HEARING LOSS IN CHILDREN

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Hearing loss in children can be present at birth because of an inherent defect or the child will develop a hearing loss subsequently. A baby's inner ear begins development at 20 days from fertilization, the external ear at 28 days, and the middle ear at 40 days. The cochlea is fully formed at 20 weeks. The sense of hearing is developed anatomically very early so any thing that happens to a baby during the first 20 weeks can lead to hearing loss. Whilst the ear is developing the other parts of the body are developing, and there may be a combination of different conditions, which one may have to look out for or consider. Some defects will not be apparent which we call a silent condition. Unless we go looking for these things, we may not know what else is wrong. So of the children who have hearing loss, 40 per cent of these children will have a defect which will lead to congenital hearing loss. It could be an infection or it may be a genetic defect that that child has developed. The genetic defect can lead to just problem of hearing in the children, or it may be to other problems and other organs that are developing at the same time as the ear.

40% of children will have a prenatal hearing loss as shown in Figure 1 and 13% will have a perinatal hearing loss as shown in Figure 2. These children will have hearing loss, either soon after birth, and may not be because of the defect during pregnancy or during genetic loss, but can be because the child was born prematurely. This may have led to an insult causing hearing loss. It may be because the child may have other abnormalities which do not lead to hearing loss directly, but either indirectly or the treatment of diseases can lead to hearing loss. Infections being one of them, so newborn babies developing an infection for which they need antibiotics can lead to it and these antibiotics have to be given in doses that can damage the hearing. One of the common drugs that are used can cause this.

There can sometimes be a combination. So a person may be born with a genetic defect that on its own does not lead to hearing loss. But if that person with that genetic defect is given a drug to treat that condition, the hearing loss may happen because they are more at risk of developing complications from the drug, such as hearing loss, than perhaps an ordinary person who does not have that genetic defect. So these are all very complex things but they are important from a medical legal point of view as well.

A small proportion, 7% of children will have

Causes of SNHL

Pre natal 40 %
- intra uterine infection
- genetic
  - isolated
  - part of a syndrome

Figure 1

Causes of SNHL

Perinatal 13 %
- pre maturity
- metabolic disorders
- infections
- ototoxic esp. Aminoglycosides
- noise

Figure 2

Causes of SNHL

Postnatal 7%
Meningitis
Other infections
Mumps and Measles
Trauma (physical esp. surgery and noise)
Ototoxic drugs
Neoplastic and metabolic condition

Figure 3

hearing loss secondary to acquired losses from a postnatal condition, see Figure 3. These acquired causes can mostly happen at any stage in life. For example meningitis. In my centre the commonest cause of acquired hearing loss is meningitis. Then there are other infections, such as mumps and measles, thankfully with the vaccination they are less, but nonetheless you still see them. Trauma is another cause of hearing loss and trauma can be head trauma or it can be surgical trauma. Sometimes children have to undergo certain procedures which may lead to hearing loss.

Drugs can lead to hearing loss. There are many such drugs that can cause hearing loss, and my largest population of children outside my cochlear implant work that I do, who have hearing loss are the children who are undergoing cancer treatment e.g. Cispaltin and Carboplatin. A lot of cancer medications, which are given in order to cure the cancer, can lead to hearing loss. And that is a very difficult decision for the parents, because they are given a choice between a combination of drugs which will lead to hearing loss, or a combination of drugs which will lead the child to become infertile. So when the parents ask me what I would wish for my child, I say 'I would rather my child be deaf, because we can do something about it, than be infertile'. Some oncologists may not give that advice is because they haven't seen how the science has progressed, particularly with cochlear implants. I think that I would rather go for a deaf child whose deafness we can perhaps optimise, than allow them to have no grandchildren.

Another condition is a disease which is spread from cats to pregnant mothers! Perhaps there may be an old grandmothers saying that pregnant women must not come in contact with cats yet that is now based on very good scientific evidence that we have. Unfortunately, there is no immunisation available for this condition

Mothers who are diabetic may also have certain metabolic changes that can lead to hearing loss in children. And there is also a question mark about certain anti epileptic drugs that epileptic mothers might be taking. And finally there are complications that can arise from syphilis and HIV. Usually all the mothers who have had infections and their babies are born, they will have hearing problems and they will also have vision problems. Often, although not always, the hearing loss is not profound at the time of diagnosis, so it may be mild or moderate hearing loss, but usually it progresses and becomes severe.

. A syndrome is a condition and it is a term used when there is a collection of abnormalities that occur together. So, for example, a child with a syndrome may have problems with eyes, with heart, with the ear. And they all come together. The commonest involved syndrome that most people would know of is Downs Syndrome. Syndrome hearing loss accounts for 30 per cent of hearing losses in children that we see. A lot of these conditions can be passed on from the child to their offspring when they become older, and I think it is only right that we actually identify that and educate the child and their parents, because this may be a first child that these parents have had and they want to know what are the chances of their next child having the same problem. So, it is important that we diagnose the syndrome and it is important that we also identify whether it is actually a genetic syndrome which can lead to other children being born with the same condition, or whether it is an environmental factor. A dominant condition is a term which is difficult for people to remember, but a dominant condition means that at least one of the parents has that condition. So if the parent has blue eyes their children may have blue eyes.

Some children may have abnormal outer ears, they may have little pits before the eyes, and they may have little pits or swellings in their neck. Some patients will have abnormal outer ears and they will have various abnormalities. Often they have bilateral ear abnormalities, they cannot be fitted with standard hearing aids. One syndrome is Ushers Type I Syndrome which is a very sad condition where the child will ultimately lose their vision as well as their hearing but because hearing loss is the first defect to be diagnosed, it can be overlooked. Overall there are 16 identified syndromes that can cause hearing loss.

In a study published 3 years ago of 106 children, we found that for the majority, we could not identify a cause, see Figure 4. We looked at whether the children who had hearing loss had other abnormalities. We found that again, a majority had balance problems. They had difficulty running, jumping, walking in the dark. From this study we developed a combined scoring for evaluation of both hearing loss issues and other complications which gave us a disability rating which helped in identifying the best choice of assessment technique. Should we assess these children in standard

CI Outcome Study-
Distribution of Aetiology

Hereditary syndromic	15%
Acquired –Perinatal	3.8%
Acquired-Interuterine	2.8%
Acquired-Meningitis	17.9%
Hereditary-Non syndromic	26.5%
Unknown causes	34%

Figure 4

continued on page 4

manner or should we do something different for these children who may have additional disabilities? They do need more detail, more prolonged assessment. We need to be more aware of their likes and dislikes, how they expressed themselves, how they communicate. And it is often very difficult to assess their hearing, as well, to see whether they are deaf enough to benefit from a cochlear implant, whether they are getting enough through their hearing aids.

Often these children have a lot of other professionals involved in their care, so we had to develop a special protocol for multi discipline assessment. So, we had a group of teachers, we had speech therapists and audiologists who were involved and a lot of people are involved in these children for cochlear implants. The people had to stay the same and they had to identify themselves in a certain way that the child consistently recognised. We have to work very patiently with these children for they are a most deserving section of our community.

(Dr Rajput acknowledged the following contributions to this work: Ms Alex Wheeler, Mr Ken Nischal, Everyone in C.I Dept @ GOSH)

The RNID, with the backing of Penelope Brock, a world renowned consultant paediatric oncologist at GOSH, is calling on the pharmaceutical industry to invest in otoprotectants (to counteract potential hearing loss), especially as the drug mentioned above, Cispaltin, is used to treat about a third of all childhood cancers — Ed. Comment.

FUND RAISING

As with all voluntary associations, we rely heavily on donations and gifts to carry out our work. Despite continual efforts by our Chairman, Lord Ashley, to raise funds, we need more in order that we may continue helping people to secure and enjoy the benefit of a cochlear implant.

We are now appealing for your help and ask all readers the following.

- Have you family, friends or colleagues whom you could approach for a donation?
- Would your employer consider making a donation?
- Have you any contacts with companies or Trusts who might make a donation?
- Do you have any knowledge of fundraising?

If you can help in any way please contact Richard Byrnes, Editor (see page 8 for details). Please would all readers try to help us help others.