UK Care Standards for the Management of Patients with Microtia and Atresia

BAA – British Academy of Audiology (baaudiology.org)

BAAP – British Association of Audiovestibular Physicians (baap.org.uk)

BAPA – British Association of Paediatricians in Audiology(bapa.uk.com)

BAPRAS - British Association of Plastic, Reconstructive and Aesthetic Surgeons (bapras.org.uk)

Changing Faces (changingfaces.org.uk)

ENT UK- Ear, Nose and Throat- United Kingdom (entuk.org)

Microtia UK (microtiauk.org)

Microtia Mingle (microtiamingle.co.uk)

NDCS – National Deaf Children's Society (ndcs.org.uk)

PPN UK – Paediatric Psychology Network UK (www.ppnuk.org)

Contents **Page**

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SECTION 1

Executive Summary

Patients born with microtia and aural atresia have a complex craniofacial condition that may impact on all aspects of their lives. It is essential that these patients and their families have access to specialised microtia teams able to provide up to date and unbiased information. A multidisciplinary approach should be taken to provide holistic, individualised assessments and interventions. This should encompass cosmetic, audiological and psychological aspects of their care.

Close liaison between local services and the ear reconstruction team is a key component to achieving the best outcomes for these patients. There is a recognised need for agreed care standards for these patients, and this document aims to reflect a consensual view of how this care can be provided in an integrated fashion. It has been produced by a collaboration of professionals working in the field and with stakeholder organisations.

MICROTIA and ATRESIA - THE CARE OF PATIENTS IN THE UK

KEYPOINTS

- Children with congenital microtia and atresia should be referred at the earliest opportunity to clinicians with appropriate professional expertise and knowledge of these conditions.

- Complex aspects of microtia and atresia care should be offered by specialised multidisciplinary teams

- The specialist team should work in close collaboration with local teams and professionals supporting families.

- Regular review within a multidisciplinary setting offers the patient and family holistic assessment and management.

- The multidisciplinary team should consider audiological, psychological and reconstructive aspects of care in an individualised manner.

- Information about support groups and organisations should be provided.

- Patients should be offered a point of contact for ease of access to the service. This may be a specialist nurse or other appropriately trained individual.

- It should be recognised that unilateral atresia and associated hearing loss may have an impact on a child's development, and that the child's progress and hearing should be closely monitored.

- For hearing restoration, options may include: educational support, conventional hearing aids, bone conduction hearing aids, bone anchored hearing aids and implantable hearing devices.

- Patients should be offered all appropriate reconstructive options for both the external ear and auditory restoration.

- For external ear reconstruction options include: no intervention, reconstruction with rib cartilage, reconstruction with a subcutaneous prosthesis, or an external moulded prosthesis.

- Patients and families should be supported in an unbiased manner in making informed decisions about which, if any, treatments are most appropriate for them.

- Patients and families should be able to access services at any age. For example, if a decision is made not to intervene as a child this should not preclude the same patient being offered intervention as an adult.

- Centres should be nationally designated and centrally funded.

- Specialist centres should undertake sufficient numbers of cases annually to be able to maintain and audit acceptable results. Surgeons should perform a minimum of 20 ear reconstructions per year with 10 of those being ‘total’ reconstructions for microtia.

- Specialist units should work on a hub and spoke basis in close collaboration with local teams to provide outpatient care in a patient-convenient location.

- Centres should be embedded within established reconstructive surgery units and should offer ear reconstructions for acquired as well as congenital conditions.

- All processes should be subject to local clinical governance standards and policies.

- Outcome measures should be routinely audited and reviewed using standardised agreed national measures.

- Regular UK national audit meetings should take place to review outcomes and to share best practice.

- Surgeons embarking on a career in microtia should be able to demonstrate a significant period of training devoted to acquisition of the necessary skills in a recognised centre. Certification of competence in all the techniques they offer should be evidenced in an appropriate manner.

- In the early period of practice, surgeons should enter a period of mentorship with a recognised expert. Centres offering such surgery should appoint consultants on a proleptic basis to facilitate this arrangement.

SECTION 2 – INTRODUCTION AND BACKGROUND

The UK health community has, since the inception of the NHS, been driven in its collective desire to improve standards of care in all arenas. Developments and improvements have often been related to improved understanding, new accumulating evidence and the development of new treatments and technologies. Arguably some of the greatest improvements in healthcare have resulted from evaluating services and understanding the current shape of care, and how this manifests for individual patients in their experiences and in their outcomes.

In the field of reconstructive surgery for congenital difference, the most striking example of such an evaluation came in the field of cleft lip and palate care. The UK had numerous small units providing variable levels of care with variable outcomes. Direct comparison with international units was unfavourable and there was general acceptance that standards of care and outcomes had to be improved.[1]

Patients born with microtia and atresia have by definition complex craniofacial deformations. The impact of this on all aspects of their life can be significant. Their need for information, support and, in many cases intervention, to restore form and function is considerable. Some of the interventions are highly complex and the outcomes are significantly dependant upon the quality of care that they are offered. Yet to our knowledge there has not, until now, been a co-ordinated national attempt to examine the shape and structure of care in the UK, and no means or methodology to assess the outcomes achieved nationally.

This document is the result of collaboration between interested parties to examine the current shape and structure of care. It is an attempt to define how that care should be best provided, and to suggest outcome measures which could be collated by all concerned. It has been commissioned by, and

sanctioned by, stake-holding associations and organisations with an interest in the field.

2.1 DEFINITIONS

The deformation in microtia can vary in its severity from an ear that is virtually absent to an ear that is perfectly formed but smaller than its fellow. The incidence is estimated at around 1 in 6,000 live births but varies between ethnic groups. In 90 percent of microtia cases only one side is involved, with twice as many on the right side compared to the left. Microtia affects boys in 65 percent of cases and girls in 35 percent. Microtia is frequently associated with atresia and can also be associated with more complex craniofacial conditions such as hemifacial microsomia and Treacher Collin's syndrome.

Aural atresia describes failure of development of the external auditory canal and is present in 80% of patients with microtia. Aural atresia arises embryologically from abnormal development of the 1st and 2nd branchial arches and branchial cleft, and ranges in severity from a patent auditory meatus with a "blind-ending" auditory canal, to complete absence of development of the meatus and canal with associated abnormal development, or absence of the middle ear structures. Aural atresia results in conductive hearing loss, with normal inner ear function (as the inner ear has a different embryological origin, it is normally developed in the majority of cases) as indicated by normal masked bone conduction thresholds in over 90% of cases.[2]

2.2 HISTORY OF EAR RECONSTRUCTION

The history of external ear reconstruction dates back to India and the Susruta Samhit and in Europe to 1597 when the Italian surgeon Tagliacozzi described and illustrated repair of the upper and lower ear using skin flaps. Various methods to achieve total ear reconstruction have been attempted including the use of maternal rib cartilage by Gillies in 1920 and the use of diced

cartilage buried in a metallic mould by Young and Peer in 1948. (http://www.microtia.us.com/)

However, the modern era of reconstructive ear surgery essentially began when Tanzer published his 6-stage technique using autologous costal cartilage in 1959. [3-8] His ideas were developed and refined significantly by Brent who was the first to demonstrate consistent, satisfactory and reproducible results in large patient numbers (published between 1973 and 2011). Brent relied on costal cartilage as the primary building block for his ear frameworks and reduced the number of stages to between 3 and 4. [9-16]

Through the 1980's and 1990's the work of Nagata[17-23] in Tokyo, and Firmin[24] in Paris, has significantly enhanced and improved techniques using costal cartilage to produce ever more convincing ears in two surgical stages.

The use of synthetic implants as the framework for ear reconstruction was first advocated by Cronin[25-27] in 1968. The idea was taken up but complication levels led to a general abandonment of that implant. More recently a porous polyethylene implant has been produced, and reports satisfactory outcomes when this is routinely covered by a temporoparietal fascial flap [28-32]. Concern exists in the UK and internationally regarding extrusion and fracture rates.

An ear prosthesis is an alternative option for restoration of form. Accounts of prostheses to replace the ear date back several centuries. Today, aural prostheses are made of medical grade silicone rubber and the shape and colour are customized for each patient. Ear prostheses can be retained with skin adhesives. While adhesives provided a means of retention, they have several problems: the application of the adhesive may be messy and time-consuming; the edges of the prosthesis must often be thickened or reinforced with fabric to resist tearing that may occur as the adhesive is cleaned from the prosthesis on a daily basis, which may detract from the appearance of the prosthesis; and the adhesive may cause skin irritation, particularly in those patients who have undergone radiation therapy.

Branemark (1985), a Swedish orthopaedic surgeon, defined "osseo-integration" as a direct structural and functional connection between ordered living bone and the surface of a load-carrying implant. This phenomenon was based on his bone healing research begun in the 1950s. In 1965, the principles of osseo-integration were applied to dental implants and this procedure has been widely recognized as safe and effective. Implant retention offers several advantages over skin adhesives, for example there is less wear and tear on the prosthesis and daily cleaning is faster and easier. Both of these factors contribute to the extended life of the prosthesis. Additionally, adhesive-related skin irritation is eliminated and implants provide more reliable retention of the prosthesis. Precise placement of the prosthesis is assured as the retentive elements automatically guide the prosthesis to its correct position. Implant-retained auricular prostheses have been used successfully in cases where the pinna is missing. [33-42]

Consideration of surgery for canal atresia was traditionally based on the Jahrsdoerfer scale [43] which scores the affected ear depending on presence or absence of structures in the middle ear and aeration of the mastoids. Patients scoring greater than 6 would be considered eligible for surgery. This challenging surgery, often combined with auricular reconstruction, involves drilling a new ear canal risking damage to the existing hearing and the facial nerve and so should only be performed by experienced Otologists.

Short term hearing results are good in patients with a score greater than 7 with 85-90% chance of achieving normal or near-normal hearing (as defined by an SRT ≤ 30 dB HL) but patients with lower Jahrsdoerfer scores had only a 45-50% chance of achieving this result [44]. Long term results are poorer, with reported a long-term (≥ 6 months) air-bone gap (ABG) of 30 dB or less in 51% of primary cases and 39% of revisions[45]. As a result most UK otologists are moving towards hearing implants.

SECTION 3 - IMPACT UPON PATIENTS AND FAMILIES

3.1 EVIDENCE BASE FOR IMPACT OF MICROTIA AND ATRESIA ON DAY TO DAY HEARING

1. UNILATERAL ATRESIA

1.1 Impact on listening

Lab-based tests of listening in noise with children with unilateral hearing loss and normally-hearing children suggest that children with unilateral hearing loss perform poorly when speech and conflicting noise are presented to each ear [46], which could therefore translate to listening difficulties in noisy places such as the classroom. Evidence suggests that right-ear impaired children perform worse than left-ear impaired children [46, 47, 48].

1.2 Academic performance

Studies of children with unilateral hearing loss (including children with any degree of hearing loss and many different aetiologies of hearing loss) show varying results. Some suggest that without treatment, children with unilateral hearing loss may need to repeat a grade at school [47, 48] and are rated as performing lower than their normally hearing peers on general educational performance dimensions by their teachers [49]. However, some studies find that there are no differences in academic performance between children with unilateral hearing loss or normal hearing [50, 51].

These studies are of varying quality, and most have small sample groups with children with mixed aetiologies of hearing loss, therefore making it difficult to generalise the results to all children with unilateral hearing loss.

1.3 Language skills

One longitudinal study of children with unilateral hearing loss suggests that over time, children with unilateral hearing loss seem to catch up with their normally hearing peers on measures of their oral-language [52]. However, it has also been shown that initially, on average, toddlers with unilateral hearing loss develop two-word phrases later than toddlers with no hearing loss [53].

1.4 Intervention options

For children with unilateral hearing loss, there is no evidence to suggest specifically when is the best time (if at all) to intervene. It is recognised however, that it is likely that there is a critical time window for maximum development of true binaural processes. Therefore there is a need to discuss with families whether to intervene as soon as possible to maximise acceptance of hearing aid and stimulation of a working cochlea, or whether to wait until potential problems arise such as delay in speech development, or poor academic performance, and then provide amplification as an option. Experience suggests that parents typically accept an offer of intervention in unilateral hearing loss when they perceive difficulties in their child's development and not based on theoretical considerations and reasoning.

1.5 Intervention outcomes

Studies looking at interventions typically report audiometric results rather than functional outcomes. Few studies assess the outcome of treating unilateral hearing loss with a bone conduction hearing device (BCHD). One study does show a significant effect of using a BCHD in children with unilateral hearing loss[54] tested with speech in spatially separated noise, but showed no difference on localisation tests. It has been shown that patients with an acquired unilateral conductive hearing loss may see an improvement in listening in noise with a BCHD. However, the same cannot be said of children with a congenital loss[55].

2. BILATERAL ATRESIA

Children with bilateral atresia will need some form of auditory stimulation via bone-conduction to hear speech clearly at a normal level. With this group of children, the decision is whether one bone conduction hearing device is adequate or whether a bilateral fitting is more appropriate. As we are aware, there is little to no transcranial attenuation of bone-conducted sound, although this has been shown to vary between patients and is also frequency dependent, with high frequencies demonstrating slightly more attenuation than low frequencies[56]. Therefore, we could expect that with one bone

conduction hearing device, and given that we have determined that both cochleae have hearing function within normal limits, both cochleae would receive some stimulation from this device, although this may not be equal. However, the positioning of the device which conventionally is on the mastoid, means that sounds arriving from the environment on the contralateral side will be blocked by the head, either introducing a slight delay in processing, or a reduction in level caused by the head shadow, suggesting it may be beneficial to have a device on both sides so that the wearer has bilateral input.

As for unilateral hearing loss, evidence for bilateral bone conduction hearing implants is very limited. A study investigating the effect of adding a contralateral bone conduction hearing implant to patients who had had a unilateral bone conduction hearing implant for many years found that 3/4 patients showed significant improvement on a localisation test with bilateral bone conduction hearing implants but speech recognition in noise was unchanged[57]. This study had only 4 participants. A second study again followed similar principles of providing a second bone conduction hearing implant later in life, but demonstrated a significant improvement in scores of speech recognition in both quiet and in noise[58]. Authors from a UK centre have shown marginal improvement in listening to speech in noise when fitted bilaterally over unilaterally[59].

All these studies have used patients who had a long period of usage of unilateral bone conduction hearing implant before being fitted with a second contralateral bone conduction hearing implant. This could have significant impact on trying to generalise the findings to infants who we may consider providing with two bone conduction hearing devices from initial diagnosis. We may expect that the participants in the studies could have suffered a degree of auditory deprivation of the unaided ear, although with previous mention of transcranial amplification this may not entirely be the case. Certainly they will not have had true bilateral amplification previously, therefore the brain's ability to realise binaural cues may be impaired or limited.

3.2 EVIDENCE FOR PSYCHOLOGICAL IMPACT OF MICROTIA AND ATRESIA

There are few studies exploring the psychological effects of microtia and/or atresia but there is some evidence of emotional distress as a result of microtia.

1. ATRESIA

Kesser et al (2013)[60] compared unilateral hearing loss and academic performance in school-aged children. No child repeated a year but 65% had extra resources in place and 73% reported problems with communication in groups or noisy environments. Ren et al (2012)[61] developed a quality of life inventory, the 'Congenital Aural Atresia Questionnaire', informed by the opinions of both patients and healthcare professionals, and consisting of 3 domains (physical, social, psychological). The questionnaire was piloted on 140 patients aged 6-18 years with unilateral and bilateral atresia and found to be sensitive to the severity of the condition and changes pre- to post-surgery.

A small scale study by the National Ear Reconstruction Service in Scotland (2013, unpublished) investigated parental desire for children with unilateral or bilateral hearing loss to access surgical hearing implants. Parents of 11 children (aged 2-12 years old) attending the microtia clinic were interviewed. Responses indicated that all parents felt their child's hearing could be improved and would consider surgical hearing implants for their children. 27% felt improved hearing to be of greatest importance and 73% felt improved hearing **and** ear reconstruction to be of equal importance, indicating the importance of the appearance of the ear as well as its function.

2. MICROTIA

The natural prominence of the ear allows disfigurements such as that seen in microtia to be visible, which can have an impact psychosocially on patients and their families. Anecdotal reports include avoidance behaviour such as reluctance to wear hair tied back or have short hair, with some parents

describing their children as isolated, refraining from taking part in exercise or playing with their peers often due to ‘playground teasing’.

The following lists of feelings and behaviours were compiled from the anecdotes of children, young people and adults with microtia attending clinics of the National Ear Reconstruction Service in Scotland (2013) demonstrating the psychological impact of microtia. Behaviours: always wear hair down, don’t let hair get wet, avoid hairdresser, avoid going out in windy weather, always wear hat/headband, avoid photographs or looking at photos, avoid mirrors, avoid questions about ear/appearance, avoid social situations, avoid physical activities, avoid special occasions, avoid school because of teasing, get into fights; and feelings: feel self conscious/ugly/angry/paranoid, lack confidence, suffer low mood and increased anxiety, have sleep difficulties. These are real issues that we hear repeatedly.

Two studies carried out in China investigated psychosocial difficulties in individuals with microtia. Du et al (2008)[62] found some evidence of depression, social difficulties and aggression in their sample group of 410 patients aged between 5 and 37 years old (mean age 12.2). Teasing from peers was found to be a risk factor for all three issues, as was the emotional impact on parents, highlighting the influence of the family on the individual. Li et al (2010)[63] compared the psychological profile of microtia patients aged 5-50 years old to a control group. They found significantly more social problems and aggressive behaviour in male microtia patients aged 8-10 years and 14-16 years, and a significantly higher prevalence of mood disorders in female microtia patients (aged 17 years and over). A significantly higher prevalence of interpersonal sensitivity, depression and anxiety was found among mothers of children with microtia. These results suggest that microtia is a condition that can have adverse psychological effects on patients and their families.

The following studies investigated the decision to undergo ear reconstructive surgery. Steffen et al (2010)[64] found the most frequently selected reason to decline surgery was stress associated with the frequency and length of hospital stays. Kristiansen et al (2013)[65] surveyed 78 patients (age range 9-

23 years) with unilateral congenital microtia who had undergone autologous ear reconstruction before the age of 12 and found the desire to have identical ears, perceiving their ear to look strange, frequent comments/questions from others, the wish to wear sunglasses, and getting teased to be motivational factors for surgery. Horlock et al (2005)[66] found teasing to be a motivational factor in children, and dissatisfaction with appearance the main reason for surgery in adults. Horlock et al (2005)[66] also found that following ear reconstruction 74% adults and 91% of children reported improvements in self-confidence, leading to enhanced social life and leisure activities. They concluded auricular reconstruction to be of significant psychosocial benefit to the majority of adults and children. Soukop et al (2012)[67] found that auricular reconstruction using autologous cartilage in children (aged 9-17 years old) resulted in significant improvements in health-related quality of life (physical health status, psychological state and social functioning) as measured by the Glasgow Benefit Inventory[68], with better surgical outcomes leading to greater improvements in these areas.

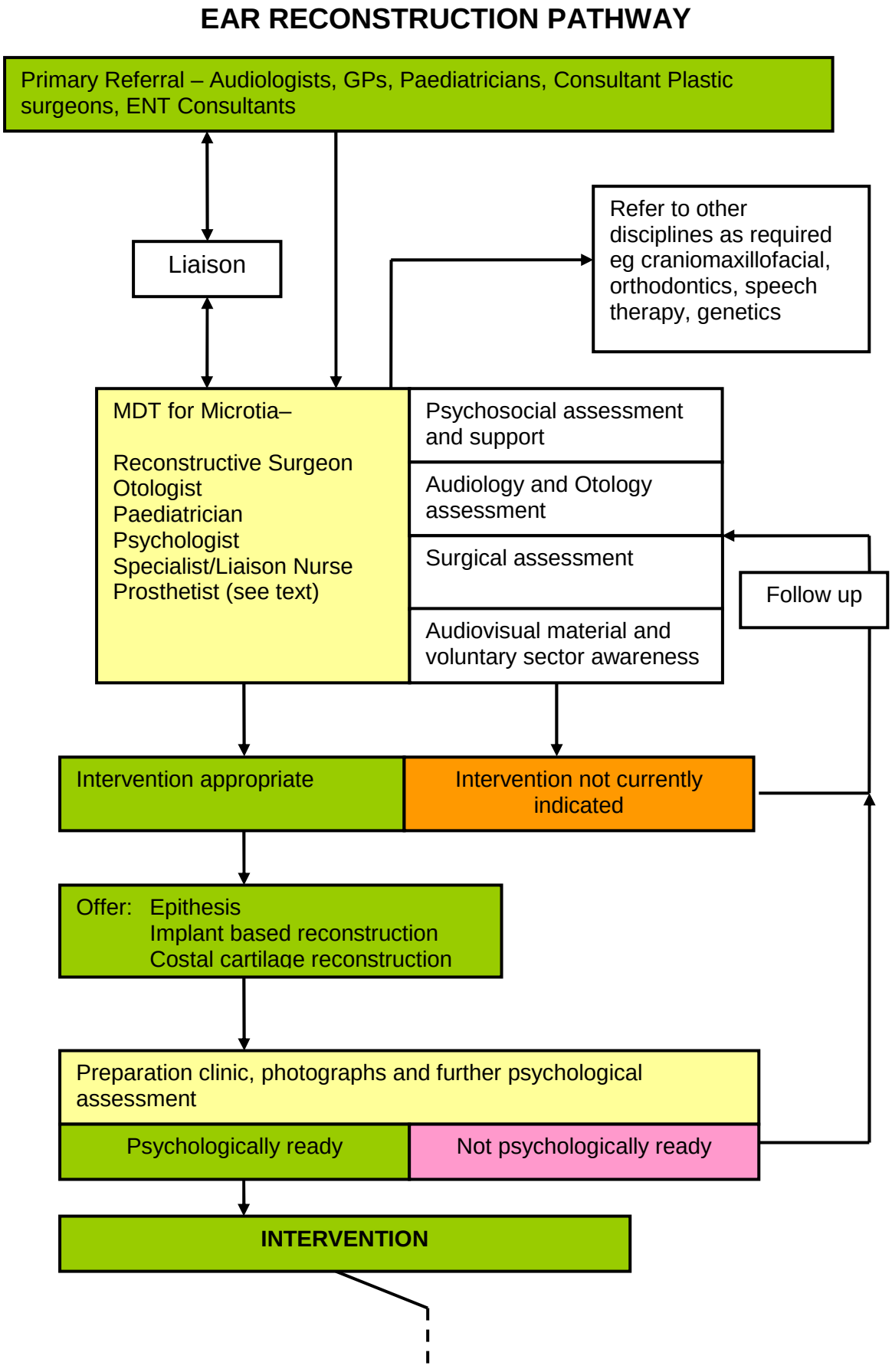
Low self-esteem, depression and anxiety are therefore problems that can occur in children and adults with microtia. From both the research carried out thus far and anecdotally, auricular reconstruction has been found to improve overall psychosocial outcomes.

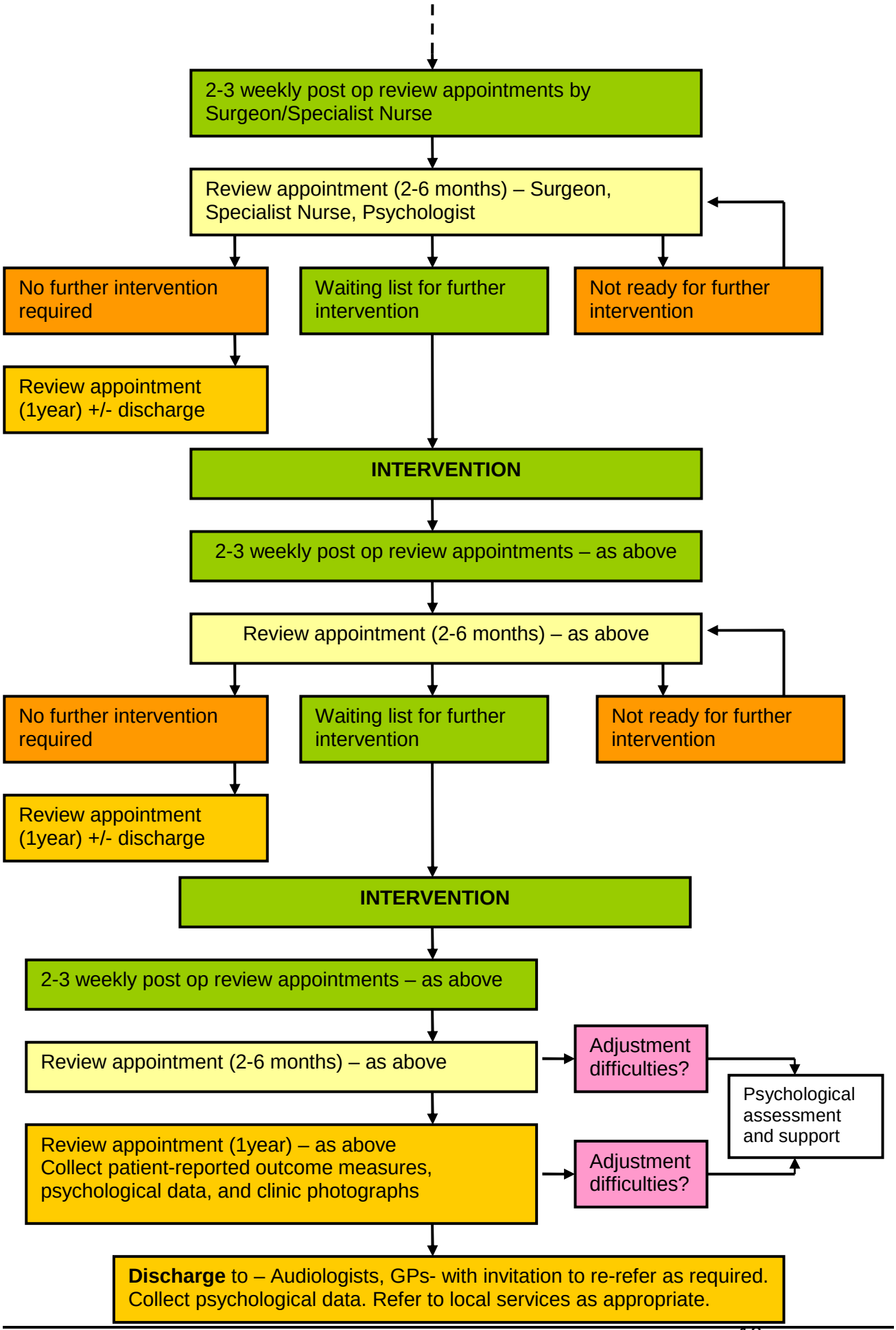
SECTION 4 - CARE PATHWAY

The age at which patients are referred for consideration of intervention for microtia varies. Below we describe a pathway applicable from birth. However, some patients may have avoided early referral or may have moved to the UK without prior intervention. All care must, self-evidently, be individual, and age and health appropriate. However, the core care standards are the same regardless of age. Figure 1 demonstrates an idealised pathway for a patient with congenital microtia, with or without atresia. Patients should be able to access services regardless of age although clearly age and co-morbidities may influence the care offered. If a collective decision is made not to intervene at one point in life this should not preclude future access to services. Patients should be able to access services at any age.

Clearly the assessment process needs to be integrated. The assessment of the patient with microtia therefore requires comprehensive evaluation of any associated hearing loss, and consideration of appropriate and timely intervention.

4.1 CARE PATHWAY FLOW CHART




2-3 weekly post op review appointments by Surgeon/Specialist Nurse
Review appointment (2-6 months) – Surgeon, Specialist Nurse, Psychologist
No further intervention required
Waiting list for further intervention
Not ready for further intervention
Review appointment (1year) +/- discharge
INTERVENTION
2-3 weekly post op review appointments – as above
Review appointment (2-6 months) – as above
No further intervention required
Waiting list for further intervention
Not ready for further intervention
Review appointment (1year) +/- discharge
INTERVENTION
2-3 weekly post op review appointments – as above
Review appointment (2-6 months) – as above
Adjustment difficulties?
Psychological assessment and support
Review appointment (1year) – as above
Collect patient-reported outcome measures, psychological data, and clinic photographs
Adjustment difficulties?
Discharge to – Audiologists, GPs- with invitation to re-refer as required.
Collect psychological data. Refer to local services as appropriate.

SECTION 5 - ASSESSMENT

5.1 THE MULTIDISCIPLINARY TEAM

Ask any parent of a child with microtia and/or atresia what could have been better about the initial weeks after the birth of their affected child and the answer will always be “information”: considered, knowledgeable and reliable information delivered by a professional who has a clear understanding of what issues their child might face, what services are available to help them and whom they should contact in the event of issues. Too frequently we meet parents who have in the past been given well-intentioned misinformation. This is not surprising given the relative rarity of these conditions. With an incidence of 1 in 6,000 patients, most midwives, health visitors and general practitioners will rarely meet such patients.

Parents should be given the opportunity for an early appointment with a healthcare professional with significant experience and understanding of microtia, atresia and conductive deafness. In some cases this may be provided in the first few weeks by a local health care professional. However, most families find it invaluable to have an early meeting with a multidisciplinary team as occurs with other conditions such as cleft. This team can provide expertise and up-to-date knowledge about the different aspects of these conditions and the related issues.

The aim of the multi-disciplinary team is to provide holistic child and family-centred care through regular contact with the professionals involved, so that children and their families are fully informed and supported, and are actively involved in any decision-making process. The exact constituent members of the multidisciplinary team who sit in the clinic may vary from centre to centre. The core members of the team should include a reconstructive surgeon, an otologist, an audiologist, a paediatrician with an interest in audiology, a clinical psychologist, a specialist nurse and an anaplastologist (maxillofacial prosthetist). Members of the wider team, in no particular order, include anaesthetists, educational audiologists and teachers of the deaf, craniofacial

or orthognathic surgeons, geneticists, nurses, orthodontists, radiologists and speech therapists.

The exact constitution of the team that meets each patient in the clinic may vary between centres but it is essential that the core presence includes a professional who can discuss hearing, assess audiological investigations, describe and indeed prescribe, interventions to aid hearing; and a professional who can describe and prescribe potential interventions for restoration of form including external prosthesis (epithesis), buried prosthesis and autologous reconstruction. The team should also be able to assess the patient's and family's emotional and psychological stressors, and if that person is not a clinical psychologist then there should be a streamlined process for onward referral to an appropriate Paediatric Psychology Service, or adult equivalent.

5.2 INITIAL ASSESSMENT

The team must ensure that appropriate initial investigations have been performed. Consideration should be given to renal ultrasound as it is recognised that there is an increased frequency of structural renal anomalies associated with external ear malformations[69]. A CT scan of the patient's head to assess the presence and anatomy of the ossicles is not generally indicated in the early years. Usually this would be undertaken when it might influence decisions regarding interventions. Baseline clinical photographs are helpful as are age-appropriate audiological assessments. Onward referral to other healthcare professionals such as geneticists should be streamlined and communication with referring doctors, general practitioners and local audiology teams is essential.

As per Newborn Hearing Screening Programme (NHSP) guidance, babies with microtia and atresia should be referred directly to the local Audiology department performing diagnostic assessment by ABR. The standard newborn hearing screen should not be performed. The goal of diagnostic assessment is both to establish the hearing threshold in the unaffected ear and to understand as much as possible about the hearing in the atretic ear.

The priority is testing of the cochlea for the ear with atresia (bone conduction testing). Knowledge about the cochlear function in this ear is essential to determine management and intervention options, and to inform discussions with the family.

The need for masked behavioural assessments in unilateral cases and for babies over 3 months of age means that the initial assessment by auditory brainstem response testing (ABR) is a crucial time to determine information about the true hearing levels of the affected ear, and in particular the cochlear function. For bilateral cases the goal is to demonstrate the level of cochlear function in both ears, without the requirement for masking, before 3 months of age. Clear guidance on the diagnostic testing that should be performed is outlined in the early assessment guidance published through NHSP and should be followed. Babies should be seen within 4 weeks of referral as per standards for newborn hearing screening.

Most cases of congenital ear malformations are obvious at birth. It should be noted that patients with stenotic ear canals could escape diagnosis and be mistakenly managed as having persistent middle ear effusions. Any narrow canal with non-visualised tympanic membrane and persistent tympanometry findings of immobile tympanic membranes should be treated with caution.
In smaller centres where there is less experience with permanent childhood hearing loss and atresia, advice on testing and management should be sought. Ideally this should be done ahead of the appointment so that the family can be fully informed and have all their questions answered. The ABR traces obtained and their interpretation should be reviewed by an external reviewer within an established local peer review programme or external expert.

The cause of the microtia and atresia should also be investigated - it is acknowledged that the physical manifestations of microtia and atresia mean that the initial identification will be with neonatologists and paediatricians.
It is important that the presence of any associated syndrome is investigated as early as possible. The options for investigations and professional contacts

arranged before hospital discharge will depend on the presence of other medical conditions, the birth hospital and the professionals involved. Guidelines for aetiological investigation of children with permanent hearing loss have been produced by the British Association of Audiovestibular Physicians (BAAP) and British Association of Paediatricians in Audiology (BAPA) (see Additional References[9]). Any investigations should be coordinated by the local lead for aetiological investigations for permanent childhood hearing impairment (PCHI). These may involve working with a different group of professionals than for sensorineural hearing loss, but it is important that the investigations are coordinated by one professional and used as part of the management plan for hearing and reconstruction interventions. The local audiology team performing the audiological assessment are responsible for notifying the local lead for aetiological investigations for PCHI. The lead for aetiological investigations is responsible for liaising with the other professionals involved and coordinating investigations, sharing of information and informing the management plan.

5.3 FOLLOW UP AFTER INITIAL ASSESSMENT

Following initial consultation and information exchange there is a need for regular follow up of children particularly when conductive hearing impairment is an issue. In most cases this can be offered as shared care between local audiology teams and the central multidisciplinary microtia team. Some families may prefer to have intermittent follow up annually or biannually; this fosters good relationships between professionals and the family and allows the team to discuss possible ear reconstruction in an age-appropriate manner. It also allows the family to ask about new developments in the field and if necessary to discuss (in liaison with local audiology services) psychosocial, educational and hearing issues which may arise.

Many parents and families find it useful to be provided with verbal explanations using pre- and post-operative clinical photographs of previous patients. In addition, written information to take home can be helpful and signposting to reliable online resources invaluable. It is during these early consultations that a family may benefit from time spent with a specialist/liaison

nurse. They will be able to help families make sense of the information they are being given and will be easily contactable for advice and support in between appointments. It is good practice to advise families of active voluntary sector support groups such as (in the UK) Microtia Mingle (www.microtiamingle.co.uk) which has a website and facebook page and Changing Faces (www.changingfaces.org.uk). Those with associated hearing impairment may be directed to the National Deaf Children's Society (www.ndcs.org.uk) and those with hemifacial microsomia to the Goldenhaar's Association. Families should be provided with information specifically about unilateral hearing loss and its impact, and information about support groups and information resources. It is important that information can be provided at the initial appointment even if not a specialist centre. These support groups and charities can help children and their families to deal with difference, and promote confidence, resilience and positive self-image.

As noted earlier, it should be recognised that a unilateral atresia and hearing loss may have an impact on a child's development and that the child's progress and hearing should be closely monitored. Each case should be managed on an individual basis. Factors additional to the microtia, and those yet to be identified, may increase the functional impact from a unilateral hearing loss. In cases of bilateral atresia with normal cochlear function, the impact on speech and language development is clear. This scenario should be managed as for any bilateral permanent childhood hearing impairment. The need for an intervention using a bone conduction hearing device (BCHD) and other options should be discussed and agreed with the family as soon as possible, with the introduction of local support services as per local pathways.

SECTION 6 – CONSIDERING EAR RECONSTRUCTION

6.1 THE DECISION MAKING PROCESS

The age at which patients, families and professionals consider intervention to restore form varies. A number of factors need to be taken into consideration. This includes the concerns and wishes of the child, the physical size of the child and their face, and the type of reconstruction being considered. The options for restoration of form include: doing nothing, external silicone prosthetics, buried synthetic frameworks and reconstruction with costal cartilage.

Children become aware of facial differences relatively early although this does not always mean they are psychologically distressed by their own facial differences. An old study by Dion (1973)[70] showed that pre-schoolers aged between 3.5 and 6 years old could distinguish differences in facial attractiveness, and Slater et al (2000)[71] found that 2-3 day old infants prefer to look at attractive rather than unattractive caucasian faces. Some parents and professionals have argued for early intervention as a prophylactic measure to prevent teasing and psychological distress. Certainly, early provision of external silicone prosthetics can be considered. Alternatively the use of buried prosthetic porous polyethylene frameworks can be provided as young as three.

The counterargument to early prophylactic intervention is that one is not treating the child but instead treating the family. Most young children are not concerned about their facial appearance, certainly infants and preschool age children do not typically engage in much social comparison[72], and experience from microtia clinics tells us that most children below the age of 8-10 when asked are simply not interested in any intervention. Social comparison begins in primary school and increases at least into adolescence, with comparisons to peers and the media, as well as direct comments from peers and parents, transforming beliefs and stereotypes about attractiveness into self-evaluation concerning appearance[72]. There is therefore a strong argument for waiting until the child is old enough, with the necessary level of abstract conceptual

thinking and emotional maturity, to enable informed decision making. Certainly deferring surgery beyond age 8 lends itself to reconstructions based on costal cartilage as the ribs are of insufficient volume until that age.

6.2 RECONSTRUCTION OPTIONS

The choice to undergo an intervention to restore the appearance of the external ear is an individual one. Some adults who have never had surgery are happy with their appearance. The MDT should enable patients and families to understand their choice either to have intervention or no intervention. Teams should offer a choice of the various forms of intervention available. The pros and cons of each form of intervention should be discussed in an objective manner with the aid of models and pre and post intervention photographs. The advantages and disadvantages of each type of intervention should be discussed to allow patients and their families to make an objective choice as to which form of reconstruction is best suited to their needs and desires. Once again, a specialist/liaison nurse can help the family make sense of the options available and take the time to ensure the child is suitably informed through simple discussion and picture presentations.

External ear prosthesis

External ear prosthesis, made from silicone, can be customised to achieve an excellent match in colour and shape. It is possible to attach the prosthesis with adhesives around the microtic ear at a very young age. Clinical opinion is divided on the benefits of such an approach. Some argue that early provision of a prosthesis may help the child and parents accept prosthetic camouflage as an integral part of body image and sense of self, at a stage of development where energy is focused on attainments, pursuit of interests and where social interactions are key. Some also feel that the advantage is of most benefit when the child starts school or nursery where they are suddenly mixing with lots of other children, as the child is less self-conscious of their prosthesis. Even if the child is not concerned with the appearance of their ear they might be subject to staring, teasing and comments by other children which could affect their confidence and self esteem. Early visits to the

hospital for non-invasive/non painful prosthetic treatment can often set the tone for future treatment options. Alternatively, instituting a regime of camouflage at an early age may indeed set the tone that the microtic ear needs to be camouflaged and indeed might be something associated with negative feelings.

A more solid attachment requires placement of titanium osseointegrated implants into the skull which is the preferred choice for long-term prostheses; the prosthesis is then attached onto the side of the head with either magnets or a bar and clip system. This type of attachment usually requires total ablation of natural ear tissue. If that choice is being made at an early age it should be made clear to the family that this process impairs others forms of ear reconstruction and can diminish the potential outcome by depriving the patient of a soft natural ear lobule formed from the microtic remnant.

Advantages of the external prosthesis are that the surgery involved is relatively simple and the ear can be made to look very realistic. There are several disadvantages with long-term external ear prostheses for microtia. The prosthesis needs to be repeatedly removed and replaced and the pin sites cleaned. Children may lose the device when engaged in sports or rigorous activities. It may be hard to hide the seam where the prosthesis is next to the normal skin, especially as skin colour changes throughout the year. A significant percentage of patients, who have osseointegrated screws, have pin site problems such as infection and over-granulation which require that the prosthesis is not worn until the problem has resolved. Finally, the prosthesis will need periodic replacement as it ages which adds to the long-term requirements of this procedure

High density porous polyethylene (Medpor™) is the most established buried prosthesis for ear reconstruction. This is a biocompatible material which is supplied in two individual pieces that are molded together by the surgeon to form an ear framework. The Medpor™ construct is covered by a flap of tissue taken from under the scalp (temporo-parietal fascial flap) which is then covered by a skin graft. The patient's own vascularised tissue completely covers and integrates into the implant.

Buried prosthesis

Medpor® reconstruction has been popularized by an American surgeon, John Reinisch who began using this technique in the 1990s. Medpor based reconstruction has distinct advantages. The surgery can be performed at a young age (3-5 years old), although the reconstructed ear has to be made bigger to account for the growth of the contra-lateral ear. Recovery is quicker than in autologous reconstruction as no rib cartilage is harvested.

The main disadvantage of Medpor® framework is the risk of prosthesis extrusion. Small extrusions may be managed conservatively but larger extrusions and infections will require that the construct be removed. Also since Medpor® ear reconstruction began in 1991, the outcome of the implant over a lifetime is not known. Extrusion rates as high as 13% and fracture rates as high as 15% have been reported.

Medpor® reconstruction is less technically demanding than rib cartilage reconstruction. However a high level of technical expertise is still required to attain good results and thus surgeons who perform this type of reconstruction should be doing so on a regular basis.

Reconstruction with autologous rib cartilage

Reconstruction with autologous rib cartilage has for many years been regarded as the gold standard in microtia reconstruction. Success with this technique was reported as far back as the 1970s when popularised by an American surgeon (Burt Brent). Brent has performed over two thousand cases demonstrating excellent aesthetic outcome, durability and longevity of ear construct. Two other surgeons who further advanced this technique and who have performed similar numbers of cases are Francoise Firmin in Paris and Satorou Nagata in Japan.

The original description of the technique by Brent was in four stages. This has been refined by Firmin and Nagata into two stages which most microtia surgeons now follow. The first stage is performed when the child is 9-10

years old. In this operation rib cartilage[7, 8, 9, 10] is harvested and a detailed three dimensional ear framework is constructed. A skin flap is raised at the site of the ear whilst the lobule is rotated into its normal position. The framework is placed in the pocket and gentle suction allows the definition of the ear to become visible. At the end of this stage the patient will have a normal looking ear which is adherent to the head. The second stage is performed after 6 months in which the ear is elevated from the head and a sulcus is created.

Experience demonstrates that the ears reconstructed with rib cartilage are durable for the rest of the patient's life. The cartilage is revascularised and responds to trauma by standard wound healing mechanism. The follow up by Brent (up to 17 years) showed no softening or shrinkage of the cartilage. There is also no doubt about the emotional and psychological benefit of the treatment. As well as the excellent results that can be achieved, the other main advantage is that this treatment has minimal long-term complications compared to prosthetic ears and thus over the years the cost incurred will be less than treatment with a prosthetic ear.

Autologous ear reconstruction is technically a very demanding operation with a steep learning curve. Surgeons who choose to perform this surgery must specialise in this field and should be dedicated to ear reconstruction. Poor results are very difficult to rectify and the best outcome for the patient is when reconstruction is performed in virgin tissue. Such surgery should be performed in designated national centres with a multidisciplinary approach.

6.3 PERIOPERATIVE CARE

Any inpatient care offered to individuals for either ear reconstruction or auditory device implantation should be conducted in an age-appropriate inpatient facility. All care should be subject to local clinical guidelines for infection control, pain control, patient safety and clinical governance.

The psychologist should be involved at preadmission stage to assess psychological readiness for surgery, including identifying any procedural

anxiety that will impact on the surgery itself, and psychosocial issues that may affect treatment adherence post-operatively. Post-operative follow-up should occur following completion of ear reconstruction to assess emotional state, satisfaction with surgical process, and body image issues. Pre-operative preparation of the patient can also include a ward visit, surgical planning, anaesthetic assessment, and the chance to ask questions or discuss anxieties with the specialist nurse.

Because of the high preponderance of difficult airways in patients with Treacher Collins Syndrome and patients with hemifacial microsomia, anaesthetic staff should be appropriately trained in difficult airway management and fibreoptic intubation. Post-operative care should be in a high dependency, or if necessary, an age-appropriate intensive care facility. Post-operative management of patients should involve nurses with appropriate training, surgical staff including the ear reconstruction surgeon and specialist nursing input as required. Implant based reconstructions may be discharged as early as 23 hours. Costal cartilage based reconstructions may require up to one week post-operative inpatient stay for surgical drains and pain management. The average inpatient stay following first stage costal cartilage reconstruction is 4 days. Subsequent stages of the autologous reconstructions can be performed on a 1 night or day case basis as geography allows.

Patients should be followed up regularly wherever possible by the operating surgical team in the early weeks following discharge. If geographical considerations make this difficult then follow up in a more local surgical care or dressing care facility may be appropriate, provided there is close communication between relevant professionals.

Patients should always be provided with a telephone point of contact to allow them to contact an appropriately trained member of the clinical team after discharge for advice and to facilitate early review as required. In some centres this may be a specialist nurse but other team members can provide this role.

6.4 TRAINING IN EAR RECONSTRUCTION

All surgical procedures improve with knowledge, experience and practise, and no surgeon is immune from this learning curve. Surgeons gain competencies at different rates and some are adept and perhaps 'suited' to different skill sets. Surgical training aims to provide surgeons with the surgical skills they need for independent practice and the learning skills they need for continuing acquisition of skill throughout a career. The Intercollegiate Surgical Curriculum Programme is an online platform which facilitates the documentation of surgical competencies during training.

Ear reconstruction is a curriculum option for senior trainees in reconstructive surgical specialties. Completion of this module should be regarded as mandatory for any surgeon wishing to embark upon a career in ear reconstruction. Any surgeon who has been trained in the era of ISCP and who wishes to undertake ear reconstruction must be able to demonstrate competence. Competence can be defined as procedure-based assessments signed at level 4 (competent to perform the procedure independently and to deal with any complication that may arise) by a recognised expert in the field.

Whilst ISCP-defined competencies are mandatory they do not ensure satisfactory aesthetic outcomes and ears with artistic merit. It is highly desirable that surgeons undertaking ear reconstruction have a specific period of training in their final years, or even beyond CCT recognition, devoted to ear reconstruction. They should be able to demonstrate prolonged commitment to the area of endeavour and their training should be endorsed as satisfactory by a surgeon with recognised expertise.

Ear reconstruction centres in the UK need to work together to produce a template that offers optimal training for the small number of surgeons required to support ear reconstruction surgeons for the UK. This may follow the interface specialty training programmes offered in other areas of reconstructive surgery.

Ear reconstruction centres should also consider, when appropriate, the proleptic appointment of surgeons to replace retiring surgeons with a significant period of overlap. Such a system will facilitate mentorship and would go a long way to reducing the risk of unsatisfactory results associated with the early period of a surgeon's ear reconstruction career.

Some have argued that the use of prosthetic materials such as Medpor® eliminate the learning curve for ear reconstruction. However, much of the learning curve and indeed risk of complications, relates to soft tissue and skin cover. Soft tissue cover for buried prosthetics is often more complicated as it involves the routine use of pedicled fascial flaps.

SECTION 7 - INTERVENTION FOR HEARING LOSS ASSOCIATED WITH ATRESIA

Families should be offered a BCHD (bone conduction hearing device) for all babies and children identified with a unilateral microtia[*1] with atresia. The rationale for this and the options available have been outlined.

From experience in a number of UK centres, a BCHD on a softband can be safely used typically from 3 months of age[*2]. It is expected that appropriate up to date information about the current options and rationales can be provided by audiologists performing diagnostic auditory brainstem response (ABR) assessments. Local centres with no hands on experience with these devices should offer referral to the nearest specialist centre providing BCHD for further discussion and information and potential trial.

Great care should be taken in the implantation of hearing aid devices since inappropriate access incisions or implant positioning may compromise ear reconstruction. Implantation should take place either in, or in very close liaison with, an ear reconstruction surgeon from an ear reconstruction centre. Surgery for an implanted device to aid hearing could occur earlier than ear reconstruction. Placement of the device has to take in to account the requirements of ear reconstruction. This is to ensure that any hearing device does not interfere with options for future ear reconstruction. This applies even if the family have not decided on ear reconstruction to enable consent to be given once the child is above the age of consent.

[*1] Experience in centres offering BCHD to families at this early stage is that families do not always take up the offer at this stage. The drivers for acceptance of this intervention include speech and language delay and presence of middle ear effusion in the unaffected ear.

[*2] The placement of the BCHD may need to be varied and include the forehead in addition to the mastoid bone for ease of use in a very young infant. However evidence is now emerging to suggest that the transfer of sound to the cochlea is not as efficient as previously thought.

7.1 AUDIOLOGY- BASED MANAGEMENT AND INTERVENTION OPTIONS

The following table summarises the roles and responsibilities at each level of service potentially involved in the management of children with unilateral atresia.

For those children with bilateral atresia the case for use of intervention is essential for the development of spoken speech and language skills. Referral to the specialist audiology centre should be carried out as soon as possible in discussion with the family. Many of the principles summarised in the table still apply to how services work with families, the child and other services to achieve successful outcomes.

	Audiology: Phases of management plan and interventions for unilateral microtia and atresia		
Service	**Birth/diagnosis**	**3 months onwards with or without BCHD**	**Surgical options –Implantable hearing device / reconstruction**
Local audiology and ENT team	Diagnostic assessment according to national standards. Priority to determine hearing in unaffected ear[*3] and bone conduction levels in atresia ear as minimum. Provide up to date information on the management of unilateral atresia. Provide information about parent groups. Provide information about keeping unaffected ear healthy. Discuss use of BCHD on softband for binaural hearing experience. Discuss role of education sensory support services and refer on agreement. Refer to local lead for aetiological investigations for PCHI to ensure coordinated assessments and sharing of information. Refer to specialist audiology team for further advice about BCHD if family interested, or local service not able to provide all the information required by the family. Refer to specialist ear reconstruction team for advice on future implantable hearing device and reconstruction options as requested and as meets needs of family. Named audiologist to act as link for family and other services. Issue with family-owned microtia plan.	Monitoring of hearing in unaffected ear. Monitoring of progress in speech and language development. Behavioural assessment commences around 7-8 months of age. In cases of persistent middle ear effusions discuss ▪ BTE aid for unaffected ear. ▪ BCHD on softband ▪ Liaise with otologist Monitor hearing in unaffected ear until 5 years of age with reviews every 3-4 months in first 2 years and every 6-9 months until 5 years of age. Refer to community paediatrician if developmental concerns. The child may already be involved because of previously identified co-morbidities, but it is possible that delays will become evident during audiological monitoring and prompt response is required to ensure early intervention. Revisit involvement of education sensory support services if not previously involved at nursery and school age. Support any requests for further information and advice with onward referral to specialist teams as appropriate.	

[*3] Unaffected ear refers to the ear without the atresia

	Audiology: Phases of management plan and interventions for unilateral microtia and atresia		
Service	**Birth/diagnosis**	**3 months onwards with or without BCHD**	**Surgical options –Implantable hearing device / reconstruction**
Specialist audiology team[*4]	Advice and information to the family about current and future BCHD options. Named audiologist to act as link for family and other services.	Advice and information. Opportunity to meet with other families. Provision of BCHD on softband. Agreed plan with family and local audiology to monitor progress. Possible that some monitoring could be done locally[*5]. Refer for information or consideration for implantable hearing device at parent request or if issues with use of band at any stage. Named audiologist to act as link for family and other services.	
Specialist surgical centre[*6]			Advice and information about implantable hearing devices and reconstruction options. Opportunity to meet other families/young people. Implantable hearing device surgery should ideally be performed by the reconstruction team[*7]. Named link professional to act as link for family and involved local and specialist services. Audiological management and monitoring of the device can be done between local, specialist audiology and specialist surgical ear teams as best fits the needs of the family. Surgery, fitting of device and ongoing maintenance.

[*4] Where an audiology service provides BCHDs the local and specialist otology/ audiology team will be the same.
[*5] This agreed plan will be dependent on location, individual families and knowledge and experience within the different audiology teams involved with the family. Not all follow-up support for the BCHD will need to be done by the specialist team.
[*6] Refers to specialist centre able to perform ear reconstruction and implantable BCHD surgery. This may or may not be the same service as the specialist audiology team.
[*7] Local arrangements for surgery to be carried out elsewhere can be agreed with full knowledge, advice and agreement with the specialist reconstruction team.

7.2 HEARING DEVICES

Research on early intervention for sensorineural hearing loss attests that early intervention with hearing aids is crucial to maximise both auditory and linguistic development in infants. For example, there is evidence that without access to speech sounds, children with hearing losses will not keep pace with their normally hearing peers in communication, cognition, social/emotional development and reading[73, 74]. The goal should be that unilateral or bilateral microtia / atresia does not have a detrimental impact on the development of the infant compared to normally hearing peers.

The review of devices prepared for this document concentrates on, and is influenced by, existing technologies available at the time of writing and those systems most commonly used in the UK. The reader should be aware that other systems, in particular implantable devices, exist and may become more appropriate to this patient group as the evidence and experience base develops, and that the devices are constantly evolving.

Bone conduction hearing device (BCHD) will be the likely device choice in most cases, although a low grade microtia with an ear canal stenosis rather than atresia, may mean that a behind-the-ear (BTE) hearing aid fitting is possible.

Bilateral atresia and access to speech

Cases of bilateral atresia should be managed as per any bilateral permanent hearing loss. The only option for intervention to ensure access to sound for spoken speech and language development is the use of a bone conduction hearing device. This should be explained and offered at diagnosis without delay.

It is recognised that one device is sufficient for acquisition of spoken language in cases of bilateral canal atresia. However, best practice is to offer bilateral devices to promote binaural hearing.

Unilateral atresia and binaural hearing

In the case of unilateral atresia, sound will not be able to reach a working cochlea. Stimulating a working cochlea as early as possible using a BCHD maximises the potential for future interventions. A goal is also to achieve as close to binaural hearing as possible.

Transcranial attenuation is greater in young infants than adults and this decreases throughout maturation. It is predicted that infants have at least 10-30 dB of transcranial attenuation to bone-conduction stimuli compared to adults[75]. Therefore, using a BCHD when there is a unilateral profound loss would be less effective for infants compared to adults, as the vibrations produced would be severely attenuated as they passed to the better hearing ear. However, when there is unilateral conductive hearing loss, and the purpose of amplification is to target the ipsilateral cochlea, infants and young children will have much less routing of the signal to the contralateral ear, and a much more binaural experience than adults.

It is recognised that families may not take up options of BCHD at the early stages following diagnosis of unilateral canal atresia. A trial of a BCHD for unilateral canal atresia should be available to families at any time and the advantages for future development and listening skills explained. This should be done in the context of neural plasticity and the potential for greater benefit from intervention, if that intervention occurs early.

Bone conduction aids

Traditional bone conduction systems typically consist of a microphone which is connected via a cable to a bone conductor/vibrator which is mounted onto a headband. The microphone may be part of a body worn sound processor, or a specially adapted linear BTE depending on the level of power needed (i.e. whether it is a mixed or a pure conductive hearing loss). Typically they use analogue sound processing.

Some limitations of transcutaneous conduction (sound travelling through skin) include attenuation of the signal as it passes through the skin. This can be up to 15dB for high frequencies[76]. To try to minimise this attenuation bone conduction transducers press firmly onto the skin. One commonly reported feature of bone conduction aids is that the tight headband can be uncomfortable to wear and/or cause headaches and sore skin/pressure points. The absence of digital sound processing and comfort issues means that they should not routinely be considered for infants or very young children.

Bone conduction hearing implants worn on softband

Bone conduction hearing implants were designed originally to attach to an implant in the temporal bone. However, it was quickly realised that bone conduction hearing implants can also be attached to a soft band that wraps around the head or mounted on a headband (often called a 'hardband'), with the sound transmitted through transcutaneous stimulation. The bone conduction hearing implant sound processor snaps onto the plastic disc on the soft or hard band, rather than onto the implant. This system is mainly used with children who are too young for surgery or who may grow out of their hearing problems, or for adults who wish to experience bone conducted sound before deciding whether to go forward with surgery for an implanted device. The bone conduction hearing implant uses digital processing of sound.

Research and specific recommendations for using bone conduction hearing implants on soft or hard bands are limited. For example, although it is widely believed that bone conduction hearing implants can be positioned on any convenient position on the child's skull (e.g. Cochlear, 2011) there is growing evidence that bone-conduction sensitivity is poorer when the transducer is placed on the forehead compared to the mastoid [77, 78].
Consideration also needs to be given to the attenuation of sound in all trancutaneous systems. As softbands are more regularly used in infants and young children than a traditional bone conduction hearing aid, when they are issued to a pre-lingual child, then the potential under amplification of high frequency sounds must be considered.

Implantable Hearing Aids and Canal Reconstruction

In the design of implantable hearing aids there have been 2 basic strategies used to stimulate the cochlea. The first strategy, as used in bone anchored hearing devices (e.g. BAHA, Pronto Pro), being to vibrate the cranium and thereby the cochlea which lies within the temporal bone of the skull. The second strategy is to produce mechanical vibrations that directly stimulate a middle ear structure causing it to vibrate (e.g. Middle Ear Implant, MEI).

Bone conduction hearing implants

Bone conduction hearing implants are an established treatment for conductive hearing loss, or single sided deafness, in children[79]. Traditionally, the external audio-processor has been clipped onto a percutaneous abutment attached to a titanium implant osseointegrated into the skull bone. This has proved to be an effective intervention in children, but is associated with a risk of implant loss due to trauma or failure of osseointegration, and recurrent skin inflammation[80,81,82]. Complication rates for bone conduction hearing implants in children can be high. Kraai et al reported soft tissue reactions in 89% bone conduction hearing implant cases in children, with implant removal or revision surgery required in 37% cases[81].

Placement of the percutaneous bone conduction hearing implant in relation to the microtic ear is of critical importance, as siting may compromise any subsequent autologous reconstruction[83]. Therefore, it is mandatory that the position for a percutaneous bone conduction hearing implant is determined by, or in close discussion with, an ear reconstruction service.

The introduction of surgical techniques and percutaneous implants that do not require soft tissue reduction have the advantage of preserving soft tissue planes, minimising the impact on future autologous ear reconstruction. However, it remains imperative to ensure that the position of the percutaneous bone conduction hearing implant does not adversely affect subsequent pinna reconstruction. To further negate the risk of skin infections and improve

cosmetic acceptance, passive transcutaneous bone conduction hearing aid systems have been developed and are now licensed for use in children.

Middle Ear Implants

Middle ear implants (MEI) (eg. Vibrant Soundbridge^{R} (VSB), MED-EL, Innsbruck) have been demonstrated to be an option for hearing rehabilitation in children with canal atresia, and can be used in conjunction with autologous ear reconstruction[84]. MEIs have an internal and external component as for transcutaneous bone anchored hearing devices, but the internal component produces mechanical vibrations that directly stimulate a middle ear structure, causing it to vibrate. In the Vibrant Soundbridge^{R} the mechanical vibrations are produced by an electromagnetic element called the floating mass transducer (FMT), which is attached to the stapes or incus, or placed against the round window membrane, dependent upon the anatomy of the middle ear[84,85]. Therefore, high resolution CT imaging of the temporal bone is mandatory when considering a MEI and consent must include the risk of loss of hearing, vertigo and facial nerve injury. MEIs should also be considered with caution when the middle ear is significantly dysplastic and poorly aerated. When siting the incision for MEI surgery prior to planned autologous ear reconstruction, the same consideration must be taken as for bone anchored hearing devices, to ensure the necessary preservation of the tissue layers around the microtic ear[86]. In response to the introduction and likely evolution of transcutaneous bone conduction hearing implant systems, and the fact that the majority of atresia cases have a purely conductive hearing loss[84], the future role of MEIs in atresia cases remains to be determined. However, MEIs are a proven option for hearing rehabilitation in patients with canal atresia[84 -88].

External Ear Canal Reconstruction (Canalplasty)

Canalplasty remains an option in children with canal stenosis, but caution must be taken when considering this procedure in severe stenosis or atresia, as the outcome is often unsatisfactory[84]. The risks[84,85,87] of a chronically discharging external auditory canal, re-stenosis, and residual conductive hearing loss necessitating amplification using a conventional hearing aid in over half of cases[85], have restricted the practice of canalplasty in the UK. The

subsequent use of a conventional air conduction hearing aid can also prove difficult in reconstructed ear canals[84,85]. If a canalplasty is to be considered, the surgery should only be performed by an experienced surgeon, in the presence of very favourable middle ear anatomy (intact stapes & well aerated middle ear, Jahrsdoerfer score >7-8/10)[85].

SECTION 8 – SERVICE MODELS AND CARE STRUCTURE

8.1 CURRENT UK SERVICE MODEL

The current structure of care for microtia patients varies significantly across the UK.

In Scotland, a nationally designated and funded centre exists within NHS Lothian. This provides ear reconstruction services for 5.3 million people for both congenital microtia and acquired ear loss. (Referrals are accepted from elsewhere in the UK subject to funding). The service offers inpatient surgical care within the children's hospital and adult head and neck hospital, according to age. Regular clinics are conducted both within Lothian and, on the basis of the hub and spoke model, in Glasgow, Dundee and Inverness. The service is contracted to perform reconstruction for 10 congenital cases per year and 10 acquired cases per year for the Scottish population. Thus, a minimum of 20 cases are performed annually.

In England, a number of centres have a dedicated ear reconstruction service. Currently several are in London, one in Manchester and one in Liverpool. These centres are not nationally designated or nationally funded but instead have developed along historical lines and rely on individual funding on a named patient basis. On occasion local health commissioners have been reluctant to fund reconstruction for microtia and have dismissed such surgery as "cosmetic".

In theory, there are very few barriers to stop other centres emerging and offering complex ear reconstruction. Indeed, in the recent past such centres have been proposed and some have provided care for a transient period. Concern exists within the clinical community that such services may not be able to offer optimal levels of care or, even if they do offer optimal care, they may not be constructed in a robust, sustainable and auditable manner. There is a clear consensus that parallels exist between the current provision of microtia care and the previous relatively ad hoc provision of cleft care within the UK prior to the C.S.A.G. report.[1, 89]. Evidence would suggest that the

results of ear reconstruction surgery are highly operator-dependant and that occasional operator results may be sub-optimal, to the significant detriment of patients with microtia.

Thus, it would seem logical and desirable that across the UK, units are designated as ear reconstruction centres.

8.2 RECOMMENDED SERVICE MODEL

Designated centres

Designated centres should be defined by their capacity to provide optimal care of microtia / atresia both in the out-patient environment and with regard to all aspects of in-patient care. Designated centres should include staff with the appropriate training and experience. Centres should offer regular microtia / atresia clinics, at least once a month. All appropriate surgical equipment should be available within the facility.

The discussion regarding numbers is always difficult. However, there is increasing evidence that this is an important factor. Prior to setting up the designated service in Scotland there was anecdotal evidence of poor standards for ear reconstruction. However, since the service has been nationally designated in Scotland, it is clear that consistently high quality and improved care has been delivered. This has been shown in subsequent service evaluations looking at patient and carer feedback. [90]

Surgeons must be competent to perform total and partial ear reconstructions for all congenital and acquired aetiologies. They should be able to evidence training in ear reconstruction in their training portfolio.

A recent study looking at the number of hip arthroplasties performed by a single surgeon, assessing results and complications, concluded that they should perform more than 35 per annum [91]. Similar studies in paediatric cholecystectomy, transurethral prostate resection, shoulder arthroplasty and

paediatric otoplasty have all demonstrated a similar link between the annual numbers of cases a surgeon is performing and outcome [92-96].

The soon to be published re-examination of Cleft care in the UK is near to completion and will show highly significant improvements in care following designation of units and high case volume per surgeon. There have been improvements in speech, facial appearance and dento-alveolar growth. Currently the median surgeon volume is 78 primary cleft operations per year compared to pre-designation when 75 surgeons performed small numbers with only one surgeon performing over 35 surgeries per annum. Perhaps unsurprisingly the hub and spoke model has improved access with less patient travel for hospital care[97].

The evidence certainly backs the consensus within the ear reconstruction community that there should be a minimum numbers of cases performed by a single surgeon, and that a minimum number of cases per year should be expected to be seen in clinics. Surgeons performing microtia reconstruction should perform a minimum of 20 ear reconstructions per year, of which 10 should be total reconstructions for microtia. All cases of microtia reconstruction should be assessed and presented, with the results being discussed in an open annual audit forum in order investigate this further.

'Hub and spoke' model

An aesthetically, psychologically and functionally acceptable outcome should be a reasonable expectation for every patient born or arriving into the UK with congenital microtia. The treatment pathway should be patient-focused. This can best be delivered by a carefully co-ordinated, networked MDT working in close collaboration between a designated centre and local care providers. Each patient needs a tailored plan accessing different levels of input at different time points as close to home as possible.

The design of services should be on a hub and spoke model. That is to say that for every patient there would be a designated centre (the hub) offering highly specialised aspects of care, while peripheral hospitals (the spokes)

would offer specific services at a more local level. Certain services should be offered in the designated centre, whereas other services could be provided within the designated centre or in a peripheral unit. Where services are available centrally and locally, informed patient choice should dictate where the care takes place.

Services that could be offered in peripheral hospitals includes certain ENT procedures, audiology and hearing aid care, bone anchored prosthesis surgery, maxillo-facial surgery and so on. However, other services such as surgical ear reconstruction with rib cartilage or a buried prosthetic implant should not be provided by any unit other than the main central hub. Central microtia teams should offer outreach clinics in hub facilities. This not only provides as much care close to home as possible but also facilitates regular communication with the broader team including local audiology services.

Multi- Disciplinary Team (MDT)

Designated centres should offer MDT care. Members of the MDT should include: Anaesthetist, Audiologist, Craniofacial or Maxillo-facial surgeon, Physician, Geneticist, Microtia Surgeon, Otologist, Paediatrician, Clinical Psychologist, Prosthetist, Specialist Nurse, Speech and Language therapists.

MDT clinics will vary in their members between different designated centres. However patients and their families should be fully aware that direct access to clinics comprising any member of the MDT is possible.

The table below shows a loose timetable of involvement for the core members of the MDT.

	Microtia and Atresia – Core Disciplines			
age	**Audiology**	**Otology**	**Plastic Surgery**	**Psychology**
0	early assessment support and advice, discuss options including intervention	support and advice, discuss options	support and advice, discuss options	support and advice re. appearance differences Psychological assessment and therapy as necessary
1	ongoing review and discuss potential options including intervention			
2	ongoing review and discussion of options including intervention	ongoing review	ongoing review	
3				
4		ongoing review	ongoing review	
5				
6			discuss options with child	
7				
8			ongoing review ?intervention ?suitability for surgery	support and advice re. surgery/change of appearance/ adjustment questions/teasing ?suitability for surgery
9		ongoing review (2 yearly)+/- intervention		
10				
11				
12				
13				
14				
15				
16+	review as required	review as required	review as required	review as required

8.3 FUNDING STRUCTURE

Funding for microtia reconstruction should be centralised into designated services following the Scottish model. Units should serve a population base of between 5 and 10 million. Failure to achieve this, results in a perpetuation of the post-code lottery of funding that currently exists. Patients should be allowed to elect to change their central designated units. Patients eligible for treatment whose families are unable to afford the travel and accommodation costs involved should receive financial support.

SECTION 9 - OUTCOME MEASURES

Measures to look at all surgical outcomes (hearing intervention and reconstruction), hearing and psychological outcomes to allow local and national audit should be a matter of routine. The use of validated self-reported outcome measures should also be administered as routine. Each local and specialist service should maintain a database allowing audit of the options taken by families for intervention and reconstruction.

9.1 OUTCOME MEASURES FOR HEARING AND HEARING INTERVENTION

As there are few standard outcome measures used universally across Audiology services, measures should be sought to cover the following areas.

- Device acceptability to child and parent
- Communication and early language skills e.g. common monitoring protocol (CMP)
- Language skills
- Non-verbal IQ
- Developmental progress
- Access to the classroom and teaching e.g. SIFTER
- Academic achievement
- Listening effort
- Confidence and participation

These will enable the progress of individual children to be monitored and in particular to develop the knowledge base of the impact of unilateral atresia. Efforts should be made to ascertain this information where possible, and in liaison with local teams and colleagues. Where known measures exist these should be used.

9.2 PSYCHOLOGICAL OUTCOME MEASURES

Should the child/young person/adult decide to pursue autologous ear reconstruction, it is useful to obtain a combination of qualitative and quantitative information both pre- and post-surgery. Assessment of readiness for surgery should occur before or at the preadmission clinic. A semi-structured interview format should be used to establish current emotional state, family functioning, mental and physical health history, significant life events, risk factors for poor psychological adjustment post-surgery, unrealistic expectations, concerns regarding the process of hospital admission, surgery, post-operative pain etc. As well as collating feedback for the medical team, the aim should be to attempt to ascertain if ear reconstruction will contribute to long term psychological well being. It is recognised that there is a lack of quantitative and qualitative research looking at the psychological impact of microtia, atresia and ear reconstruction and a need for longitudinal studies to further inform psychological assessments.

As well as qualitative data, standardised measures should be included at preadmission to assess current psychological state, as well as providing information for assessing change post-surgery. Where performance related outcome measures (PROMs) are available, they should include psychosocial items. An example of a validated patient reported outcome measure developed in Edinburgh in collaboration with other centres is included in appendices 1 and 1. Other examples of useful measures are the Pi-ed[98] (Paediatric Index of Emotional Distress) and the SWA-M (Satisfaction with Appearance Scale-Microtia). The Pi-ed is a standardised measure for 8-16 year olds, validated in hospital and community samples and is effectively a paediatric version of the Hospital Anxiety and Depression Scale (HADS). The SWA-M originated as the Satisfaction with Appearance Scale© developed by the Psychology Special Interest Group of the Cleft-Palate Cranio-facial Society of Great Britain, and has been adapted by the Scottish National Ear Reconstruction Service for use with Microtia (SWA-M). The Satisfaction with Appearance Scale is not standardised but has been used in a number of published trials and is considered a valuable measure of change. It is careful not to imply that a patient should be dissatisfied with their appearance, is

accessible and brief and therefore appropriate for use in clinics. Use of these measures pre- and post-operatively provides valuable data for audit and research.

9.3 RECONSTRUCTIVE SURGERY OUTCOMES: MICROTIA QUALITY STANDARDS

The following outcome measures describe markers of high-quality care that should contribute to improving the effectiveness, safety and experience of care in microtia patients undergoing reconstructive surgery. The principles underlying high quality outcomes in microtia reconstruction are twofold. Firstly, to ensure patients have a positive experience of care, and secondly, to treat and care for microtia patients in a safe environment and protect them from avoidable harm.

Expected levels of achievement for these developmental quality outcome measures are not specified. Quality standards provide a framework for continuous improvement in quality, and therefore aspirational achievement levels are likely to be 100%. However, it is recognized that this may not always be appropriate in practice taking account of patient safety, patient choice and clinical judgement. Therefore, desired levels of achievement should be defined locally.

Units should strive to promote a service that is fair, personal and responsive to patient's needs and wishes. In order to deliver a high quality reconstructive service, it is essential to ensure equality of access and quality of services. Data collection to assess this should be routine practice and subject to transparent national review. Transparency will provide evidence of service effectiveness to commissioners, and also serve to assure the public that the service, wherever provided, is safe and of an acceptable quality. Such national review would also allow performance to be benchmarked in a standardised manner, identify variance, and support quality improvement initiatives to address any variation, or unacceptable outcomes, in line with the new NHS outcomes framework.

Below we have highlighted for each quality measure where the data should be routinely collected and reviewed. Where local data collection is recommended, individual units should aim to generate comparisons of performance over time.

Quality statement 1: timeliness of care

a) Patients considering the option of reconstruction surgery should be seen by their locality ear reconstruction team within 12 weeks of referral.
b) Referral and consultations should be offered regardless of whether the patient is of a suitable minimal age for reconstructive surgery, to allow for information giving to patients and their families.
c) Patients should be informed of the anticipated schedule of each stage of reconstruction and the estimated timeframe to completion of ear reconstruction. This information can help patients in planning their surgery at a time when it will be least disruptive to school/work.
d) Once listed for reconstructive surgery, waiting time to first surgery should follow in a timely fashion that, where possible, accommodates for patients' school/work commitments. Further surgery and outpatient appointments should also adhere to the above.
e) Should patients wish referral to a different reconstruction centre, communication links should be in place to allow for expedited referral and consultation.

Data source:

a), b), c), and d): Local data collection
e): National data collection.

Equality and diversity considerations:

Should patients wish to change their reconstruction centre this should not impact on the timeliness or the quality of care. The referring unit should make information regarding all aspects of the MDT care plan available at the time of referral in order to facilitate this.

Quality statement 2: access to reconstructive services

a) Reconstructive surgery should be offered to patients with microtia irrespective of older age, geographical location and socioeconomic status, unless significant co-morbidity precludes it.

Data sources:

a): Local data collection.

Definitions:

People should receive an age-independent assessment of co-morbidity that includes performance status to determine the presence of significant co-morbidity. All areas in the UK should be assigned a reconstruction centre for referral of patients.

Equality and diversity considerations:

Ear reconstruction surgery should be based on clinical need and fitness for treatment rather than age. Treatment and care of all patients with microtia should take into account patients' needs and preferences.

Quality statement 3: support and aftercare

a) Patients having ear reconstruction surgery for microtia are offered personalized information and support, including a written follow-up care plan and details of how to contact a named healthcare professional.

b) The details of such support and the care plan should be shared with the patient's named general practitioner.

c) The named healthcare contact should be a member of the reconstructive team. Their role is to co-ordinate reconstructive care and to provide continuity of care and support. They should be easily accessible to patients, be able to offer referral to psychological services if required, and liaise with the reconstructive team and other members of the MDT.

Data source:

a), b), and c): Local data collection.

Definitions:

Personalised information and support should include:

- Details of the named healthcare professionals and how to contact them.
- Dates of any follow-up appointments or planned future surgery.
- Explanations of incidence and symptoms of post-operative complications, and who to contact if they occur.
- Practical information about how to care for their reconstructed ear including information on how to clean the ear, when they can start sport, return to work/school, and when they can go swimming.
- Where to find further sources of information and support.

Equality and diversity considerations:

All information about treatment and care should be personalised and tailored to the individual needs of the patient. The information provided should be in an appropriate format for the patient's age. It should also be accessible to patients with additional needs such as physical, sensory or learning disabilities, and to people who do not speak or read English.

Quality statement 4: care delivery

a) Patients should be satisfied with the delivery of both outpatient care and hospital care.

b) Patients should be adequately informed about how their care will be delivered and satisfied that this was met.

c) The use of patient reported experience measures to assess the above should be standard practice. Local audit should seek to identify areas to improve patient experience based feedback.

Data source:

a), b), and c): Local data collection.

Quality statement 5: surgical outcome

a) Operative results should be documented with pre and post-operative photographs for all patients undergoing reconstructive surgery. This is recommended after each surgery, and must occur in all patients after the completion of reconstructive surgery.

b) Patients should be satisfied with the aesthetic outcome of their reconstructive surgery.

c) Reconstructive surgery should aim to improve patients' self-confidence and microtia-specific behavioural issues.

d) The use of patient reported outcome measures (PROMs) to assess the above should be standard practice, and results subject to local and national audit. . An example of a validated patient reported outcome measure developed in Edinburgh in collaboration with other centres is included in appendix 1.

Data source:

a), b), and c): Local data collection.

d): Local and National data.

Definitions:

Patient reported outcome measures (PROMs) should be collected in a standardised manner across the UK. The measure should be validated and specific to microtia. It should include information on aesthetic outcome and specific psychosocial behaviours associated with microtia. Units should aim to complete such measures pre and post-operatively.

Quality statement 6: safety

a) All surgical processes should be subject to local clinical governance standards and policies.

b) Data on operative complications should be collected freely by each reconstructive unit. This information should be submitted centrally and reviewed annually amongst units. The information should be made available to service providers, commissioners and patients.

c) Units providing reconstructive surgery for microtia should provide a safe peer-reviewed service with a low incidence of intra-operative pneumothorax and an acceptable rate of post-operative complications.

Data source:

a), and b): Local data collection.

c): National data.

Definitions:

Operative complications to be collected should include:

- Pneumothorax
- Haematoma
- Infection
- Extrusion
- Exposure
- Loss of graft

SECTION 10 - REFERENCES

1. Bearn D, Mildinhall S, Murphy T, Murray JJ, Sell D, Shaw WC, Williams AC, Sandy JR. Cleft Lip and Palate Care in the United Kingdom—The Clinical Standards Advisory Group (CSAG) Study. Part 4: Outcome Comparisons, Training, and Conclusions. *The Cleft Palate-Craniofacial Journal*. 2001;38(1): 38-43.

2. Alasti F, Genetics of microtia and associated syndromes. *J Med Genet* 2009;46:361-369

3. Tanzer RC. Total reconstruction of the external ear. *Plast Reconstr Surg Transplant Bull*. 1959 Jan;23(1):1-15.

4. Tanzer RC. Total reconstruction of the external auricle*. Arch Otolaryngol*. 1961 Jan;73:64-8.

5. Tanzer RC. An analysis of ear reconstruction. *Plast Reconstr Surg*. 1963 Jan;31:16-30.

6. Tanzer RC. Total reconstruction of the auricle: a 10-year report. *Plast Reconstr Surg.* 1967 Dec;40(6):547-50.

7.Tanzer RC. Total reconstruction of the auricle. The evolution of a plan of treatment. *Plast Reconstr Surg*. 1971 Jun;47(6):523-33.

8. Tanzer RC. Microtia--a long-term follow-up of 44 reconstructed auricles. *Plast Reconstr Surg.* 1978 Feb;61(2):161-6.

9. Brent B. The correction of microtia with autogenous cartilage grafts: II. Atypical and complex deformities. *Plast Reconstr Surg*. 1980 Jul;66(1):13-21.

10. Brent B. The correction of microtia with autogenous cartilage grafts: I. The classic deformity.? *Plast Reconstr Surg*. 1980 Jul;66(1):1-12.

11. Brent B, Upton J, Acland RD, Shaw WW, Finseth FJ, Rogers C, Pearl RM, Hentz VR. Experience with the temporoparietal fascial free flap. *Plast Reconstr Surg*. 1985 Aug;76(2):177-88.

12. Brent B. Auricular repair with autogenous rib cartilage grafts: two decades of experience with 600 cases. *Plast Reconstr Surg*. 1992 Sep;90(3):355-74.

13. Brent B. Technical advances in ear reconstruction with autogenous rib cartilage grafts: personal experience with 1200 cases. *Plast Reconstr Surg*. 1999 Aug;104(2):319-34; discussion 335-8.

14. Brent B. The pediatrician's role in caring for patients with congenital microtia and atresia. *Pediatr Ann*. 1999 Jun;28(6):374-83.

15. Brent B. The team approach to treating the microtia atresia patient. *Otolaryngol Clin North Am*. 2000 Dec;33(6):1353-65.

16. Brent B. Microtia repair with rib cartilage grafts: a review of personal experience with 1000 cases. *Clin Plast Surg*. 2002 Apr;29(2):257-71

17. Nagata S. A new method of total reconstruction of the auricle for microtia. *Plast Reconstr Surg*. 1993 Aug;92(2):187-201. PubMed PMID: 8337267

18. Nagata S. Modification of the stages in total reconstruction of the auricle: Part I. Grafting the three-dimensional costal cartilage framework for lobule-type microtia. *Plast Reconstr Surg*. 1994 Feb;93(2):221-30; discussion 267-8. PubMed PMID: 8310014

19. Nagata S. Modification of the stages in total reconstruction of the auricle: Part II. Grafting the three-dimensional costal cartilage framework for concha-type microtia. *Plast Reconstr Surg*. 1994 Feb;93(2):231-42; discussion 267-8. PubMed PMID: 8310015.

20. Nagata S. Modification of the stages in total reconstruction of the auricle: Part III. Grafting the three-dimensional costal cartilage framework for small concha-type microtia. *Plast Reconstr Surg*. 1994 Feb;93(2):243-53; discussion 267-8. PubMed PMID: 8310016.

21. Nagata S. Secondary reconstruction for unfavorable microtia results utilizing temporoparietal and innominate fascia flaps. *Plast Reconstr Surg*. 1994 Aug;94(2):254-65; discussion 266-7. PubMed PMID: 8041816.

22. Nagata S. Total auricular reconstruction with a three-dimensional costal cartilage framework. *Ann Chir Plast Esthet*. 1995 Aug;40(4):371-99; discussion 400-3. PubMed PMID: 8561450.

23. Kawanabe Y, Nagata S. A new method of costal cartilage harvest for total

auricular reconstruction: part II. Evaluation and analysis of the regenerated costal cartilage. *Plast Reconstr Surg*. 2007 Jan;119(1):308-15. PubMed PMID: 17255687.

24. Firmin F. [Reconstruction of the pinna in cases of microtia]. *Rev Laryngol Otol Rhinol (Bord)*. 1997;118(1):11-6. French. PubMed PMID: 9206299.

25. Cronin TD. Use of a silastic frame for total and subtotal reconstruction of the external ear: preliminary report. *Plastic Reconstr Surg*. 1966 May; 37 (5): 339-405.

26. Cronin TD, Greenberg RL, Brauer RO. Follow-up study of silastic frame for reconstruction of external ear. *Plast Reconstr Surg*. 1968 Dec;42(6):522-9..

27. Cronin TD, Ascough BM Silastic ear construction.. *Clin Plast Surg*. 1978 Jul;5(3):367-78.

28. Wellisz T. Reconstruction of the burned external ear using a Medpor porous polyethylene pivoting helix framework. *Plast Reconstr Surg*. 1993 Apr;91(5):811-8.

29: Wellisz T. Clinical experience with the Medpor porous polyethylene implant. *Aesthetic Plast Surg*. 1993 Fall;17(4):339-44.

30. Romo T 3rd, Fozo MS, Sclafani AP. Microtia reconstruction using a porous polyethylene framework. *Facial Plast Surg*. 2000;16(1):15-22.

31. Romo T 3rd, Presti PM, Yalamanchili HR. Medpor alternative for microtia repair. *Facial Plast Surg Clin North Am*. 2006 May;14(2):129-36.

32. Reinisch JF, Lewin S. Ear reconstruction using a porous polyethylene framework and temporoparietal fascia flap. *Facial Plast Surg*. 2009 Aug;25(3):181-9.

33. Brånemark PI, Adell R, Albrektsson T, Lekholm U, Lundkvist S, Rockler B. Osseointegrated titanium fixtures in the treatment of edentulousness. *Biomaterials*. 1983 Jan;4(1):25-8.

34. Parel SM, Branemark PI, Tjellstrom A, Gion G. Osseointegration in maxillofacial prosthetics. Part II: Extraoral applications. *J Prosthet Dent*. 1986 May;55(5):600-6.

35. Parel SM, Holt GR, Branemark PI, Tjellstrom A. Osseointegration and facial

prosthetics. *Int J Oral Maxillofac Implants*. 1986 Summer;1(1):27-9. PubMed PMID: 3462128.

36. Jacobsson M, Tjellström A, Fine L, Jansson K. An evaluation of auricular prosthesis using osseointegrated implants. *Clin Otolaryngol Allied Sci*. 1992 Dec;17(6):482-6.

37. Federspil PA. Ear epistheses as an alternative to autogenous reconstruction. *Facial Plast Surg*. 2009 Aug;25(3):190-203.

38. Federspil PA. Auricular prostheses. *Adv Otorhinolaryngol*. 2010;68:65-80.

39. Korus LJ, Wong JN, Wilkes GH. Long-term follow-up of osseointegrated auricular reconstruction. *Plast Reconstr Surg*. 2011 Feb;127(2):630-6.

40. Giot JP, Labbé D, Soubeyrand E, Pacini R, Guillou-Jamard MR, Compère JF, Bénateau H. Prosthetic reconstruction of the auricle: indications, techniques, and results. *Semin Plast Surg*. 2011 Nov;25(4):265-72.

41. Si Y, Fan SC, Sun W, Chen YB, Zhang ZG. Osseointegration technique in patients with acquired auricular deformities and failed previous reconstruction: a retrospective study of long-term follow-up and Chinese experience. *ORL J Otorhinolaryngol* Relat Spec. 2012;74(3):129-35.

42. Goiato MC, dos Santos DM, Haddad MF, Moreno A. Rehabilitation with ear prosthesis linked to osseointegrated implants. *Gerodontology*. 2012 Jun;29(2):150-4.

43. Shonka DC, Jahrsdoerfer RA, Kesser BW. The Jahrsdoerfer Grading Scale in Surgery for Congenital Aural Atresia. *Arch Otolaryngol Head Neck Surg*. Aug. 2008;134:873-7.

44. Chang SO, Choi BY, Hur DG. Analysis of the long-term hearing results after the surgical repair of aural atresia. *Laryngoscope*. Oct 2006;116(10):1835-41

45. De la Cruz A, Teufert KB. Congenital aural atresia surgery: long-term results. *Otolaryngol Head Neck Surg*. 2003 Jul;129(1):121-7.

46. Hartvig Jensen J, Angaard Johansem P, Borre S. Unilateral sensorineural hearing loss in children and auditory performance with respect to right/left ear differences. *British Journal of Audiology*. 1989;23:207-213.

47. Oyler RF, Oyler AL, Matkin ND. Unilateral hearing loss: demographics and educational impact. *Language, Speech & Hearing Services in Schools*. 1988;19:201-210.

48. Bess FH, Tharpe AM. Unilateral hearing impairment in children. *Pediatrics*. 1984;74:206-216.

49. Dancer J, Burl NT, Waters S. Effects of unilateral hearing loss on teacher responses to the SIFTER: Screening instrument for Targeting Educational Risk. *American Annals of the Deaf*. 1995;140:291-294.

50. Hallmo P, Moller P, Lind, O, Tonning FM. Unilateral sensorineural hearing loss in children less than 15 years of age. *Scandanavian Audiology*. 1986;15:131-137.

51. Keller WD, Bundy RS. Effects of unilateral hearing loss upon educational achievement. *Child Care & Health Development*, 1980;6:93-100.

52. Lieu JE, Tye-Murray N, Fu Q. Longitudinal study of children with unilateral hearing loss. *The Laryngoscope*. 2012;122:2088-2095.

53. Kiese-Himmel, C. Unilateral sensorineural hearing impairment in childhood: analysis of 31 consecutive cases. *International Journal of Audiology*. 2002;41:57-63

54. Kundst SJW, Leijendeckers JM, Mylanus EAM, Hol MKS, Snik AF, Cremmers CWRJ. Bone-anchored hearing aid system application for unilateral congenital conductive hearing impairment: Audiometric results. *Otology & Neurotology*. 2008;29:2-7.

55. Priwin C, Jonsson R, Magnusson L, Hultcrantz M, Granstrom G. Audiological evaluation and self-assessed hearing problems in subjects with single-sided congenital external ear malformations and conductive hearing loss. *International Journal of Audiology* 2007;46:162-171.

56. Stenfelt S. Transcranial attenuation of bone-conducted sound when stimulation is at the mastoid and at the bone-conduction hearing aid position. *Otology & Neuro-otology*. 2012;33:105-114.

57. Van der Pouw K, Snik AF, Cremers, CWRJ. Audiometric results of bilateral bone-anchored hearing aid application in patients with bilateral congenital aural atresia. *The Laryngoscope*. 1998;108:548-553.

58. Priwin C, Stenfelt S, Granstrom G, Tjellstrom A, Hakansson B. Bilateral bone-anchored hearing aids (BAHAs): An audiometric evaluation. *The Layrngoscope*. 2004;114:77-84.

59. Dutt SN, McDermott A, Burrell S, Cooper HR, Reid AP, Proops DW. Speech intelligibility with bilateral bone-anchored hearing aids: the Birmingham experience. *The Journal of Laryngology & Otology*. 2002;116(S28):47-51.

60. Kesser B W, Krook K, Gray LC. Impact of unilateral conductive hearing loss due to aural atresia on academic performance in children. *The Laryngoscope*. 2013 Sep;123(9):2270-5

61. Ren YY, Zhao SQ, Wang DN. Development and validation of a multidimensional quality of life questionnaire for congenital aural atresia. *Acta Oto-Laryngologica.* 2012;132(7):708-714.

62. Du JM, Chai J, Zhuang HX, Guo WH, Wang Y, Liu GF. An investigation of psychological profiles and risk factors in congenital microtia patients. *Journal of Plastic Reconstructive and Aesthetic Surgery.* 2008;61:S37-S43.

63. Li DT, Chin WS, Wu JF, Zhang Q, Xu F, Xu ZC, Zhang RH. Psychosocial Outcomes Among Microtia Patients of Different Ages and Genders Before Ear Reconstruction. *Aesthetic Plastic Surgery.* 2010;34(5):570-576.

64. Steffen A, Wollenberg B, Konig IR, Frenzel H. A prospective evaluation of psychosocial outcomes following ear reconstruction with rib cartilage in microtia. *Journal of Plastic Reconstructive and Aesthetic Surgery.* 2010;63(9):1466-1473.

65. Kristiansen M, Oberg M, Wikstrom SO. Patients' satisfaction after ear reconstruction with autologous rib cartilage. *Journal of Plastic Surgery and Hand Surgery.* 2013;47(2):113-117.

66. Horlock N, Vogelin E, Bradbury ET, Grobbelaar AO, Gault DT. Psychosocial Outcome of Patients After Ear Reconstruction: A Retrospective Study of 62 Patients. *Annals of Plastic Surgery.* 2005;54(5):517-524.

67. Soukup B, Mashhadi SA, Bulstrode NW. Health-related quality-of-life assessment and surgical outcomes for auricular reconstruction using autologous costal cartilage. *Plastic and Reconstructive Surgery*. 2012 Mar;129(3):632-640.

68. Gatehouse S. *The Glasgow Health Status Questionnaires Manual*. Glasgow, Scotland: MRC Institute of Hearing Research, Glasgow Royal Infirmary; 1998

69. Wang RY, Earl DL, Ruder RO, Graham JM. Syndromic ear anomalies and renal ultrasounds. *Paediatrics*. 2001 Aug;108(2):E32.

70. Dion K. Young Children's Stereotyping of Facial Attractiveness. *Developmental Psychology.* 1973;9(2):183-188

71. Slater A, Quinn PC, Hayes R, Brown, E. Newborn infants' preference for attractive faces: the role of internal and external facial features. *Infancy*. 2000;1:265-274.

72. Smolak, L. Chapter 12: Appearance in Childhood and Adolescence. In Rumsey N, Harcourt D. *The Oxford Handbook of the Psychology of Appearance*. 2012 Oxford University Press

73. Holden-Pitt, L, Diaz J. Thirty Years of the Annual Survey of Deaf and Hard of Hearing Children & Youth: A Glance Over the Decades. *American Annals of the Deaf.* 1998 Apr;143(2):72-76.

74. Moeller MP, Tomblin JB, Yoshinaga-Itano C, Connor CM. Jerger S. Current state of knowledge: Language and literacy of children with hearing impairment. *Ear Hear.* 2007;28:740-753.

75. Small SA., Stapells DR. Maturation of bone conduction multiple auditory steady-state responses. *International Journal of Audiology.* 2008;47(8):476-488.

76. Zarowski AJ, Verstraeten N, Somers T,Riff D, Offeciers EF, Kompis M, Caversaccio MD (eds): Implantable Bone Conduction Hearing Aids. *Adv Otorhinolaryngol*. Basel, Karger, 2011;71:124–131.

77. Small SA, Hatton JL, Stapells DR. Effects of bone oscillator coupling method, placement location, and occlusion on bone-conduction auditory steady-state responses in infants. *Ear & Hearing.* 2007;28(1):83-98.

78. Mackey A. Maturation of skull properties with implications for the fitting and verification of the soft band bone anchored hearing system for infants and young children. Unpublished doctoral dissertation, University of British Columbia (Vancouver). 2013.

79. Christensen L, Richter GT, Dornhoffer JL. Update on bone-anchored hearing aids in pediatric patients with profound unilateral sensorineural hearing loss. *Arch Otolaryngol Head Neck Surg.* 2010 Feb;136(2):175-7.

80. Fuchsmann C, Tringali S, Disant F, Buiret G, Dubreuil C, Froehlich P, Truy E. Hearing rehabilitation in congenital aural atresia using the bone-anchored hearing aid: audiological and satisfaction results. *Acta Otolaryngol*. 2010 Dec;130(12):1343-51.

81. Kraai T, Brown C, Neeff M, Fisher K. Complications of bone-anchored hearing aids in pediatric patients. *Int J Pediatr Otorhinolaryngol*. 2011 Jun;75(6):749-53.

82. McDermott AL, Williams J, Kuo M, Reid A, Proops D. The birmingham pediatric bone-anchored hearing aid program: a 15-year experience. *Otol Neurotol*. 2009 Feb;30(2):178-83

83. Bajaj, Y., Wyatt, M.E., Gault, D., Bailey, C.M., & Albert, D.M. How we do it: BAHA positioning in patients with microtia requiring auricular reconstruction. *Clinical Otolaryngology*. 2005, 30, 468–471

84. Frenzel H, Hanke F, Beltrame M, Steffen A, Schonweiler R, Wollenberg B. Application of the Vibrant Soundbridge to Unilateral Osseous Atresia Cases. *Laryngoscope*. 2009;119:67–74.

85. Kiefer J, Staudenmaier R. Combined Aesthetic and Functional Reconstruction of Ear Malformations. Staudenmaier R (ed): *Aesthetics and Functionality in Ear Reconstruction. Adv Otorhinolaryngol.* Basel, Karger, 2010, vol 68, pp 81–94

86. Frenzel H, Hanke F, Beltrame M, Wollenberg B.
Application of the Vibrant Soundbridge in bilateral congenital atresia in toddlers. *Acta Oto-Laryngologica*. 2010; Early Online, 1–5

87. Verhaert N, Fuchsmann C, Tringali S, Lina-Granade G, Truy E. Strategies of Active Middle Ear Implants for Hearing Rehabilitation in Congenital Aural Atresia. *Otol Neurotol*. 2011 Jun;32(4):639-45

88. Colletti V, Soli SD, Carner M, Colletti L. Treatment of mixed hearing losses via implantation of a vibratory transducer on the round window. *Journal of Audiology.* 2006; 45:600-608

89. Clinical Standards Advisory Group. *Report on cleft lip and/or palate*. London: Her Majesty's Stationery Office, 1998:1-119.

90. Personal Communication from National Specialist and Screening Services Directorate (NSD) Procurement, Commissioning, and Facilities NHS National Services Scotland

91. Ravi B, Jenkinson R, Austin PC, Croxford R, Wasserstein D, et al. Relation between surgeon volume and risk of complications after total hip arthroplasty: propensity score matched cohort study. *British Medical Journal (Clinical research ed.)* 2014 May;348:g3284.

92. Evans C, van Woerden HC. The effect of surgical training and hospital characteristics on patient outcomes after pediatric surgery: a systematic review. *Journal of pediatric surgery.* 2011;46(11):2119-27.

93. Chen K, Cheung K, Sosa JA. Surgeon volume trumps specialty: outcomes from 3596 pediatric cholecystectomies. *Journal of pediatric surgery*. 2012;47(4):673-80.

94. Singh A, Yian EH, Dillon MT, Takayanagi M, Burke MF, Navarro RA. The effect of surgeon and hospital volume on shoulder arthroplasty perioperativequality metrics. *Journal of shoulder and elbow surgery / American Shoulder and Elbow Surgeons.* 2014 August;23(8):1187-1194.

95. Kim JH, Park JY, Shim JS, Lee JG, Moon du G, Yoo JW, et al. Comparison of outpatient versus inpatient transurethral prostate resection for benign prostatic hyperplasia: Comparative, prospective bi-centre study. *Canadian Urological Association Journal*. 2014;8(1-2):E30-5.

96. Arkoulis N, Reid J, O' Neill C, Stewart KJ. Otoplasty- The case for skin incision by higher volume operators . Submitted JPRAS. 2014

97. Personal Communication. Prof Jonathan Sandy, Oral and Dental Sciences, University of Bristol.

98. O'Connor S, Carney T, House E, Ferguson E, O'Connor R. *PI-ED: The Paediatric Index of Emotional Distress*. 2010 GL Assessment, London.

Additional References

1. Department of Health. (2011). *The NHS Outcomes Framework 2012-13*. Available at: http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/documents/digitalasset/dh_131723.pdf .

2. Department of Health. (2011) Policy paper Confirmation of Payment by Results (PbR) arrangements for 2011-12.

3. *Standards for better health*. Department of health. July 2004.

4. *The NHS Improvement Plan: Putting People at the heart of public service*. Department of Health. June 2004.

5. Darzi. L. High Quality Care for All: NHS Next Stage Review Final Report. 2008. CM 7432. Available at: http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_085825.

6. Marshal M et al. Dying to know: public release of information about quality of healthcare. Nuffield Trust and Rand, 2000.

7. Outcome measures in plastic surgery. Editors Chung K, Pusic A. Clinic Review Articles: Clinics in Plastic Surgery, April 2013.

8. A Child with Microtia and Atresia, A guide for families. NDCS (2013) www.ndcs.org.uk.

9. Guidelines for aetiological investigation of infants with congenital hearing loss identified through newborn hearing screening.
(2009)
Aetiological investigation into severe to profound permanent hearing loss in children.
(2008)
Aetiological investigation into bilateral mild to moderate permanent hearing loss in children.
(2008)
Medical evaluation of children with permanent unilateral hearing loss.
(2009)
www.baap.org.uk

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Appendix 1

Ear Reconstruction Pre-operative Patient Questionnaire.

Thank you for taking time to complete this questionnaire. Below are questions relating to your ear. Please answer as best as possible, if you do not understand any questions please leave blank.

Age: _____ **Date of birth:** _____ **Gender: M / F** *(circle)*

Reason ear reconstruction required: (e.g. Microtia/Trauma) _____

_____

Right ear/Left ear or Bilateral: _____

1. How much do you agree with the following statements regarding the ear that you are seeing the surgeon about? *(tick)*

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
I often hide my ear with my hair or a hat.					
I am anxious about attending a hairdresser due to my ear.					
I hide my ear when having a photograph taken.					
I avoid looking at my ear in the mirror.					
I feel self-conscious about my ear.					

2. How much do you agree with the following statements regarding the appearance of the ear that you are seeing the surgeon about?

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
I am satisfied with the appearance of my ear.					
I think my ear is similar to my other ear.					
I am satisfied with the size.					
I am satisfied with the shape.					
I can wear glasses/sunglasses behind my ear.					

Date completed: _____

Thank you for taking the time to fill out this questionnaire.

Appendix 2

Ear Reconstruction Post-operative Patient Questionnaire.

Thank you for taking time to complete this questionnaire. Below are questions relating to your ear and surgery. Please answer as best as possible. If you do not understand any questions please leave blank.

Age: _____ **Date of birth:** _____ **Gender: M / F** *(circle)*

Reason ear reconstruction required: (e.g. Microtia/Trauma) _____

_____

Right ear/Left ear or Bilateral: _____

PART 1

1. Since having ear reconstruction surgery, how much do you agree with the following? *(tick)*

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
I often hide my new ear with my hair or a hat.					
I feel anxious about attending a hairdresser due to my ear.					
I hide my new ear when having a photograph taken.					
I avoid looking at my new ear in the mirror.					
I feel self-conscious about my new ear.					

2. Since having ear reconstruction surgery, how much do you agree with the following? *(tick)*

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
I am satisfied with the appearance of my new ear.					
I think my new ear is similar to my other ear.					
I am satisfied with the size.					
I am satisfied with the shape.					
I can wear glasses/sunglasses behind my new ear.					
I have trouble around the area of my chest scar.					

3. How much do you agree with the following regarding the **events** surrounding your ear surgery? *(tick)*

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
The surgeon was good at explaining how surgery would be.					
I got enough pain relief after surgery.					
My new ear looks like the surgeon explained it would.					
It has been explained well how to take care of my new ear.					
I am satisfied with the care I received.					

4. If you had to choose the management of your ear again, how much do you agree with the following? *(tick)*

	Strongly agree	Agree	Neither	Disagree	**Strongly disagree**
I would have an ear made from rib again.					
I would prefer to have a prosthetic ear fitted.					
I would prefer to do nothing about my ear.					

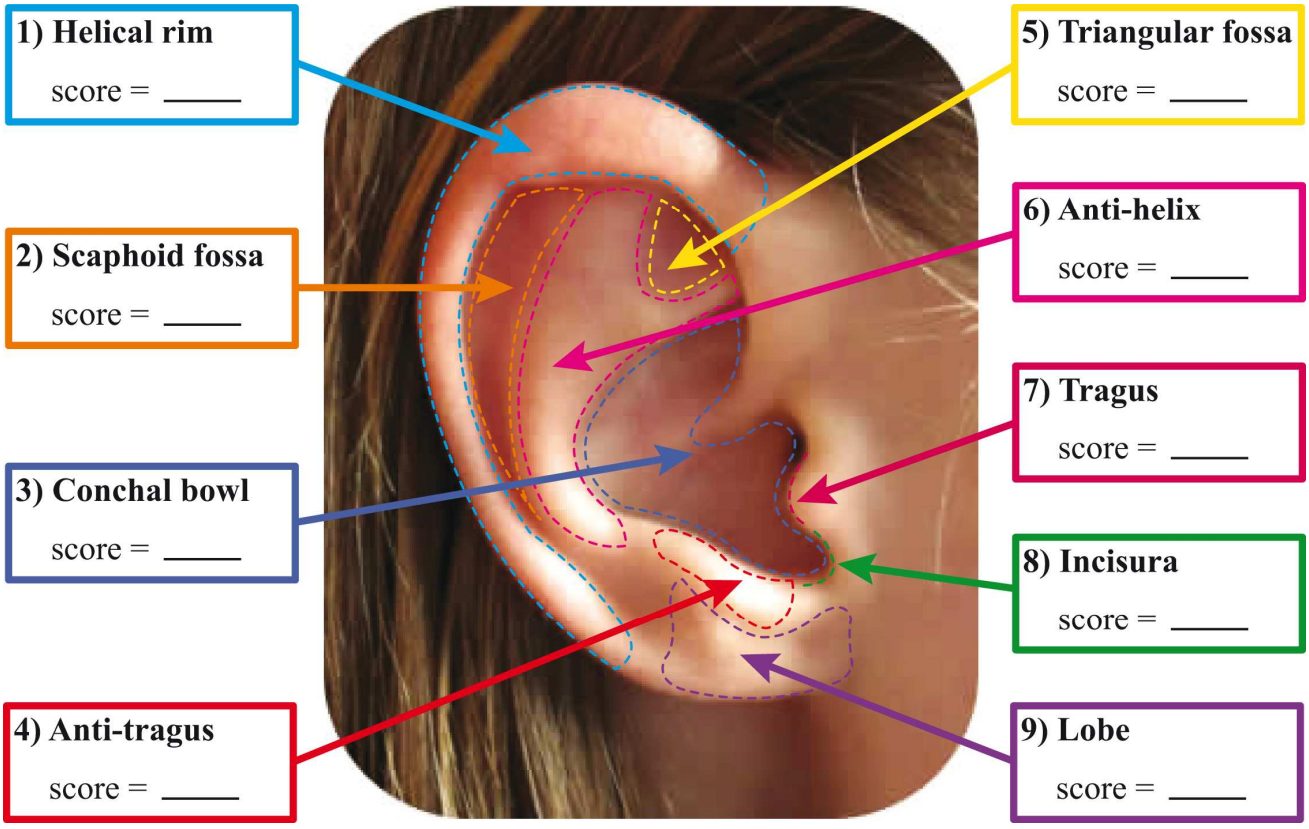
PART 2

*The following questions are also related to **appearance**, but in more detail. We would like you to answer by giving a score from 1 to 5, using the scale below:*

very dissatisfied	quite dissatisfied	alright	quite satisfied	very satisfied
1	2	3	4	5

Section A:

We've marked out **9 key areas** of a 'normal' ear below. We'd like you to find them on your reconstructed ear and give them a score using the scale above.



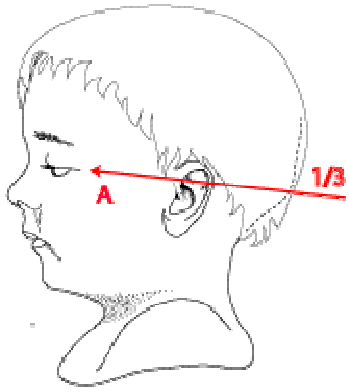
Section B:

How satisfied are you with the following *(again please use the scale above)*:

10) The size of your ear? (*Big or small)* score = _____

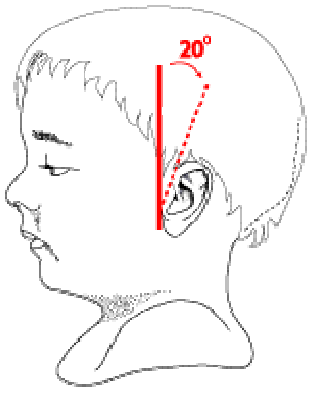
11) The projection of your ear? (*How much it sticks out*) score = _____

12) The position of your ear? (*Too high or low, see Picture 1*) score = _____



Picture 1.

13) The rotation of your ear? (*How much your ear tilts)* score = _____



Picture 2.

14) The skin covering your ear? (quality, *colour, hair*) score = _____

15) The scars around your ear and scalp? score = _____

Date completed: _____

Thank you for taking the time to fill out this questionnaire.