microtia" after Nagata)
Grade III (severe)	– microtia ("lobule type microtia" after Nagata) – anotia

Clinical significance

Malformations of the external ear by themselves do not lead to hearing impairment, as long as they are not associated with additional auricular developmental anomalies. This, however, is often the case; in a cohort of deaf children, significant external ear malformations were found in 3% (e6). The main clinical significance is the aesthetic problems, which even in the case of prominent ears can lead to a reduced quality of life, reduced self-esteem, social avoidance behavior, and poor performance in school (e7–e9)

Diagnostic approach

Evaluation by both an otorhinolaryngologist and a pediatric audiologist is usually required to evaluate malformations of the auditory canal, middle and inner ear as well as determine if hearing impairment is present. The external ear changes should be photographed. If a syndrome is clinically suspected, clinical genetic consultation should be obtained. A pediatrician should evaluate the child looking for other visible malformations and searching for signs of internal involvement.

Therapy

Conservative therapy—Matsuo suggested in 1984 using individually molded splints to re-form the still flexible infantile ear cartilage (1). There are numerous reports on using ear splints for relatively minor malformations such as prominent ears, cup ears and Stahl ears (evidence level 3 and 4) (review by van Wijk [2]). The results from three studies with control groups reach evidence level 2– (3–5). There is still uncertainty about the spontaneous course of such malformations as well as at what age to start using ear splints, how long to continue the therapy and how effective it is in the long term. The risks are so minimal that ear splints can be recommended for a trial in infants (recommendation grade D).

Prosthesis—The external ear can be replaced with prostheses which are attached with adhesive or osseointegrated, using titanium pegs anchored in bone. The advantages are highly satisfactory and rapid aesthetic correction as well as the avoidance of operative effects, such as donor site defects (for example, after removing rib cartilage).

Although the benefits of an ear prosthesis on the quality of life are well-proven in adults with microtia (review by Federspil [6]), there is only a single small (n = 8) pediatric series. Although there are no validated instruments to assess effects on quality of life, about 2/3rds of patients appear satisfied with their prosthesis (evidence level 3, recommendation grade D) (7).

Other authors feel that a prosthesis should not be employed in children and adolescents for psychological reasons (e2), because the patients continue to regard the prostheses as foreign objects (8). Another concern is that a child or adolescent's prosthesis may come loose in public. Finally, the prosthesis must be replaced every 2–3 years as the child grows.

Surgery—While there are many surgical approaches to malformations of the external ear, we will only consider correction of prominent ears and reconstruction of the pinna in patients with microtia.

Surgical correction of prominent ears *(Figure 1)*—Initially prominent ears were corrected by excision of post-auricular skin (e10, e11), until Gersuny recognized the elastic restorative capacity force of the ear cartilage (e12). One must determine exactly what change leads to the prominent ears in each patient. The most common causes are:

- defective development of the antihelix
- (pseudo-) hyperplasia of the concha
- protruding ear lobe.

Even though more than 100 methods of correcting prominent ears have been published, three basic approaches have become established:

- combined excision and suture techniques: after Becker (e13) and Converse (e14)
- cartilage abrasion or scoring: after Chongchet (e15) and Stenström (e16)
- suture-only techniques: after Goldstein (e17), Morestin (e18), Luckett (e19), Mustardé (e20) and Fritsch (e21).

Suture-only procedures are preferred to the more aggressive incisional or excisional approaches. Severe complications with unacceptable, often uncorrectable, alternations in ear positioning—such as unnatural edges and defects in the underlying support—are less

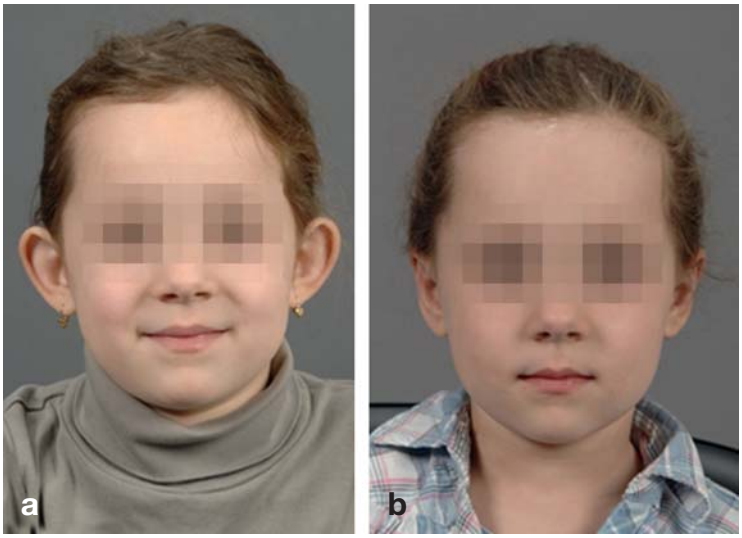

Figure 1: Prominent ears before (a) and after (b) corrective surgery. Through the use of a suture-only technique with appropriate modern suture materials, the cartilage framework of the external ear is maximally spared greatly reducing the risk of post-operative complications and deformities (surgeons T. Braun and J. M. Hempel)

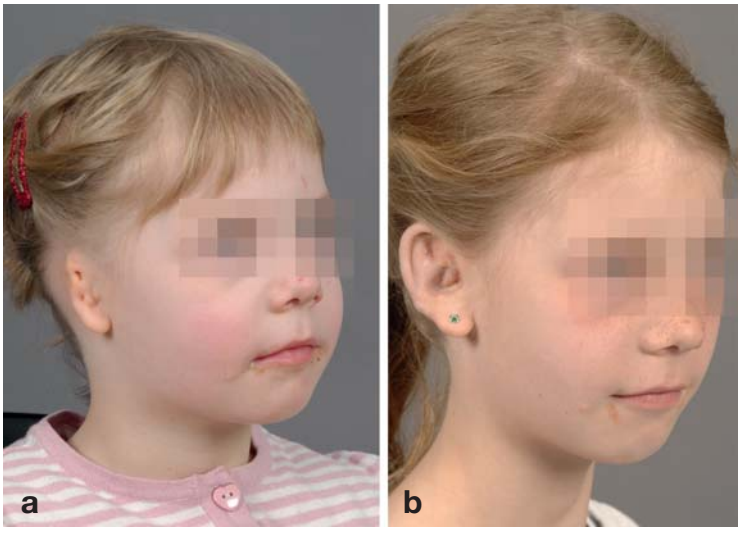

Figure 2: Reconstruction of microtia. Implantation of a porous polyethylene framework matching the normal ear, which is combined with a temperoparietal fascial flap and covered with local skin and full-thickness skin grafts. (a) Preoperative appearance; (b) after healing is complete (surgeon J. M. Hempel)

common in cartilage sparing suture techniques (evidence level 2+, recommendation grade C) (9, 10). Non-resorbable suture material must be employed to obtain satisfactory long-term results when suturing cartilage (11).

Surgical reconstruction of external ear *(Figure 2)*—The partial or complete reconstruction of moderate or severe malformations of the pinna is a surgical challenge because of the need to replace both the skin and the underlying cartilage framework.

For many decades, reconstructive measures were dominated by the use of skin flaps and autologous rib cartilage. This method was advanced by Converse, Tanzer, Brent, Nagata, Weerda and Firmin among others and requires multiple procedures, usually two or three (e22). Because of donor site defects after the removal of rib cartilage (scars, thoracic deformities [e23]) and uncertainty about long-term results because of resorption of the cartilage framework as reported by several authors (e24), a variety of alloplastic support materials were tried. All proved unsatisfactory—as for example silicone implants—because of increased infection and extrusion rates (e25). The introduction of porous polyethylene (PE) into ear reconstruction by Berghaus in the 1980s (12, 13) made available an alloplastic material that when combined with a suitable surgical approach (after Berghaus [13] and Reinisch [14]) produced durable long-term results. The biocompatible plastic allows the ingrowth of tissue components and has become the most widely used alloplastic material for reconstructing the external ear (12, e22).

Validated measures have shown an improvement in health-related quality of life following reconstruction both with rib cartilage and PE for children and adolescents (evidence level 2+, recommendation grade C) (15–17). Rib cartilage is generally first used after 8 years of age because before that there is generally an inadequate supply of donor cartilage (e2). Reconstruction with PE can be done in a single procedure after 4 years of age (17) and can thus help at an early age to reduce teasing which generally starts as affected children enter kindergarten or school. The teasing can lead to considerable psychosocial morbidity (e8). When such a correction is combined with simultaneous restoration of hearing using a bone-conduction hearing aid or an active middle ear implant, the child is both functionally and aesthetically corrected prior to exposure to peer pressure (17).

Developmental defects of the auditory canal and middle ear

Embryology

The external and middle ear both arise from the pharyngeal arches. Thus, combined malformations are commonly seen. For example, a grade 3 dysplasia of the external ear is almost always accompanied by auditory canal atresia. In contrast, inner ear defects are uncommon (e26).

Epidemiology and classification

Duplicated external auditory canals with fistula formation can be surgically repaired if recurrent infections are a problem. Middle ear defects often lead to conductive hearing impairment and are thus clinically significant. Minor middle ear malformations affect only the middle ear per se (usually defects in the auditory ossicles); major defects feature atresia or stenosis of the external auditory canal (congenital aural atresia) (*(Figure 2a)*. The incidence of congenital aural

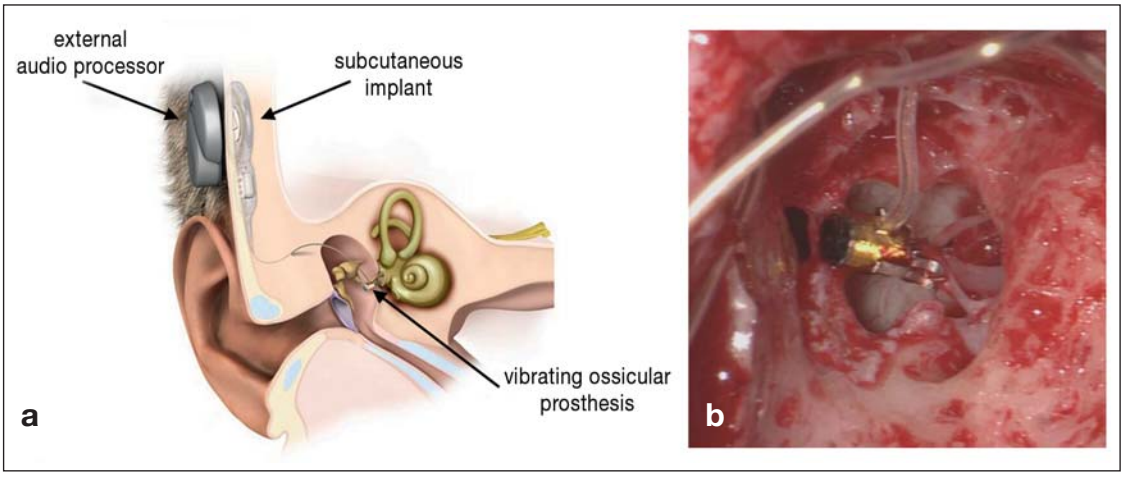

Figure 3: Functional correction of congenital aural atresia. Major middle ear malformations with low Jahrsdoerfer scores often do poorly with reconstructive surgery. Functional repair is better achieved with bone-conduction hearing aids or an active middle ear implant (a) (with permission of MED-EL, Innsbruck, Austria). The classical attachment of the vibrating ossicular prosthesis to the incus is usually impossible in congenital aural atresia; here it is attached to the stapes (b) (surgeon J. M. Hempel)

atresia either isolated or in combination with malformations of the external, middle and (rarely) inner ear is estimated as 1:10 000 (e2).

The Jahrsdoerfer CT score is used to classify the severity of such malformations and provides prognostic information regarding the utility of a surgical intervention (e27).

Clinical significance

Unilateral and bilateral middle ear malformations are separated because of their considerably different functional effects. Infants with the uncommon bilateral congenital aural atresia have bilateral absolute hearing thresholds of around 60dB. Prompt provision of hearing aids, usually bone-conduction type, in infancy is required to insure normal speech development (e28). Children with unilateral congenital aural atresia and normal hearing on the other side traditionally were not fitted with hearing aids because normal speech development was anticipated. Towards the end of the last century, it became accepted that unilateral hearing impairment led to marked reduction in the ability to localize sounds and to hear amidst background noise (e29). Therefore such children should also be evaluated for hearing aids. Children with unilateral hearing impairment who do not receive hearing aids have to repeat a school year more often (22–35%) and have more behavioral problems than children of the same age with normal hearing (e30).

Minor middle ear defects may lead to varying degrees of hearing impairment.

Diagnostic approach

External auditory canal stenosis or atresia can be recognized on otoscopic examination. Imaging to calculate the Jahrsdoerfer score (high resolution CT of the petrous portion of the temporal bone) involves considerable radiation exposure, so it is only used when the information has an immediate clinical impact, such as when auditory canal or middle ear surgery is imminent. Minor middle ear malformations (such as fixation of the malleus) often cannot be recognized on CT.

Middle ear malformations are usually found on mandatory newborn hearing screening (measurement of transient evoked otoacoustic emissions or conduction of early auditory evoked potentials). Further diagnosis consists of examination of early auditory evoked potentials in air and bone by an otorhinolaryngologist or pediatric audiologist. In older children, diagnostic evaluation of hearing impairment includes hearing thresholds and speech audiometry (see AWMF S2 Guideline „Peripheral hearing impairment in children“ [18]).

Therapy

Minor middle ear defects can frequently be corrected by tympanoplasty (restoring functionally mobile ossicles). Larger defects with higher Jahrsdoerfer scores are usually treated with classical surgical approaches (creation of an external auditory canal and reconstruction of the tympanic membrane and middle ear) to restore hearing capacity (evidence level 2+, recommendation grade C) (19, e27).

The benefits of classic surgical management of congenital aural atresia with low Jahrsdoerfer scores must be regarded as uncertain (e19, 27). Re-stenosis of the external auditory canal is common; about 50% of the patients still require a hearing aid after the procedure (20). Bone-conduction hearing aids generally provide satisfactory amplification (21) and are preferred over surgical correction of atresia (evidence level 2+,

TABLE 2

Classification of inner ear malformations (after [e33])

Severity	Malformation
I	Michel deformity: complete absence of all cochlear and vestibular structures
II	Cochlear aplasia; sometimes vestibule
III	Common cavity deformity; rudimentary labyrinth without separation of cochlea and vestibule
IV	Cystic incomplete separation of cochlea and vestibule
V	Cochlear hypoplasia, sometimes rudimentary vestibule
VI	Cochlea with reduced number of turns (Mondini malformation)

recommendation grade C) (22). They are initially fixed with a headband or clip but after 3 years of age they can be anchored to bone with a titanium screw. These devices have been shown to improve the quality of life (23, 24). Percutaneous fixation is better accepted by patients than using head bands, clips or modified glass frames (25). Another option is a subcutaneous double magnet system for fixing the hearing aid (26).

Another new option is active middle ear implants, hearing aids which are partially implanted *(Figure 3a)*; they stimulate only the affected ear and achieve better amplification of higher tones than do bone-conduction devices. This leads to better understanding of speech in noisy surroundings (27). A system from Ball for implantation in children has been approved since 2009 (28): an electro-acoustic transformer is coupled with the ossicles and amplifies their oscillation. It can be attached to the incus, stapes *(Figure 3b)* or even directly to the oval or round window, depending on the malformation (28–31). Through this approach functional restoration of conduction or combined hearing impairment can be achieved (evidence level 2+, recommendation grade C) (17, 30, 32). Improvement in the quality of life of children and adolescents with congenital aural atresia following implantation of an active middle ear implant has been shown with validated questionnaires (17). Because of these advantages, today the treatment of congenital aural atresia with middle ear implants is preferred over classical corrective surgery (17, 30, 32).

Developmental abnormalities of the inner ear

Epidemiology

Malformations of the inner ear can be demonstrated with modern imaging techniques in 40% of children with sensorineural hearing impairment (e31).

Embryology

The inner ear develops out of an ectodermal invagination. This then differentiates into the pars utriculoampullaris (utricle and vestibular canals) and pars sacculocochlearis (sacculus and cochlea). The inner ear and vestibulocochlear nerve (cranial nerve VIII) develop in intimate contact (e26).

Classification

There are both malformations of the membranous and bony labyrinths. The classifications of Jackler (e32) as well as Sennaroglu and Saatci (e33) are widely used. The latter grades the severity of the most common bony defects that can be identified with imaging according to the likely time of developmental arrest (*Table 2)*. In addition there can be developmental defects of the endo- and perilymphatic systems (vestibular and cochlear aqueducts) and the internal auditory canal.

Clinical significance

Inner ear malformations are important because of the resulting sensorineural hearing impairment. Depending on whether the defect is uni- or bilateral and on its severity, untreated children may develop speech disorders as well as have problems with sound localization and hearing in noisy environments. Malformations of the endo- and perilymphatic systems can present with progressive hearing impairment or recurrent meningitis. Although not mentioned in *Table 2*, the large vestibular aqueduct syndrome deserves special mention; progressive hearing impairment develops and may become much worse following even minor skull trauma (e26), apparently because variations in the cerebrospinal fluid pressure are transmitted in non-modulated form to the endolymph.

Diagnostic approach

Malformations of the bony labyrinth can be identified with imaging but those of the membranous labyrinth are usually only recognized histologically (e26). Because of a lack of therapeutic implications, imaging is rarely necessary for making the initial diagnosis of sensorineural hearing impairment. However, imaging must be done before cochlear implantation (e26).

When congenital sensorineural hearing impairment is diagnosed, associated syndromes should be excluded by a pediatrician or other specialists. Examples include:

- Pendred syndrome with disorders of thyroid function
- Alport syndrome with renal and ocular involvement
- Usher syndrome with ocular involvement
- Jervell–Lange–Nielsen syndrome with cardiac involvement

(see AWMF S2 Guideline „Peripheral hearing impairment in children“ [18]).

Therapy

Bilateral sensorineural hearing impairment can usually be managed with audiologic devices, usually hearing aids or if these prove inadequate for appropriate speech development, then cochlear implants (see AWMF S2 Guideline „Peripheral hearing impairment in children“ [18] and the consensus paper on hearing aid use from the German Society of Phoniatrics and Pediatric

Audiology [Deutsche Gesellschaft für Phoniatrie und Pädaudiologie] [33]). Malformations of the inner ear can make the use of a cochlea implant difficult and associated with poor audiologic results (for example with a common cavity) or even impossible (for example aplasia of the labyrinth, absent vestibulocochlear nerve) (34, 35). The audiologic improvement achieved with brain stem and mid-brain implants is clearly less than with cochlear implants (36).

In the case of unilateral hearing impairment, a trial of therapy with a bone-conduction hearing aid is recommended to improve spatial speech perception. This device makes possible pseudo-stereophonic hearing; surveys of affected children using validated instruments have shown improvement in quality of life measures (maximum evidence level 2+, recommendation grade C) (37, 38). Recently, cochlear implants have been employed in adults with unilateral hearing impairment; in contrast to bone-conduction hearing aids, these implants make true binaural hearing possible (e34, e35). Because there is so little experience in children and adolescents (39, 40), each case must be assessed individually (evidence level 2+, recommendation grade D).

KEY MESSAGES

- Prominent ears are best corrected with cartilage-sparing suture techniques in order to reduce the risk of post-operative framework defects.
- Successful aesthetic reconstruction of major malformations of the external ear is possible with rib cartilage (main risks: donor site defects, resorption of implant) or porous polyethylene (main risk: extrusion of implant) frameworks.
- Functional correction of conduction or combined hearing impairment in congenital aural atresia is best accomplished with bone-conduction hearing aids or active middle ear implants.
- Unilateral hearing impairment should also be corrected to improve ability to localize sounds and hear in noisy surroundings.
- One can try to treat unilateral deafness with a bone-conduction hearing aid or in individual cases with a cochlear implant.

Conflict of interest statement
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eTABLE 1

SIGN Grading System, Levels of Evidence (www.sign.ac.uk/guidelines/fulltext/50/index.html)

Level	Description
1++	High quality meta-analyses, systematic reviews of randomized controlled trials (RCTs), or RCTs with a very low risk of bias
1+	Well conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1–	Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++	High quality systematic reviews of case control or cohort studies High quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
2–	Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion

SIGN, Scottish Intercollegiate Guidelines Network

eTABLE 2

SIGN Grading System, Grades of Recommendation (www.sign.ac.uk/guidelines/fulltext/50/index.html)

Grade	Level of Evidence	Description
A	1++ or multiple 1+	Strong recommendation "definitely should"
B	multiple 2++ or extrapolated from 1++/1+	
C	multiple 2+ or extrapolated from 2++	Recommendation "should"
D	3 or 4 or extrapolated from 2+	Recommendation open "could"

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