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Congenital Ear Abnormalities (microtia): Position Paper ENT UK 2010

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Introduction

As the NHS faces increasing financial pressure, patients and purchasers alike wish to ensure that resources are spent on those areas of healthcare with clear benefits. It would perhaps seem self evident that a child born with an absent ear warrants treatment, yet some recent documents have questioned the value for money of treating congenital ear abnormalities. This brief position paper summarises the incidence, impact and the evidence base supporting the current management of congenital ear abnormalities.

What are congenital ear abnormalities and what is their incidence?

This summary document deals with the more major congenital ear abnormalities, most commonly an underdeveloped or absent pinna (external ear), a condition also known as microtia. Microtia is unfortunately a very obvious and eye catching congenital abnormality. Most children with microtia also have an associated hearing loss due middle ear abnormalities and ear canal atresia (blockage).

The reported incidence of congenital ear abnormalities varies, with the more typical lobular or Grade III microtia (where the pinna or external ear is largely absent) occurring in approximately I in 15,000 live births.[1] In the majority of cases microtia is an isolated abnormality, although hemifacial microsomia (where one side of the face is underdeveloped) may be present to a lesser or greater degree in otherwise normal children. Microtia is more common in males, and in approximately 10% of cases the condition is bilateral. Microtia is also a feature of inherited syndromes .[2] In England in 2008-09 there were over 500 external ear reconstructions, some inevitably following tumour resection (www.hesonline.nhs.uk). The patients were aged < 15 years in 30% - ie about 150 patients in England annually are under consideration here.

Impact of the abnormality

A congenital ear abnormality affects both the child and the family of the child. When a child with a congenital ear abnormality is born, the problem causes great concern, anxiety and indeed distress to the parents. As a child becomes older he or she develops an awareness of self-image, and a desire to be 'normal'. Teasing and bullying are a well recognised phenomenon in children with congenital ear abnormalities and are often a motivation to seek surgery. Such an abnormality can have a negative impact upon a child's psycho-social behaviour.[3] In the majority of cases children with microtia have reduced hearing, and this may also affect their educational performance and possibly their safety.[4,5]

Management of Congenital Ear Abnormalities

The management is complex and multidisciplinary, requiring input from experts in Paediatric Ear Nose and Throat Surgery, Facial Plastic & Reconstructive Surgery and Audiology. On occasion other specialists may be required such as Clinical Psychologists and Prosthetists. Ideally management should take place from a clinic able to offer advice relating to all these aspects.

Management begins soon after birth. Parents find it useful and reassuring to have an informed discussion about their child's visible deformity and options for treatment. Reconstruction is not normally considered until a child is 7 years of age, at which time they are generally developing body image consciousness, themselves request surgery and are involved in the consent process.

The technique for ear reconstruction using autogenous (the child's own) rib cartilage is well established, safe and gives very satisfactory results.[6,7] Treatment may involve middle ear & ear canal surgery, a bone anchored hearing aid or a middle ear implant [8]. The psychological, social and long term quality of life benefits are well recognised and evidence based. [9-12] Children with hearing loss require a specialist audiological assessment, and advice from a Paediatric Otologist as there is increasing evidence of treatment benefit. [13]

Conclusion

A congenital ear abnormality will have both a cosmetic and functional impact upon a child due to hearing loss, and will also cause great concern to their family. A useful comparison is a child born with a cleft palate, and as in this condition a multi-disciplinary approach is required. Withholding treatment is potentially damaging for both children and their families, and risks adversely effects on education. Surgical treatment gives cosmetic, and long term quality of life benefit.

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