



# CHARGE Association

**CHARGE Association has only been recognised since 1979, so it is very hard to be precise about the number of people with the condition. Because of this, knowledge is still sketchy. David Brown explains how CHARGE Association is identified and details our knowledge of it to date.**

*David Brown  
Head of Family  
Centre, Ealing*

**C**HARGE Association was first noted by an American doctor in 1979, and the acronym "CHARGE" was first used in 1981. This acronym is used to describe a very varied group of people who exhibit at least four of the features prefixed by the letters of the acronym and including one or other of choanal atresia and/or coloboma.

### Identifying Characteristics

**C**oloboma is an eye deformity involving an absence of part of the eye. Coloboma of the iris (at the front of the eye) may limit the person's ability to adjust to bright light; coloboma of the retina (at the back of the eye) will create a blank area in the person's visual field. Sometimes the eyes may be small and in extreme cases missing altogether.

**H**Heart defects may be of various kinds. Sometimes these problems can resolve themselves over time but often emergency surgery is needed very soon after birth.

**A**Choanal atresia is a blockage of the passages at the back of the nose, and is one of the major criteria for diagnosis of CHARGE Association. The blockage may be on one side or both sides, and it may be formed of a membrane of skin or of bone. Again emergency surgery is often necessary immediately after birth to open these passages.

**R**etarded growth and development may become evident as the child matures; most of the people identified as having CHARGE Association are below the third percentile of physical growth norms. There are multiple factors leading to retarded growth including severe feeding difficulties, reflux, breathing problems, chest infections, and multiple surgical procedures with repeated and prolonged hospitalisation.

**G**enitalia anomalies, specifically the incomplete development or under-development of the external genitals is very common in males. Both males and females with CHARGE Association often experience hormonal problems.

**E**ar anomalies can affect the external ear (which may be unusually large or small or of an unusual shape), middle ear (bone malformations or chronic glue-ear infections), and/or the internal ear (especially high frequency hearing loss). The most common form of hearing loss found in people with CHARGE Association is mixed i.e. a conductive loss because of middle-ear problems combined with a sensori-neural loss because of problems with the cochlea. It seems likely that at least some people with CHARGE Association may have central auditory processing problems (difficulties processing auditory information in the brain) which are very difficult to detect and can be superimposed over the other hearing problems.

Apart from these critical features of the condition there are other anomalies which are often found in people with CHARGE Association, including:

- cleft lip and/or palate
- facial palsy
- kidney abnormalities
- malformations of the larynx, oesophagus and trachea
- abnormal tongue size
- delayed and abnormal dental development
- malformed or absent semi-circular canals which means that the balance sense will be affected or absent.

At the CHARGE Conference held in Sydney in 1996 a paediatrician talked about two other problems which may be common in young children with CHARGE Association. The first was sleep apnoea, periods of severely obstructed breathing, which are sufficiently severe to wake the child and to cause persistent sleep disorder. The second was hypoglycaemia or low blood sugar, which can result in periods when the child becomes sweaty, cold, difficult to arouse, or unusually irritable; this situation has been found to improve if a glucose feed is given

# CHANGING DEAFBLIND POPULATION



IN DEPTH ▼

to the child at the appropriate time.

In the literature about CHARGE Association, which is mostly medical, the letter "R" in the name has often been interpreted as standing for mental retardation. Recent literature suggests that delayed or retarded development is now coming to be seen as an outcome of the other combined anomalies rather than as an integral part of CHARGE Association itself. Evidence from families and involved professionals suggests that people with CHARGE Association cover the whole spectrum of intellectual ability from severe retardation through to normal or even high intelligence.

## Cause of the Condition

The exact cause of CHARGE Association is still unknown and research suggests a variety of factors which may induce the irregularities which are present in this condition. The majority of cases are thought to be sporadic with no obvious evidence of genetic involvement. There is some evidence in the literature of cases where certain characteristics of CHARGE have been inherited, and also some evidence to suggest chromosome deletion in some cases. References in the literature, and many parents of people with CHARGE Association, suggest the possibility of linking CHARGE with exposure to certain chemicals (pesticides, fungicides, and wood preservatives) but this has not been proven.

## Size of Population

Very little is known about the precise numbers of people with CHARGE Association anywhere in the world. Most of the people identified as having this condition are children, though there are some young adults in their mid twenties. Because of this very little is known about the life expectancy of people with CHARGE Association, nor about the likelihood or nature of late onset complications. One difficulty is that this is a comparatively recently identified condition; if people were born with CHARGE Association well before 1982 it is unlikely that they will be identified as having the condition in their adult years. Another possible explanation is that babies born with CHARGE Association generally died because medical science was unable to support them through the first immensely complex and difficult years of life; nowadays we have the technology to achieve this in most cases.

In the United Kingdom the CHARGE Association Family Support Group has recorded over a hundred people with CHARGE Association. The Sense Family

Centre in Ealing is currently working with 130 young children, eight of whom have CHARGE Association.



*Joshua Levey who has CHARGE Association and whose father, David, is the new CHARGE Development Officer*

## Development and Education

Most of the literature about CHARGE Association is medical and until about six or seven years ago it showed little understanding of the educational needs or potential for development of people with CHARGE Association. This has improved in more recent literature and, as explained above, with growing awareness of the importance of early educational intervention and of the need for appropriate educational approaches, it should become increasingly rare for "mental retardation" to be automatically ascribed to children with CHARGE Association. As with all children born with multiple disabilities it may take years to determine fully the extent and nature of each disability. I have worked with several young children with CHARGE Association who presented as having severe visual and hearing impairment but who, subsequently, made excellent use of residual vision and hearing after appropriate early intervention and a great deal of hard work by their families and local professionals. Many people with CHARGE Association received no input at any time from a specialist in deafblindness, but as CHARGE Association becomes better known in the field of deafblindness it is to be hoped that this situation will improve. I know of no identified sub-group within the

---

*Recent research suggests that delayed development is probably the outcome of other problems rather than part of CHARGE itself*

---

# CHANGING DEAFBLIND POPULATION



## *CHARGE Association continued*

population of people with multi-sensory impairment who have so many medical problems, of such complexity and severity, and with so many hidden or delayed difficulties, and yet no sub-group has shown such a consistent ability to rise triumphantly above these problems, admittedly with massive help from medical professionals, therapists, educationalists, and above all their families. People familiar with CHARGE Association often speak about iron willpower, great spirit, and a marked sense of humour being distinctive features of most people with CHARGE Association, though as far as I know this is the first time these particular aspects of the condition have appeared in the literature!

*Further information about CHARGE Association can be obtained from:*

*David Brown, Sense Family Centre, Ealing, 0181 991 0513*

*Sheila Draper, CHARGE Association Family Support Group, 0181 540 2142*

# CHARGE Association and Sense

*David Levey  
CHARGE  
Development  
Officer*

The Sense Weekend Away last September attracted a wonderful turnout from the CHARGE Family Support Group. This impressive turnout caught the eye of Rodney Clark, Chief Executive of Sense, who picked up on various requests from parents for more support. After consulting with David Brown, Head of the Family Centre in Ealing, Rodney then invited a small group of parents to a meeting at Sense to see if things could be taken further.

In attendance at this meeting were family members: Sheila Draper, Jackie Turner, Yvonne Arnold, Ian Kirkbride, David Levey and a representation from Sense covering a wide range of activities all of which were relevant to the CHARGE Family Support Group. From this meeting it was agreed to set up an Action Group to develop things further. Members of this group are: from CHARGE - Yvonne Arnold, Ian Kirkbride, and David Levey, from Sense - Rodney Clark, David Brown, Malcolm Matthews, Director of Policy and National Services, and Joff McGill, Voluntary Services Officer.

In addition to the Action Group, Sense have decided that initially they will fund a 6 month development project and have appointed a CHARGE Development Officer to

carry out the work for one day per week. Sense thought it best that this appointment was given to a CHARGE parent and offered the position to me which I have accepted. I welcome the opportunity to address some of the major concerns of the Family Group and am also looking forward to deepening our relationship with Sense, who have already given us tremendous support in the past and clearly wish to carry on their good work into the future.

In more detail my job specification is as follows:

- Produce a first information leaflet
- Establish a database
- Liaise with Family Support Group
- Service the Action Group
- Support the preparatory work for medical involvement
- Liaise with David Brown and the Ealing Family Centre on FEAS (The Sense Family and Education Advisory Service) involvement.

Anybody wishing to send or receive information on CHARGE Association can contact me at Sense Head Office.

**David Levey**  
CHARGE Development Officer