FACTSHEET

Hearing loss in children with Trisomy 21 (or Down syndrome)

Hearing loss is common in children with Trisomy 21 (also known as Down syndrome). Children with Trisomy 21 have issues with speech and language development. If there is also a hearing loss, then a very severe language delay can occur. All children with Trisomy 21 should be seen regularly by both the family doctor and a paediatrician.

Do all children with Trisomy 21 have hearing loss?

Many children and adults with Trisomy 21 have temporary, permanent or even fluctuating hearing loss. There are a number of causes of this hearing loss.

How severe is the hearing loss?

Hearing loss may be mild or on one side only, up to profound and permanent hearing loss in both ears. It may also be a **fluctuating** hearing loss ie, reasonably good hearing at times and poor hearing at other times.

How will a hearing loss effect my child with Trisomy 21?

As almost all children with Trisomy 21 have some degree of intellectual disability, they will have more difficulty developing speech. If a child with Trisomy 21 does not have a hearing loss the child's ability to develop speech is based on the level of intellectual ability (ability to learn). A child with a mild intellectual disability (and no hearing loss) usually develops a good amount of speech and can both read and have a simple conversation with family or friends. If a child with Trisomy 21 has a very severe degree of intellectual disability, they are likely to have trouble developing spoken language, and may need to use signs and/or pictures (augmentative communication) to communicate. However, if a child with Trisomy 21 has hearing loss as well as intellectual disability, then they will have more difficulty with speech development, unless we treat this problem very quickly. Constantly watching for hearing loss in any child or adult with Trisomy 21 is essential. Even adults with Trisomy 21 can develop hearing loss for the first time in adult life. How do we know if a child with Trisomy 21 has hearing loss?

A permanent hearing loss is often picked up at the newborn screen of hearing (SWISH). Unfortunately, many children with Trisomy 21 develop hearing loss after birth. These children may be picked up by behavioural testing (the "puppet" test) or play audiometry with an audiologist. If the child is not able to do behavioural testing, hearing can be tested (if needed) by special electrophysiological testing (brainstem auditory evoked responses or electrocochleography). These tests are done while a child is sedated or under an anaesthetic.

What types of hearing loss do children with Trisomy 21 have, and how do we treat them?

Some children with Trisomy 21 will have a permanent **sensorineural** hearing loss. A sensorineural hearing loss is caused by a problem in the inner ear (cochlea or auditory nerve). These children may need hearing aids from birth.

Many other children with Trisomy 21 have poor eustachian tube function and small nasal passages. These children become congested, often for long periods and fluid builds up in the middle ear (behind the ear

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drum). All children with allergic rhinitis ("hay fever" or "sinus") are at particular risk.

When the child with Trisomy 21 has to listen through this layer of fluid (which may be thick and sticky ie; glue ear) they often have a big hearing conductive loss (as much as 50 decibels). Ear, Nose and Throat (ENT) Surgeons may treat this hearing loss by inserting ventilating tubes ("grommets") (sometimes needed repeatedly). Antibiotics may also be needed when there is an acute infection.

If the child often accumulates middle ear fluid, the ENT Surgeon may request the child's audiologist to fit hearing aids. If hearing aids are fitted because of chronic middle ear fluid, the hearing should be tested often (at least once every four months) to check if the middle ear fluid is still present. An ENT Surgeon will also need to check the condition of the eardrums often. If the child has repeated or constant discharge from the ear or if the eardrum collapses inward over the middle ear structures (atelectasis) then surgery might be needed. Surgery can correct this problem and prevent further ear disease developing.

Can a child with Trisomy 21 have permanent/long term conductive hearing loss?

Despite careful management, many children with Trisomy 21 seem to have some degree of conductive hearing loss each time they are tested ie, hearing loss due to problems in the middle ear. This may go on for many years and sometimes it may -be permanent. Rarely, there may be structural abnormalities of the middle ear in children with Trisomy 21, but usually permanent or long term conductive hearing loss occurs because the mucous membrane lining the middle ear is always slightly congested or swollen, or the eardrum has become scarred or stiffer than normal. If long term or permanent conductive hearing loss occurs in children with Trisomy 21 hearing aids are usually helpful.

Surgical advances now mean that implantable hearing devices, which give a clearer, sharper and more reliable sound than hearing aids, are available in certain situations.

Glossary

Conductive hearing loss- Hearing loss caused by a problem conducting sound waves through the outer or middle ear.
E**ustachian tube**- A passage that links the ears to the back of the nose .
Intellectual disability- A child with an Intellectual Disability learns and develops more slowly than other children.
Play audiometry- a hearing test for young children
Electrophysiological testing - A test of the auditory system. It looks at electrical activity generated in response to sound. It is performed in natural sleep or under sedation, or under General Anaesthesia
Sensorineural hearing loss- Hearing loss caused by a problem in the inner ear.

What are "mixed" hearing losses in children with Trisomy 21?

A 'mixed' hearing loss means that the electrical (sensorineural) and the mechanical (conductive) parts of the ear are both not working well. The sensorineural loss is permanent and the conductive loss may be temporary, permanent or fluctuating. The sensorineural loss needs a hearing aid and often this can be adjusted up to help the conductive loss. Fluctuating hearing loss is difficult to handle because at times the hearing loss will naturally improve and the amount of power that the hearing aid provides will be too much.

Children with Trisomy 21 who have a mixed hearing loss have some permanent hearing loss due to abnormalities in the inner ear ie, the cochlea or the auditory nerve. Together with this permanent hearing loss, they have an extra hearing loss due to problems with the middle ear. In these children, we aim to treat the middle ear problems if possible, but the hearing aids should be set to cater for the extra degree of hearing loss. A fluctuating hearing loss often seems to make language development difficult for a child with Trisomy 21. If speech is not developing in a child with mixed hearing loss, other ways to communicate should be introduced along with spoken words.

A speech pathologist or teacher of the deaf may also be very helpful and may recommend Makaton signs, photos or picture systems to help with communication.

Close collaboration between the audiologist, speech pathologist, family doctor, paediatrician, ENT surgeon and carers will help make the best treatment decisions.

Remember:

- Children with Trisomy 21 often have temporary, fluctuating or permanent hearing loss. This may lead to very severe speech delay if left untreated.
- Children with Trisomy 21 who have middle ear disease need regular hearing tests and supervision by both an ENT surgeon and a paediatrician.
- Fitting hearing aids in a child with Trisomy 21 is often very helpful in maximising the child's opportunity to hear well and to develop good speech. For information and fitting of hearing aids contact Australian Hearing 131 797.