



Factsheet – 15

THE CHANGING DEAFBLIND POPULATION

Since 1955, when Sense was first set up, we have learned a great deal more about the population of deafblind and dual-sensory impaired people living in the UK - although this knowledge is far from complete. As well as children and adults who have been born deafblind as a result of a range of conditions such as congenital rubella syndrome or CHARGE, around 4,000 people affected by Usher syndrome have been identified, as well as an estimated 250,000 older people whose sight and hearing is significantly affected.

In some cases, knowledge of the causes of deafblindness have led to changes in the deafblind population - there are now far fewer children affected by congenital rubella syndrome as a result of vaccination. In other cases, as with people with Usher syndrome, our knowledge of this population is more incomplete, so that it is not possible to identify if there have been any changes.

A slow dawning of knowledge

It is only recently that we have been able to develop our understanding of how parts of the deafblind population are changing. This is largely because deafblindness and dual-sensory-impairment (a combination of sight and hearing difficulties) have only been recognised for a fairly short time. The first deafblind school in the UK that we know of, was set up at the Homerton School for the Deaf in London in 1900, and by 1911 50 deafblind children had been identified in the UK.¹ We do not know what caused these children's deafblindness, and it seems likely that this figure hugely under-estimates the number of deafblind people in the UK at this time.

A number of other developments in the first half of this century - the formation of the National Deafblind Helpers' League (now Deafblind UK) in 1928; deafblind pioneer Helen Keller's Honorary Doctorate for her work with deafblind people in 1932; and the opening of a deafblind unit at RNIB Condover Hall School in 1951 - reflect a gradual rise in awareness of deafblindness. But our knowledge of the deafblind population over this period remains sketchy.

Changing causes of deafblindness amongst children

Congenital rubella syndrome

When Sense was established in 1955 (known at first as 'The Rubella Group'), rubella epidemics were a common occurrence and hundreds of babies were born

with congenital rubella syndrome (CRS). With CRS, a child's ears, eyes and heart are the most likely to be affected, although a number of other conditions can also occur:

- A child's **eyes** develop during the early stages of pregnancy. Some babies will be born with cataracts (cloudy lenses) in one or both eyes; others may have unusually small eyes, colouring of the retina or a condition called Nystagmus - where the eyes flicker from side to side. Other children may suffer from rarer conditions or find that their sight gets worse as they get older.
- Many children have hearing loss in one or both **ears**. This is because the inner ear, which links the ear to the brain, has been damaged. A child's hearing may also get worse over time.
- The **heart** is a complicated organ and many different problems can be caused by the virus. Rubella can also affect a child's **brain**, and difficulties can vary from mild to severe.

As a result of the introduction of the rubella vaccination for schoolgirls in 1970, and then the Measles, Mumps and Rubella (MMR) vaccination in 1988, the numbers of children affected by rubella have fallen dramatically. Between 1971 and 1979 there were 416 children born with severe disabilities resulting from congenital rubella syndrome. Then, between 1991 and 1996, following the introduction of the Measles, Mumps and Rubella (MMR) programme, fewer than 33 babies were born with CRS.

Recent research by Norman Brown from Sense, amongst others, has shown that adults who have been affected by congenital rubella syndrome, may develop additional problems as they move into adulthood. This may include further deterioration in sight and hearing, and behavioural problems are commonly reported. See also Sense's factsheet **About rubella**.

Deafblindness as part of multiple disability

Today, congenital rubella syndrome is no longer the most common cause of deafblindness. Increasingly, as well as a sensory impairment, children have other complex and multiple disabilities to cope with. David Brown, formerly Head of Sense's Family Centre at Ealing, London writes:

"In 1983, Chris Best from Sense published a survey of deafblind children in the UK.² This showed that within the previous two years, the number of young children with congenital rubella syndrome appeared to be declining whilst the number of children who had multi-sensory defects plus additional disabilities due to other causes was increasing.

Research by professionals in North America, Western Europe and Australia has confirmed that this change in population has continued. In 1991, Mike Collins reported on students within four states in New England identified as having deafblindness, and found congenital rubella syndrome to account for only 15% of the total, and this was expected to drop to about 5% by 1995.³ Marianne Riggio, surveying changing population in the whole New England region in 1992, produced similar results.⁴

Any discussion of the causes of these changes must be a matter of opinion. Considering the significant increase in children with multiple anomalies is not straightforward and requires educated guesses. From our experience it seems likely that a growing proportion of this group, including children born prematurely and those left with multiple disabilities following early severe infections, are now able to survive because of developments in medical knowledge and technology. In other words, we are now working with growing numbers of children who could not have survived much beyond birth or the trauma of severe infection had they been born ten or even five years ago.”⁵

Survey of 100 children at the Ealing Family Centre

In 1996, David Brown collected information about 100 children who were currently receiving services from the Sense Family Centre in Ealing - 51 boys and 49 girls with ages from four months to six and a half years. He writes:

“It is often very difficult to pinpoint the precise cause of a child’s disabilities, but parents were asked their opinion and this was often backed up by written medical information. The largest group of children (26) had no known cause for their disabilities, and 29 separate causes were mentioned for the other 74 children. Most numerically significant were prematurity (12 children), birth trauma/asphyxia (8 children), CHARGE Association (7 children), Cytomegalovirus infection (5 children), and Meningitis infection (5 children). There were 9 different named syndromes including congenital rubella syndrome (2 children), Cri du Chat, Cat Eye, Goldenhar, Noonan’s, and Pallister Killian, and seven different rare identified chromosome abnormalities (7 children).

Anomalies

“Twenty-five separate anomalies (or characteristics) were identified within the group. In addition to visual and hearing impairment these included severe hypertonica (54 children), epilepsy (45 children), severe feeding problems (43 children), heart defects (18 children), hydrocephalus (10 children), hernia (5 children), choanal atresia (4 children), and cleft palate (3 children). The child with the most identified anomalies (9) had CHARGE Association but is developing and learning at a more satisfactory rate than any other child in the study. Some of the most severely disabled children in the group have only 3 or 4 identified anomalies. Clearly the severity of each individual anomaly is a crucial factor in determining rate of development rather than the number of separate anomalies present, although severe hypertonica and epilepsy were very common features of the children who are showing extreme delay in their development. The majority of children in the study had 3 or more identified anomalies (58 children); 35 children had between 5 and 9 identified anomalies, whilst only 7 were reported as having vision and hearing difficulties with no other anomalies.

The implications of these Trends

“Within the population of children with a multi-sensory impairment the increase in the incidence and severity of multiple disability means that every aspect of living and learning becomes more challenging, both for the children themselves and for the families and professionals involved with them. The sheer number of different areas of professional expertise which need to be called upon means that this is more

complex than working with a population consisting of mostly children with congenital rubella syndrome”.⁶

Usher Syndrome

Usher syndrome - a genetic condition where people are born deaf or partially hearing, and then start to lose their sight in their teens or earlier due to Retinitis Pigmentosa - has only been recognised as a distinct condition since 1860. It was named after C.H. Usher, an ophthalmologist who published case studies in 1914. Sense started working with people with Usher in 1983, and our knowledge has grown enormously since then.

Three types of Usher syndrome have now been identified:

- People with **Usher 1** are born profoundly deaf and usually grow up as part of the deaf community. Typically their vision starts to change in the first and second decade of life, with tunnel vision and nightblindness often being the first symptoms. Babies who have Usher have poor balance.
- People with **Usher 2** are born partially hearing, are often hearing aid users, and will have probably grown up as part of the 'hearing' community. Again, vision often starts to deteriorate during the teenage years. Balance is unaffected.
- People with **Usher 3** may have normal hearing from birth, but develop Retinitis Pigmentosa in adolescence or later when hearing loss also occurs.

Since Sense started offering support to people with Usher the profile of people it helps has changed. At first, most of the referrals it received were from people with Usher 1 and their families. Now, about 70% of the referrals are from people with Usher 2 who face very different challenges.

Also, as a result of improved diagnosis and awareness amongst doctors, children with Usher syndrome are being identified much earlier. While it will certainly be a hard blow for families to hear this news, it does mean that planning for the child's future can be carried out much more effectively.

Older people

As more people live longer, the incidence of sight and hearing loss is growing. Over half of people over 60 have impaired vision. Hearing loss, though less common, affects one in 12 people in this age group.

No two older people with disabling loss of hearing and sight are the same. The effects depend on the conditions that caused the loss, the severity of the loss and how it happened. Most people have had hearing and sight in their younger days and may not have had time to adjust to one loss before the other occurs. Others have been deaf or hard of hearing for some time then lose their sight, while there are also people who have been partially sighted or blind all or most of their lives who then lose their hearing.

References

1. *Homerton 1900-1921* by Doreen Woodford. Available from: British Deaf History Publications, 288 Bedfont Lane, Feltham, Middlesex, TW14 9NU.
2. Best, C (1983) *The "New" Deaf-Blind?* British Journal of Visual Impairment 1, 2, 11-13.
3. Collins, M, Majors, M, and Riggio, M (1991) *New Deaf-Blind Population: Etiological Factors and Implications for the Future*. Proceedings of the 10th IAEDB International Conference, Orebro, Sweden.
4. Riggio, M (1992) *A Changing Population of Children and Youth with Deaf-Blindness: Reaction Paper in Proceedings of the National Conference on Deaf-Blindness*. Hilton/Perkins National Program; Boston.
5. Brown, D (1997) *Trends in the population of children with multi-sensory impairment* Talking Sense Volume 43 No. 2

See also Sense factsheets:

- About rubella
- About Usher syndrome
- Hearing and sight loss in older people

Where can I go for help?

If you:

- would like to find out more about deafblindness or the services for deafblind people in your area
- require information in alternative formats including braille, large print, audio or disk - or would like this factsheet to be translated into your first language -
please contact Sense's Information Team.

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