BASIC MEDICAL SURVEILLANCE ESSENTIALS FOR PEOPLE WITH DOWN'S SYNDROME.

HEARING IMPAIRMENT

(One of a set of guidelines drawn up by the Down's Syndrome Medical Interest Group. Approved by BACDA and BAAP. Sept 2000. Reviewed and revised by BACDA 2004)

1. Well over 50% of people with Down's syndrome have significant hearing impairment which may be mild, moderate, severe or profound. Sensorineural and/or conductive loss may be present at any age. [3.4.5.7.17]. Hearing impairment can be successfully managed in this population. If undetected it is likely to be a significant cause of preventable secondary handicap [3.10.12.13.21]. Lifelong audiological surveillance is essential for all. The main cause of conductive loss is persistent otitis media with effusion (OME, glue ear). The natural history of OME and response to intervention differ from that in the general population hence local surveillance and management protocols need to be set up specific to people with Down's syndrome.[3.5.9.19]

2. People with Down's syndrome of all ages should have rapid access to specialist audiology services [3].

3. Because of an increased incidence of congenital sensorineural loss newborns should be included in targeted newborn hearing screening programmes wherever universal newborn hearing screening is not yet in place.[1.14]. This does not preclude the need for ongoing surveillance [8].

4. Guidance for parents of children with Down's syndrome should include discussion about hearing problems and their management, supported by good quality written information.[15]

5. All babies, regardless of any previous hearing screening results, should have a full audiological assessment between age 6 and 10 months. This should include measurement of auditory thresholds, impedance testing and otoscopy [18]. To ensure inclusion of the child with Down's syndrome participation in existing child health hearing surveillance programmes should be encouraged.

6. Therefore by 10 months it should have been established whether or not a child has any degree of permanent hearing loss with or without OME. A clear management plan must have been agreed with the parents and intervention instigated where necessary.

7. In the second year (usually around 18 months) all children – whatever their previous hearing status - should have further audiological review carried out in a manner appropriate for a child with learning disability. This should include assessment of auditory thresholds, impedance testing and otoscopy. This should be repeated at least yearly until age 5 and thereafter 2 yearly for life. More frequent testing will be necessary if problems exist.

8. Transition of care from paediatric to adult services should involve direct transfer of care to a named person.

9. At all ages people with Down's syndrome have narrow ear canals which predispose to accumulation of wax[4]. This may affect impedance testing and hearing.

10. Most people with Down's syndrome are able to respond to standard tests – eg distraction; speech discrimination; pure tone audiometry (*play or standard*); and visual reinforcement audiometry – as long as these are carried out by testers with expertise in working with people with learning disability. Threshold measurement tests appropriate to developmental age must be used [6.20].

11. Because of increased incidence of sensorineural as well as conductive loss the frequency range tested should include 8000Hz whenever feasible as this may be an early warning of impending sensorineural deafness [11.22].

12. Diagnostic Auditory Brain Stem (ABR) responses in people with Down's syndrome must be interpreted with caution [7.22]

13. As in the general population all those who are hearing impaired should have access to specialist hearing support services (Speech and Language Therapy; Teachers of the deaf; hearing Therapists etc)

14. At all ages particular attention should be paid to the treatment of suppurative nasal and ear conditions [3.16].

15. In adults with the syndrome hearing assessment is essential in the differential diagnosis of depression and dementia [7].

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Enquiries to:
Down's Syndrome Medical Information Services
c/o Staff Library, Duncan Macmillan House, Porchester Road, Nottingham NG3 6AA
0115 9691300 ex 13268 info@dsmig.org.uk