Hearing screening in children—state of the art(s)

SIR,—Haggard emphasized the extreme importance of early identification of hearing impairment in infants at risk of sensorineural hearing loss and advised evoked response audiometry as a sensitive and specific determinant. 1 Reardon and Pembrey, in the same edition of the journal, documented that 30% of all genetically determined deafness occurs in syndromic form.2 Anatomical abnormalities 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. Although screening tests would detect those infants with hearing impairment, for children with head and neck syndromes or external ear deformities it can only be complementary to imaging, which is the most important investigation, particularly for children in whom improvement is possible by surgical intervention. We believe that for all infants with syndromes associated with ear abnormalities—for example, Klippel-Feil, Treacher Collins, craniofacial microsomia, Wildervanck, Goldenhar, X-linked deafness—or for infants with external ear deformities, that conventional tomography or high resolution fine section computed tomography in the neonatal period is essential, and that evoked response audiometry and tomography are complementary procedures.3

We have previously reported our series of 17 patients with Klippel-Feil syndrome who underwent hypoplastic or absent cochlea and axial computed tomography and in whom we found a high proportion of severe inner and middle ear abnormalities, many of which were incompatible with any degree of hearing (RR Phillips, PD Phelps, paper presented at the 50th annual meeting, Royal College of Radiologists, Liverpool 1989). In the inner ear there was a range of abnormalities, the most severe including dysplasia of the bony spiral of the cochlea (known as the Mondini defect), which occurred in six of our patients, or absence of any recognisable cochlear structure (the Michel deformity), which we demonstrated in five patients. There was a range of severity of middle ear abnormalities, which included abnormal ossicles in six patients, absent oval window in two, and small or slit attic in two.

Severe cochlear abnormalities are incompatible with auditory function and therefore bilateral abnormalities imply that education must use methods not involving sound. The Mondini defect allows some degree of cochlear function. For some patients with conductive deafness, surgical correction of the structural abnormalities may be successful in providing a degree of auditory function.

The radiological assessment of congenital deafness must assess inner ear structure, cochlear function, and the risk of cerebrospinal fluid fistula due to structural abnormalities. In the middle ear it must assess the feasibility of surgery for better sound conduction and the presence of any surgical hazards—for example, high jugular bulb. Any deformity of the bony labyrinth indicates a severe degree of sensorineural deafness and is therefore a relative contraindication to surgical attempts to improve the sound conduction mechanism.

Thus in patients with head and neck syndromes or external ear deformities, the early radiological assessment of the inner and middle ears (by conventional imaging and computed tomography in the neonatal period) is essential in order that the range of structural abnormalities may be demonstrated. At this stage imaging may be performed with little, if any, sedation and with a minimal radiation dose. Guidance may then be given regarding the possibility of corrective middle ear surgery and to allow the best methods of communication to be taught from the earliest opportunity.

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