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Sudden Sensorineural Hearing Loss – Causes – a Two Years Study – Treatment Results

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Sensorineural Hearing Loss may be either sensory (affecting the sense organ of hearing in the cochlea) or neural (affecting the nervous pathways which connect the cochlea to the hearing centers in the brain). Generally, sensorineural hearing loss is described as sudden if you notice a drop in your hearing instantaneously or over a short period not exceeding three days. You may notice a popping sound when it happens, or you may detect it when you first wake up or try to use the impaired ear. Sudden Sensorineural Hearing Loss (SSHL) is a medical emergency and you will need to contact your doctor immediately. People with Sudden Sensorineural Hearing Loss often describe themselves as 'deafened'. They may also say they have an 'acquired hearing loss'. If you lose your hearing after learning to speak, you are said to have 'post-lingual hearing loss'. It is usually unilateral but it can be bilateral in about 4-17% (20% in our cases). The loss of hearing may be permanent , or the hearing may spontaneously return to normal or near normal.

<u>What causes sudden sensorineural hearing loss?</u>
Sudden Sensorineural Hearing Loss with no known cause: Most of the time the cause is unknown – it is only possible to establish a firm cause of Sudden Sensorineural Hearing Loss in about 15% of cases. The remaining 85% are referred to as idiopathic (with no known cause). In the majority of these idiopathic cases, the hearing is affected only one ear (unilateral) and will improve spontaneously. It can be tempting, therefore, to adopt a 'wait-and-see' policy, but you should be seen by a specialist as soon as possible if you experience SSHL, because it is important to establish a diagnosis since treatment may be needed if a specific cause is found. In an attempt to find a reason for these idiopathic cases of hearing loss, two main theories have been advanced. The first theory is that it may be of viral origin, as some viruses are known to damage the hearing, and a viral infection seems to precede the hearing loss in some cases. The second theory is that it may be due to some vascular defect (relating to blood vessels). There is no conclusive proof to support either view.

Infections : Meningitis is one of the commonest

causes of severe or profound acquired deafness in infants and children. Hearing loss due to meningitis usually affects both ears (bilateral). Anybody who has suffered from meningitis, especially a child, should have their hearing tested as soon as possible after recovery. Measles and mumps are also associated with SSHL. In measles, the loss is usually moderate to profound and bilateral, whereas in mumps it usually affects only one ear. The herpes zoster virus may on rather rare occasions produce a SSHL accompanied by weakness of the facial muscles and vertigo. This so-called Ramsay Hunt syndrome may affect adults who have been close to children with chicken pox, which is caused by the same virus. Some infections are confined to the ear itself. Labyrinthitis is an inflammation of the inner ear, caused by bacterial or viral infection, which can make you feel dizzy, give you tinnitus (ringing in the ears), and can lead to sensorineural hearing loss, but usually only in one ear. Bacterial labyrinthitis often results in permanent hearing loss.

Head injuries :
Head injuries, especially those associated with a fractured skull, may produce profound and often permanent hearing loss. Even where there is no fracture, sensorineural hearing loss may occur, caused by damage to the central nervous system or the inner ear itself.

Noise :
Noise induced hearing loss (NIHL) is usually gradual in onset and can be prevented by wearing appropriate ear protection. However, sudden hearing loss may occur from exposure to excessively loud noises, for example from blast injuries (from a nearby explosion) or from firearms and fireworks, especially in enclosed spaces. This sudden hearing loss can range from total deafness in one or both ears to a relatively minor high-frequency loss. In these latter cases, the hearing may recover spontaneously in time.
Ear surgery:

Sensorineural hearing loss may occur after any surgical procedure on the ear, and the degree of risk depends on many factors. These include the nature of the procedure, the underlying disease and the skill of the surgeon. The hearing loss may occur immediately, in the few days following the operation, or even many years later. It is important to balance the potential benefits of surgery against the risks to hearing, and these issues should be discussed with the specialist.

Barotrauma:
Barotrauma may occur when the ear is exposed to sudden pressure changes, as in flying or diving. Most commonly, the middle ear is affected, causing a conductive hearing loss. It is rarer for the inner ear to be involved, but sensorineural hearing loss can occur if the membrane of the round window ruptures, causing a leakage of perilymph.

Immunological disorders:
Disorders affecting the immune system may have a role in causing Sudden Sensorineural Hearing Loss but the link is, at best, uncertain. There is a certain amount of debate over the diagnosis of the condition known as Autoimmune Inner Ear Disease (AIED), its relationship with SSHL, and potential treatments, but it probably accounts for less than one per cent of all cases of hearing loss. Sudden deafness has been known to occur in established autoimmune diseases such as rheumatoid arthritis and diabetes. However, there is no evidence that rheumatoid arthritis or diabetes *cause* sudden deafness; they are common conditions and it is likely that some people who have them also happen to develop idiopathic SSHL.

Ototoxicity:
SSHL is sometimes attributed to ototoxic drugs (drugs that may damage the inner ear) and rare cases have been reported following the use of gentamicin. However, the deafness related to ototoxicity is more often gradual than sudden, and is often preceded by tinnitus. Drugs that are known to cause permanent hearing loss are usually given only when no other alternative exists for treating a lifethreatening disease. If your doctor prescribes ototoxic drugs, you should discuss with them how this might affect your hearing.

Ménière's Disease:
Ménière's Disease affects about one in every two thousand adults in the UK population. It is a late-onset disease usually starting between the ages of 30-50 years. Ménière's Disease is characterized by severe attacks of vertigo combined with fluctuating deafness, tinnitus and a feeling of pressure in the affected ear. Attacks often start without warning and can lead to loss of confidence, anxiety and sometimes depression.

Other causes:
Less common causes of SSHL include an acoustic

neuroma, which is a benign tumour. The hearing loss associated with acoustic neuroma is usually gradual and often unnoticed, but may be sudden in as many as 10% of cases. Even rarer is the neurological condition of multiple sclerosis, in which 'plaques' of the disease may affect parts of the brainstem associated with hearing. The diagnosis of this condition is made by ABR and a magnetic resonance imaging (MRI) scan, and fortunately the hearing often recovers spontaneously.

Objective of this study is to report 16 cases with SHL.We take consideration of 5 parameters:

1. The initial size of hearing loss,
2. The beginning of treatment after first time of SHL appearance.
3. Patients age,
4. Audiogram type
5. Accompaniment of dizziness or / and tinnitus.

For our study we accept the Wilson definition that << SHL is definited by HL 30db at least, in 3 continuously frequencies, that is being onset in interval less than 3 days >>.

Material – Method :

Sixteen patients studied (men 8, woman 8, age between 31 and 85) at our clinic in 2003 - 2004 with HL onset less than 10 days. Full personal health record, clinical examination, audiographic and laboratory control as well as petrosal CT, where there was indication, are our standard methods to estimate such cases.

The following therapeutic scheme was given to all patients:

1. Bollus 1 solu – Medrol 500 mg I.V. at patient's admission.
2. 5 amp Loftyl in N/S 0, 9% 1000 cc for 6-8 h per day.
3. I.V. presolon (prednisolone) 25mg x 3 / per day.
4. Tabl Neurobion x 3

Also, other measurements , such as salt constriction, control of diseases (diabetes mellitus, immunodeficiencies, etc.) were taken in specific cases.

According to ALFORD we have three types of SHL : medium HL (30-55db) , severe HL (55-80db) , very severe or deafness (> 80db).

As for the start of treatment we give the following score : 0 : immediate therapy after HL , 3 : 3-5 days after hearing Loss , 4 : 5-10 day after hearing Loss , 5 : > 10 day after hearing Loss .

Audiogram estimation of the other ear was done at first (day of admission) and 10nth day.

Lab test include hTSH II , T3 , free – T4 , CRP, RA test , blood glucose , IgM and IgG for CMV and toxoplasmosis , ANA , etc.

At patient's clinical examination and recording we ask about possible dizziness, tinnitus, possible virus illness of upper respiratory system, drugs reception, other diseases, SHL at previous time, etc.

Results :

Ratio men to women: 1/1. Dizziness or vertigo was considering 5 patients at the admission time. Treatment initialization was immediately for 9 patients, between 3rd & 5th day for 4 and between 5th and 10th day for 3 patients. Previous virus infection was presented in 4 cases. Average of hearing loss was 56 db (1rst acoologic estimation). The 10 days absolute improvement (gain) in db (3rd acoumetric estimation) was 33 as an average for those 16 cases and 75 % in our group had hypacousia improvement more or equal to 30 %.

Discussion:

Having reviewed all the possible causes of SSHL, here are all treatments that can be used, depending on the initial diagnosis. We are also focusing on what research is being done into sudden sensorineural hearing loss and new methods that have been developed at this time, focusing on early diagnosis.

Sudden Sensorineural Hearing Loss where the cause is unknown

Although hearing returns to normal or near-normal in about 70% of these cases, 'failure to investigate patients will inevitably lead to a missed diagnosis and a missed opportunity for treatment. Hence, in all such cases, urgent investigation is required. Where cases of SSHL may be due to a virus or vascular disorder, a wide range of treatments have been recommended, but none have had convincingly beneficial results. However, some viral infections respond to anti-viral agents like acyclovir, and in the Ramsay Hunt syndrome short-term treatment with steroids and acyclovir is justified if started early enough, as it may lead to a reversal of the hearing loss to near normal thresholds. There is no evidence of cardiovascular disease in the majority of patients suffering from SSHL , but there are certain 'vascular' conditions which may occasionally cause sudden deafness. These include reduced blood flow in the cochlea and disorders of coagulation (clotting) of the blood. Research is being carried out on the potential value of carbogen inhalation in such cases, carbogen being a mixture of 95% oxygen and 5% carbon dioxide. So far no value has been proven.

Sudden Sensorineural Hearing Loss with a known cause

In many cases of Sudden Sensorineural Hearing Loss, there is no effective treatment for the hearing loss itself, but further loss may be reduced and occasionally the hearing may even be restored by preventive measures. For example, the adverse effects of ototoxic drugs may be halted and to some extent reversed if the patient stops taking that particular drug (in consultation with a medical advisor) and replaces it with a different agent. Similarly, the risk of noise-induced hearing loss can be greatly reduced by appropriate ear defenders, and the use of helmets undoubtedly reduces the risk of deafness in skull fractures due to head injuries. Finally, timely surgery for the 'dangerous' type of middle ear infection will prevent what is called suppurative labyrinthitis, which occurs if the infection spreads to the inner ear. When a perilymph leak (a leak of one of the inner ear fluids) is suspected, bedrest is advised for up to five days. If there is no improvement in the hearing after that period, surgical exploration of the ear is needed, with repair of the leak when possible. In cases of Ménière's Disease, most forms of medical treatment are aimed at controlling the crippling attacks of vertigo, while preserving the hearing if at all possible. In severe cases, if attacks persist despite medication, a Consultant may advise surgery. A number of different operations have been devised, some to reduce pressure in the inner ear, others to diminish or abolish the abnormal impulses passing from ear to brain.

WHAT RESEARCH IS BEING DONE INTO SUDDEN SENSORINEURAL HEARING LOSS? Much of the recent research on Sudden Sensorineural Hearing Loss has been targeted, not surprisingly, on those idiopathic cases in which no firm cause can be established.

Steroids and anti-viral agents:

As mentioned above, the anti-viral agent acyclovir is occasionally used to treat SSHL, on the assumption that some cases may have a viral origin. However, researchers are still investigating how effective acyclovir is as a treatment for hearing loss.

Reducing clotting in the artery supplying the cochlea: It is still unclear whether vascular factors play a part in Sudden Sensorineural Hearing Loss, but on the assumption that they have a role, many attempts have been made to reduce the risk of clotting in the very small artery supplying the cochlea. A study in Germany on patients with SSHL found that some of them had excesses in the blood of fibrinogen (which is involved in the process of clotting) and of the 'dangerous' type of cholesterol. The researchers then tried to find out whether partially removing these harmful substances – an established procedure in patients suffering from coronary artery disease – could produce better results than standard treatments of SSHL. The results showed small, but not significant, improvements, but the author concluded that this procedure could reasonably be used as an alternative to conventional therapy.

Establishing an immune disorder connection: Many authorities include immune disorders in their lists of causes of SSHL, although the hearing loss caused by autoimmune inner ear disease (AIED) is more commonly gradual than sudden. Researchers are working on developing a test which could determine whether an immune disorder has led to SSHL.

Technology:

In addition to research into the causes of Sudden Sensorineural Hearing Loss, there is a lot of work going on to improve hearing aids and cochlear implants. Research on hearing aids includes the development of digital technology, improving evaluation and fitting procedures and the development of more effective sound processing technologies to improve the perception of music and the human voice. Research into improving cochlear implants is advancing all the time. Areas of research include improving the quality of sound, improving tuning procedures, investigating the advantages of bilateral implants (implants in both ears) and techniques which can provide high-frequency hearing via an implant whilst allowing conventional amplification (e.g. via a hearing aid) to provide low frequency hearing.

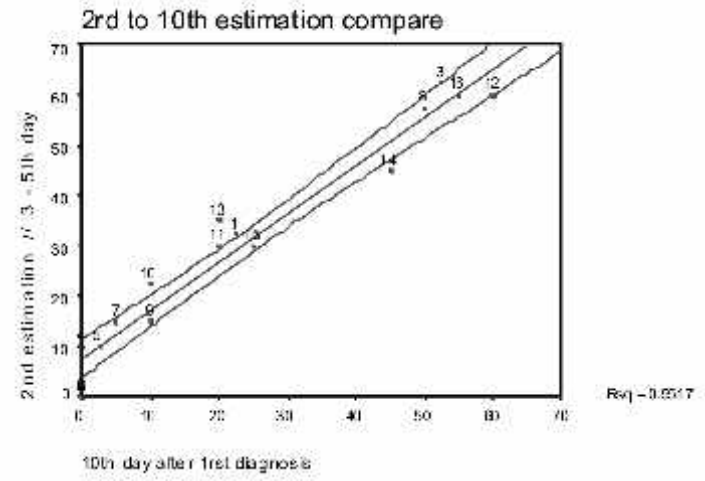
Ménière's Disease:

Researchers funded by Deafness Research UK are currently conducting a major project to improve understanding of the causes of Ménière's Disease. Many cases of Ménière's are sporadic but in 7% other family members are similarly affected. This, together with the predominantly Caucasian/Eurasian distribution, is strong evidence of a genetic predisposition. Preliminary genetic studies have pinpointed one area of the human genome which could be involved. Deafness Research UK has funded the initial molecular genetic study of this site. In due course other candidate genes will be examined. The identification of the Ménière's disease gene(s)

made possible by this funding will lead to a boost to the hypotheses as to the cellular pathways involved in the underlying cause. This should in turn help to direct the search for new therapeutic and even preventative strategies, though in the long-term. Identifying the gene(s) predisposing to Ménière's disease will bring two immediate clinical benefits; it will speed up early diagnosis, which can sometimes be difficult, and it will provide a predictive test for relatives or descendants of affected patients, especially in familial cases. Recently, new studies about Ménière's Disease reveal the usefulness of low frequency masking in early diagnosis. Specifically, this may lead to new instruments development that will further burst possibilities of early diagnosis in extended new fields.

Conclusions:

1. 40% of patients have usually mild or transient vertigo with nausea and vomiting or only dizziness.
2. Audiometry shows a unilateral sensorineural deafness in 80 %.Computed tomography of the temporal bone is indicated to avoid overlooking mastoiditis, primary cholesteatoma, acoustic neuroma and spondylosis deformans. Blood studies of value should include a complete blood count and serologic test for syphilis.
3. As for the day of treatment initiation, we see definite advantage in improvement when we start treatment under 3rd day (90 – 95% , t =5.592 , df=15 , p<0,01).Something that also lead to this conclusion is that second estimation (3-5 day) interpret 95,2% of definitive therapeutic results . When we start therapy later, improvement becomes in more slowly rates and deadlock is more usual.
4. Adding of Valtrex in therapeutic scheme within 2004 doesn't bring any positive results (2003 mean : 15.250 , sd : ± 20.188 , 2004 mean : 37.500 , sd : ± 18.097 , t = - 2.213 , df = 14 , p = 0.044).
5. Reliant on international bibliography, improvement of SHL without treatment ranges between 33 and 56%. Our improvement ratio is near piracetam's using as indicated from other studies.
6. It seems also that 2nd estimation results are critical (important predictive value) for definite total improvement (Pearson correlation = 0.976 , p< 0.01). That also indicates the value of early treatment.
7. Significant residual hearing loss concerns men-left ear (Pearson x2 = 14.186 , df =1 , p<0.01)



Summary :
SHL is a serious medical situation , which may lead to major permanent hearing loss if early treatment isn't applied where is indicated. In our two years study is revealed the importance of early treatment , especially when it is applied the first three days after the initial symptoms have been raised. In the discussion part of our paper , we review recent research on Sudden Sensorineural Hearing Loss and what treatments are going to be set in the future. It must be said that as only 15% of SHL cases have known cause we are focusing on treatments that deal with possible causes , such as virus infections and clotting or spasm in the artery supplying the cochlea.

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