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Imaging in congenital deafness

One in every 1000 children has severe or profoundly severe hearing loss (greater than 50 dB hearing loss in the better hearing ear), which is detectable in the first year of life. In 90% the hearing loss is congenital and the causes are many and varied. A further one in 1000 has moderate to severe hearing loss and is not usually found until the age of 3 years.[1] Diagnosis and recognition of congenital hearing loss at the earliest possible age is essential, as is recognition of the type and degree of deafness. Any part of the hearing organ may be affected. There is a wide range of abnormalities due to arrested or abnormal development, the aetiology and pathogenesis of which is completely or partially understood in only a few cases. Children with conductive deafness can be judiciously selected for surgical correction involving a minimum of risk to normal structures if the anatomical deformities are carefully outlined.

Radiology can play a significant part in the management of the child with congenital deafness. Evaluation of the bony structures in the ear can be made with accuracy. Structural deformities of inner and middle ears frequently coexist to give both a conductive and sensorineural component to the deafness. In the inner ear, radiological evaluation of congenital deafness must assess inner ear structure related to probable cochlear function and identify those structural abnormalities which carry the risk of cerebrospinal fluid fistula. In the middle ear radiology must assess the feasibility of surgery for better sound conduction and the presence of any surgical hazards—for example, high jugular bulb, misplaced facial nerve, position of carotid artery—in addition to assessing structure and function in the favourable ear. Anatomical abnormality of the inner ear and middle ear are well recognised in association with the head and neck syndromes and at present imaging of the middle and inner ear is not routinely performed.

We suggest that patients should be selected for imaging in the following cases:

- In any syndrome known to be associated with structural deformity of the ear (see below).
- When there are certain spinal abnormalities present and including the Klippel-Feil syndrome.
- When there are abnormalities of the external ears including auricular appendages and pits.
- After attacks of meningitis and cerebrospinal fluid rhinorrhoea.

We believe that high resolution fine section computed tomography in the neonatal period is essential and that evoked response audiometry and tomography are complementary procedures.[2] Although screening tests would detect those infants with hearing impairment, imaging is the most important investigation particularly in those children for whom improvement is possible by surgical intervention.

Severe cochlear abnormalities are incompatible with

auditory function and therefore bilateral abnormalities imply that education must use methods not involving sound.

We perform high resolution fine section computed tomography using a Siemans DRH scanner and obtaining magnified 1 mm sections in axial and, when necessary, in coronal planes (fig 1). In order to reduce the radiation dose to the orbits to the minimum we recommend a plane at 30′ to the baseline, parallel with the roof of the orbit so that the globe is mostly below the sections. The dose to the cornea using this positioning is between only 0·9 and 1·8 millisieverts. We image our thin sections in the high resolution mode using a bone algorithm on a wide window setting of 3000 to 4000 Hounsfield units.[3]

For all infants selected by our suggested guidelines we believe that high resolution fine section computed tomography should be the initial investigation, and it should be performed in the neonatal period when sedation is rarely required.

At present, classification of the underlying aetiology of hearing loss in the head and neck syndromes is difficult in view of our inadequate understanding of the pathogenesis.

Cochlear abnormalities are the most commonly observed malformation of the inherited disorders (fig 2). Of the congenital disorders, the otocraniofacial group of syndromes are among the most common and most important of those with malformations of the middle and external ears.

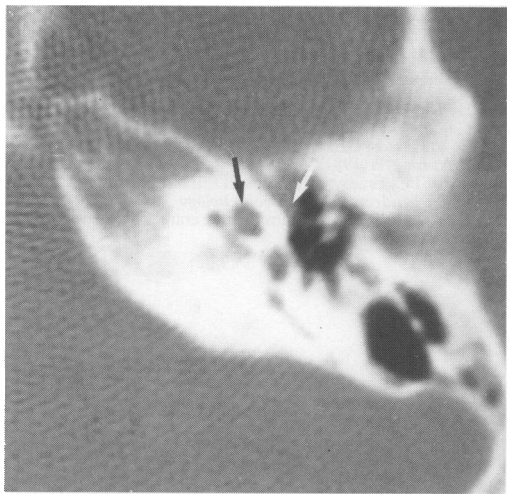

Figure 1 Normal axial computed tomogram. Arrows point to distal coils of cochlea (black arrow) and tensor tympani (white arrow).

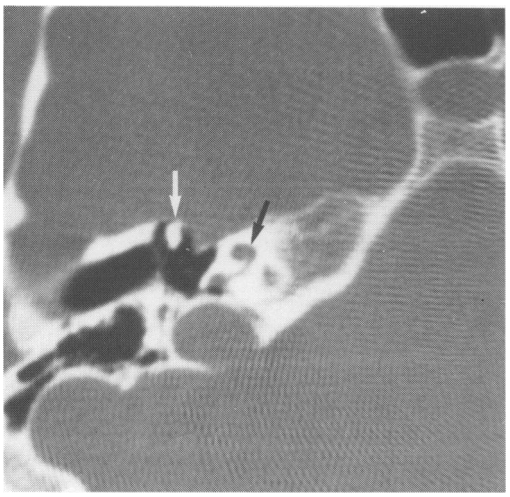

Figure 2 Branchio-otorenal syndrome (earpits' deafness). Black arrow points to small cochlea with two turns only. White arrow indicates ossicular mass attached to anterior attic wall.

This group include craniofacial microsomia (Goldenhar's syndrome), mandibulofacial dysostosis (Treacher Collins syndrome), Pierre Robin syndrome, craniofacial dysostosis (Apert's syndrome, Pfeiffer's syndrome, Saethre-Chotzen syndrome, and Crouzon's syndrome), cryptophthalmos syndrome, Waardenburg's syndrome and branchio-otorenal syndrome.

The range of abnormalities of the middle and inner ear associated with these syndromes is wide. Thus some infants with Goldenhar's syndrome are found to have a narrow internal auditory meatus and a deficient labyrinth resulting in severe deafness while others with the same underlying syndrome have global hypoplasia of the structures in the middle ears including hypoplastic or absent ossicles.[4 5] There is considerable similarity between the abnormalities in the middle and inner ears in both hemifacial microsomia and mandibulofacial dysostosis.[6] Cochlear function is usually normal. Otopathology is less common in craniofacial dysostosis than in mandibulofacial dysostosis or hemifacial microsomia.[7] Ossicular malformation or fixation is reported in the craniofacial dysostoses.[8] These children also develop conductive deafness due to serous otitis media because of impaired eustachian tube function.[9]

Otopathology occurs less frequently with primary cervical shoulder anomalies than with craniofacial malformations. Sensorineural hearing loss is more common than conductive loss.[7] This group includes the Klippel-Feil syndrome and Wildervanck's syndrome, cleidocranial dysostosis, otocervical syndrome, and Sprengel's shoulder. Severe hearing loss occurs in up to 30% of children with the Klippel-Feil syndrome. A range of abnormalities may be demonstrated by imaging including gross labyrinthine deformities such as the Mondini defect—a deficiency of the central bony spiral of the cochlea which precludes normal hearing. In addition to these malformations of the inner ear, a variety of external and middle ear abnormalities occur.[10] Infants with Wildervanck's syndrome (the combination of Klippel-Feil and Duane's syndromes) may have a particularly severe combination of abnormalities. Ossicular malformation and atretic external auditory meati have been reported in infants with cleidocranial dysostosis, both abnormalities which are clearly demonstrated on high resolution fine section computed tomography.

The ostoskeletal syndromes (bone dysplasias) include osteogenesis imperfecta, dysplasias with increased bone density, Kniest's disease, otofacial digital syndrome type II (Mohr) and otopalatal digital syndrome. Hearing loss in these infants is frequently due to compression of the inner ear and foraminal narrowing. Congenital hearing loss is also found in some chromosomal abnormalities including trisomy 13–15, trisomy 18, trisomy 21, trisomy 22, and sex trisomy (Turner's syndrome) and in chromosomal deletion 4p- (Wolf-Hirschhorn syndrome). A combination of middle and inner ear abnormalities are reported.

Deafness occurs with endocrine disorders and, in particular, hypothyroidism. Structural abnormalities of the ear have, however, not been demonstrated and the hearing loss is usually reversed by appropriate hormone treatment. The one condition in which deformities of the bony labyrinth have been shown in association with endocrine disturbances is in infants with Pendred's syndrome who have severe hearing loss due to Mondini type defects with deficiency of the modiolus.

Many drugs produce hearing loss and tinnitus, usually from temporary or permanent damage to the cochlear end organ. Thalidomide produced structural abnormalities by a teratogenic effect in utero and ear abnormalities are the second most common result of thalidomide embryopathy with an estimated 10% of patients affected. The deformities have no characteristic or distinguishing feature. They do,

however, tend to be bilateral, severe, and extensive and often involve external, middle, and inner ears.[11]

Deafness is a common feature of neurofibromatosis and is usually due to neuromas on the VIIIth nerve or bony dysplasia. The acronym CHARGE association is applied to that combination of congenital defects involving coloboma, heart disease, atresia of the nasal choanae, retarded development and/or central nervous system abnormalities, genital hypoplasia, and ear abnormalities. Computed tomography has demonstrated structural abnormalities in 50% in our series.[8] Atresia of the external auditory meatus, ossicular abnormalities, and cochlear deformities may be shown.

In some of the aforementioned conditions the hearing loss is static (for example, craniofacial microsomia) but in others, for example, the bony dysplasias it is usually progressive.

Computed tomography should be considered for any infant or child suspected of having one of these syndromes, although priority will obviously have to be given to more important defects of other systems.

We believe that in infants born with deformed pinna with or without external meatal atresia or with manifestations of one of the head and neck syndromes, imaging in the neonatal period should be performed, preferably before discharge home from the maternity unit. Examination at this stage has the following advantages:

- Sedation is rarely required.
- A full assessment of the middle and inner ears may be obtained with a few tomographic sections.
- The information obtained can be correlated with hearing assessments performed from 1–2 years of age.

The optimum age for imaging is difficult in those children in whom hearing loss is detected at a later age, particularly when the deficit is only detected at the age of 2–3 years when sedation is usually required. Severe labyrinthine deformities preclude any auditory function. Surgical attempts to improve sound conduction mechanism is rarely undertaken in unilateral malformations if the hearing in the other ear is normal.

Accurate knowledge of the structure of the inner and middle ears is essential from the earliest occasion possible, such that those infants in whom structural malformation precludes any hearing are detected and educated from the earliest age with methods not involving sound.

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