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Identifying infant hearing loss – never too early, but often too late

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Infant hearing loss is one of the most frequent disorders at birth, with an estimated 17 babies born with hearing loss every day in South Africa.[1] Approximately 6 babies in every 1 000 live births in developing countries will present with a significant bilateral hearing loss. If milder losses are included, this figure increases markedly.[2] Unfortunately the invisible nature of the condition makes it undetectable by clinical examination and it only becomes apparent once secondary symptoms such as delayed speech and language or behavioural problems appear. These are often exacerbated by inappropriate advice to 'wait and see' by clinicians who are unaware of the critical window of opportunity for spoken language acquisition, which must be accessed in the first 12 months of life.

The devastating effects of undetected infant hearing loss, of any degree, must be understood in the light of the critical first few months of life when an infant absorbs and assimilates language from the environment. Any hearing loss that is not detected and does not receive intervention within the first year of life may result in significant and persistent delays in language development.[3] As language is the cornerstone of literacy and academic performance, children with late-identified hearing loss are restricted to limited educational and vocational outcomes. Although the condition is not life threatening, those affected by it face limited opportunities, isolation and stigmatisation during their entire lives, while societal costs are significantly greater owing to increased educational costs, loss of income, and limited contribution to the economy.[3]

Fortunately, if infant hearing loss is identified early and followed up by timely intervention children can have outcomes on a par with those of their hearing peers.[3] Studies have demonstrated that intervention in the form of amplification with hearing aids or cochlear implants, followed by family-centred early intervention services initiated within the first 6 - 9 months of life, leads to significantly better outcomes compared with late-identified children who exhibit persistent delays in language, speech and socio-emotional development.[3] Therefore, screening newborns for hearing loss has been implemented as standard of care in countries such as the USA and UK where close to 95% are screened before discharge from hospital.[4]

South African estimates indicate that fewer than 10% of newborns have any prospect of being screened for hearing loss, which translates to 15 babies born with hearing loss every day who will be sent home without parents or professionals aware of the babies' condition.[1] Risk factors may give an indication of children who are at risk of permanent hearing loss in 50% of cases.[3] Common risk factors include: family history of permanent childhood hearing loss; admission to neonatal intensive care unit for more than 5 days; *in utero* infections (cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis); any craniofacial anomaly, especially those related to the ear or temporal bone; physical findings associated with a syndrome known to cause hearing loss; neurodegenerative disorders; meningitis; head trauma; and chemotherapy.[3] In addition to these risk factors it is important to recognise that any concern by a caregiver with regard to a child's hearing, speech, language, or delayed development warrants immediate attention and referral for screening or assessment with regard to hearing.[3]

The only reliable means to screen hearing in newborns and young infants is by way of two electrophysiological techniques – otoacoustic emissions (OAEs) and auditory brainstem responses (ABRs). OAEs entail a single probe inserted into the ear canal, measuring the active properties of the outer hair cells in the cochlea as low-level acoustic signals. ABRs require a probe in the ear and three electrodes placed on the scalp to measure the change in electroencephalic signals in response to sound. Automated devices are available that can provide a pass or refer response in less than 1 minute. If an infant is referred for a screening test or a physician or health care professional is concerned about the child's hearing, a referral should be made to an audiologist for a diagnostic hearing assessment to determine the type, degree, and configuration of hearing loss. If a medical condition of the ear, such as otitis media, is suspected a referral to an ENT specialist should be made.

Intervention for infants with hearing loss aims to provide access to auditory input through amplification devices as soon as possible. These may include hearing aids fitted to match the unique hearing loss of the child, or in cases of severe to profound hearing loss a cochlear implant that electrically stimulates the auditory nerve in response to sound. This process must be accompanied by and followed up with family-centred communication intervention by trained interventionists (e.g. speech therapists or early interventionists) who assist parents to facilitate auditory skills and language development for their child.

The combination of 21st century technology and dedicated family-centred early intervention has made it possible for infants with hearing loss to access the hearing world and to have equal opportunities and outcomes compared with their hearing peers.[4] Prompt referral and early identification is the first step to ensuring these outcomes – it is never too early for a hearing test, but it may be too late.

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'Doctor, my ear is blocked' - when to panic

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There are very few ear-related emergencies in ENT; however, when the eardrum appears normal, an innocuously sounding 'blocked ear' may just be one.

The causes of a blocked ear may be divided into three broad categories: (*i*) arising from the ear (otological); (*ii*) arising from other adjacent head and neck structures (non-otological); and (*iii*) idiopathic.

Otological causes

Otological causes are described logically by following the anatomy of the ear from the external to the middle to the inner ear.

Anything that physically blocks or compromises the conduction of sound waves to the eardrum will cause a blocked sensation. Wax, a foreign body, squamous debris in the ear canal and swelling of the canal due to otitis externa are common causes. It is interesting that even what seems on inspection to be a minute amount of wax can induce this blockage. It is particularly the case when the wax lies on the inferior aspect of the canal or is in contact with the tympanic membrane. Ear drops that precipitate can form a visible membrane on the tympanic membrane and compromise its movement. Otitis externa is mainly due to mechanical trauma (finger or pen scratching) or chemical trauma (swimming – water). The significant pain and canal swelling with abundant discharge and squamous debris blocks the ear.

A perforation of the eardrum, barotrauma or haemotympanum due to trauma, sudden shock waves or inadequate equalisation often leads to a blocked ear. A further cause of a conductive hearing loss is ossicular chain disruption after trauma. Middle-ear fluid may be serous, mucoid, glue like or purulent in acute and chronic otitis media and even contain CSF (after trauma). These conditions lead to conductive hearing loss and are often treatable and reversible.

Nerve (sensorineural) hearing loss is a highly significant cause of a blocked ear sensation and it is vital to recognise it immediately when the onset is acute. This hearing loss, particularly in the low frequencies, is interpreted by the patient as either a blocked feeling, sensation of wax, or foreign body in the ear. Patients who develop sudden sensorineural hearing loss (SSNHL) which is caused by a viral inflammation of the cochlea are surprisingly well and have no associated symptoms such as pyrexia, rash, or dizziness. Herpes simplex type 1 is implicated. Otoscopy is absolutely unremarkable. A patient with a 'normal' examination is not commonly sent for an audiogram and only if tuning fork testing with the Rinne and Weber tests is used, will a diagnosis of hearing loss be made. Unfortunately this is not done by many doctors. The drum looks normal and commonly a diagnosis of eustachian tube dysfunction is made. Topical and systemic decongestants and even antibiotics are prescribed and sadly the diagnosis and opportunity for reversible treatment are missed. Herpes zoster is implicated in rare cases and typical zoster vesicles appear in the ear canal and on the eardrum. Nausea, vomiting and dizziness may be present only if the vestibular system is involved (complete labyrinthitis). This makes the diagnosis easier.

There is level 2 evidence of a good chance of recovery of the hearing loss if treatment for SSNHL is given within 24 hours and perhaps within 1 week of onset. If treatment is withheld, the hearing loss will be permanent in 50 - 60% of patients. High-dose corticosteroids (60 - 100 mg/day) are used for 5 - 10 days. There is no good evidence that the addition of antiviral agents improves the outcome. However, when faced with the prospect of permanent deafness, many doctors will add antivirals for 5 days. Vascular aetiologies have been implicated but not proven. There is a range of therapeutic modalities available, none of which are evidence based. These include carbogen (combination of 95% oxygen and 5% carbon dioxide) to improve blood supply to the cochlea, vasodilators, haemodilution, anticoagulants, and homoeopathic and herbal remedies.

Ototoxic medications, e.g. chemotherapeutic agents (e.g. Cisplatin), anti-inflammatories, and aminoglycoside antibiotics, will affect hearing. Rarer causes of SSNHL are tumours of the 8th cranial nerve (acoustic neuromas) and brain tumours.

Non-otological causes

It is essential to consider adjacent structures in the head and neck when the eardrum appears normal and no otological cause can be found. The temporomandibular joint (TMJ) lies immediately anterior and adjacent to the external ear canal and is separated by a thin bony wall and capsule. Inflammation in this joint is often referred to the ear, causing a 'blocked ear' sensation. Common causes are trauma to the jaw, malocclusion, new dentures, bruxism and excessive clenching, and recent extensive dental work with patient's jaw open for a lengthy time. The ligaments in the jaw joint are supplied by the same cranial nerve as that which supplies the two tiniest ligaments in the body, i.e. stapedial tendon and tensor tympani muscle. These two middle-ear tendons retract the eardrum and stapes to dampen sound and block loud noise. With TMJ ligament and tendon spasm, this can induce spasm in these two middle-ear tendons, leading to a blocked ear sensation. Impacted wisdom teeth and other related dental problems may refer to the ear. Oral and hypopharyngeal lesions can also refer to the ear.

Neck spasm, whiplash injury and recent cervical spine surgery can all cause a referred blocked feeling in the ipsilateral ear that should also be considered.

Summary

When the eardrum and otological examination appear normal and the patient is surprisingly well, you should have a high index of suspicion of a SSNHL. If you are unable to use tuning forks, urgently refer to an audiologist for a hearing test. You will have a very grateful and relieved patient if you confirm your early diagnosis and institute immediate treatment.

The dizzy patient – a 5-minute approach to diagnosis

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It's 17h00 on a busy Thursday afternoon in your practice. You're running about 40

minutes late, there are 6 patients in the waiting room, and you are scheduled to assist at a caesarean section at 18h15. As your next patient walks in you sink in your chair and breathe a deep and disguised sigh. Mr Jones is a frail elderly man with multiple medical problems and his opening remark is: 'Doctor, I'm feeling very dizzy'.

Dizziness is one of the most confusing and poorly understood conditions encountered in medical practice. The topic is not well taught at medical school and most of us are at a loss when seeing patients with this condition.

Understanding the body's balance control systems will enable doctors to diagnose and manage the condition. This article presents a 'nuts and bolts' approach that is simple, reliable and appropriate for a rushed consultation.

There are two vital principles to remember about dizziness: (*i*) it can be classified into two broad categories – otological (arising from the ear) and non-otological; and (*ii*) the diagnosis is almost always made on history alone and then confirmed on examination.

Definition

Dizziness has different meanings for different patients – from disequilibrium, light-headedness, feeling off-balance, woozy, shaky, 'not quite right', or 'not myself', to a vertigo sensation of gentle swaying and ground moving upwards, the world spinning around, or violent incapacitating rotatory vortex feelings.

Balance control systems

There are four major inputs into the brain's balance control system and centre, i.e. (*i*) the eyes; (*ii*) receptors in muscles and joints; (*iii*) the cardiovascular system; and (*iv*) the ears.

- The visual system plays a critical role in our balance and orientation in space (vestibulo-ocular reflex). It enables us to see where we are and, more importantly, where our feet are when standing and walking. Elderly patients with deteriorating night vision and, for example, cataracts are unable to discern uneven surfaces and fall often, especially at night in poor light.
- Essential messages are relayed to the brain on a continuous split-second basis via the spinal cord from every position and stretch receptors in the joints and muscles of our limbs and spinal column (vestibulospinal system). The proprioceptors in the ankles, feet, knees, hips, and neck are fundamental in 'telling' the brain where the body, and especially the head, is positioned in space. A recent hip or knee replacement, loss of proprioception in the feet in the elderly or diabetics, or a whiplash injury to the neck will lead to a sense of momentary disequilibrium and 'dizziness' that lasts split seconds or persists. Patients feel 'disorientated'.
- It is crucial to exclude both hypertension and hypotension and associated cardiac arrhythmias. Vascular abnormalities such as atherosclerosis, emboli causing micro-infarcts, larger cerebrovascular accident, and vertebrovascular insufficiency are not uncommon causes of both light-headedness and true vertigo. An important principle is that most minor vascular events associated with vertigo are discrete, begin suddenly, and end very abruptly within hours to a few days. The patient will feel absolutely normal and be able to resume normal activities. Some types of otological vertigo tend to persist and gradually improve with time, but with residual symptoms.
- The vestibular system of each inner ear (semicircular canals, utricle and saccule) controls rotatory movements and orientation in, to and fro, up and down, and angular movements. Hence, disturbances of the ear usually result in a perceived sensation of rotation (vertigo). In each inner ear vestibular system there is an equal and opposite stimulation and inhibition between the left and right side with every movement of the head and body. In vestibular neuronitis (involving the vestibular system only) or labyrinthitis (involving the entire labyrinth: vestibular system and cochlea with associated hearing loss) one inner ear does not function and the relative 'overstimulation' of stimuli from the normal ear to the brain produces excessive vertigo. It takes days to weeks for the brain to suppress the stimuli from the normal ear while the damaged ear recovers. In benign positional vertigo each episode lasts 15 - 30 seconds and the patient is normal between episodes.

History

A thorough, targeted history will differentiate otological from non-otological causes and will usually provide the diagnosis.

Rule out the non-otological causes first. Are there any visual abnormalities such as cataracts, recent eye surgery, or dizziness at night? Is there a history of head and neck trauma, neck spasm, joint surgery or replacement, arthritis, diabetes, or peripheral vascular disease affecting the proprioceptors and mechanoceptors? Are there episodes of angina, palpitations, postural hypotension, or amaurosis fugax? Were the episodes of vertigo discrete with a sudden onset and an equally abrupt end? A detailed medication history is vital. This may give a clue as to the underlying condition. In most circumstances, a recently added drug (e.g. an antihypertensive), or the interaction and side-effects of more chronic drugs, is the cause of the dizziness and simply changing or discontinuing these drugs is all that is needed.

With regard to otological causes, there are three common pathological conditions that must be excluded on history. With acute vestibular neuronitis or labyrinthitis the patient usually wakes in the early morning with debilitating vertigo, nausea and vomiting with or without hearing loss. Symptoms are incapacitating for hours to days and generally subside completely after a few weeks of unsteadiness and a feeling of disassociation. Benign positional paroxysmal vertigo (BPPV) is experienced as an unpleasant torsional vertigo lasting 15 - 30 seconds that only occurs when the patient places his/her head in a certain position. This happens when he/she looks up (also called high-shelf vertigo) or classically when lying in bed and rolling from one side to the other or rising. In between these dizzy positioning episodes the patient 'feels perfect'. In Meniere's disease, patients can have sudden episodes of typical symptoms including the sensation of fullness in the ear, vertigo, fluctuating hearing loss and tinnitus.

Examination

This will include systemic, cardiovascular, neurological, ophthalmic, and vestibulospinal examination. Otoscopy, tuning fork and basic balance tests such as the Dix Hallpike (for BBPV), Romberg's and Unterberger's tests will confirm or exclude otological causes.

Management

This is beyond the scope of the article but is targeted at treating the underlying cause.