Congenital cytomegalovirus and deafness

We use the term 'deaf' to refer to all types of hearing loss from mild to profound. This includes deafness in one ear or temporary hearing loss such as glue ear.
We use the term 'parent' to refer to all parents and carers of children.

Introduction

Congenital cytomegalovirus (cCMV) is one of the most common causes of permanent deafness in children after genetic (inherited) causes.[1] cCMV can cause deafness in children who are otherwise well.

In the past, information has focused on more severely affected children, who are often ill at birth. This factsheet has information relevant to all children with cCMV, including those who are not as severely affected.

This factsheet may be helpful for:

- families with children and young people affected by deafness that they believe may be connected to cCMV
- families with children and young people affected by cCMV
- professionals working with these families.

What is cytomegalovirus (CMV)?

CMV belongs to the herpes virus family. It is very common, affecting people all over the world. Despite there being very little public awareness, CMV is more common than many other conditions such as Down's syndrome, spina bifida, toxoplasmosis and cystic fibrosis. Many of us will have had CMV by the time we are 40 years old. Sometimes CMV can cause cold or flu-like symptoms but often the infection has no symptoms so we may not know we have had it.

Once infected by CMV, the virus usually stays in an inactive form in the body (often for life). If your immune system becomes weakened the virus can reactivate but does not cause as much harm as the primary infection.

The CMV infection can cause illness in anyone with a weak immune system, such as an unborn baby. CMV infection in an unborn baby is called congenital CMV (congenital means present from birth). Congenital CMV (cCMV) causes about 10% to 20% of permanent deafness in children in the UK and is the leading cause of non-hereditary deafness.[1] There is currently no vaccine to prevent cCMV, but researchers are working on one.

If you have the CMV infection as a child or adult, this is called 'acquired CMV'. If a baby has a CMV infection in the first month of life, this is called 'perinatal CMV'. Acquired and perinatal CMV are not known to cause childhood deafness.

How is CMV spread?

From one person to another through close contact with bodily fluids such as urine, saliva, blood, tears, breast milk, semen, vaginal fluids and faeces (poo). You cannot catch CMV from being in the same room as a person who has the infection. CMV can also be spread through transplants and transfusions through a donor with either an active or past infection. Patients receiving transplants or transfusions are more susceptible because of their weakened immune systems.

A pregnant mother infected with the CMV virus for the first time can transmit it across the placenta to her unborn baby. The majority of these babies will not have any problems.

Breastfeeding advice

Breastfeeding is still recommended for children with cCMV and well babies with possible or confirmed deafness. CMV infection passed to a newborn baby from breast milk (called 'perinatal CMV') is not known to cause childhood deafness.

What happens during CMV infection?

There are three different ways in which CMV can occur.

Primary infection: when someone is infected for the first time and has no pre-existing immunity. Most times it remains inactive in the body.
The first time we come into contact with CMV our immune system makes antibodies (a special protein to help fight the virus). CMV becomes present in body fluids and can be passed from person to person. A child with cCMV may 'shed' i.e. pass or 'excrete' CMV in their urine for a few years. The length of viral shedding may last for a few months, but it can last up until eight years of age, and possibly longer. The virus will then lie 'dormant' (inactive) in the body, just like the chickenpox virus does.

Reinfection: when someone becomes infected again but with a different strain of CMV.

Reactivation: once infected the virus remains dormant. The chances of it reactivating are low i.e. 0.01% or 1 in 10,000.[2]
The virus may become active again at any time, but this will not be noticeable because it doesn't cause illness. However, CMV will become present in body fluids again, meaning the virus can be passed on to another person.

How common is cCMV?

cCMV is the most common infection passed from a mother to an unborn baby. If a mother becomes infected with CMV for the first time when pregnant, the virus is passed to the unborn baby in about 33% of cases.[3]
cCMV affects about 1 baby in every 150 born.[1] While the majority of these babies will not have any symptoms, around 1 in 1,000 will have a permanent disability.[2]

Symptoms of cCMV at birth

Some children with cCMV will have symptoms at birth (symptomatic cCMV) whereas others are born without symptoms (asymptomatic cCMV). Sensori-neural deafness (permanent deafness caused by damage to the hair cells within the cochlea or the hearing nerve (or both)) is one of the long term neurological symptoms caused by cCMV. The deafness does not always become apparent until later on in early childhood. Other effects of cCMV can include cerebral palsy, visual impairment and developmental disabilities.

Children with symptomatic cCMV generally have greater health problems than children with asymptomatic cCMV but the effects can vary greatly. The severity of symptoms at birth is not necessarily an indication of how severe the effects will be later in life.

Symptomatic cCMV

Only 10% of babies with cCMV are born with symptoms or signs of the infection.[1] They may include one, or a combination, of the following:

- low birth weight
- small head
- little red spots under the skin
- big liver
- big spleen
- yellow skin and eyes
- fits (seizures)
- tiny patches of calcium in the brain
- eye problems
- sensori-neural deafness.

Symptomatic cCMV can have long-term effects on a child's development and cause a range of health problems. These could include physical and motor impairment, sensory problems, developmental and learning delay, attention deficit hyperactivity disorder (ADHD) and autism. About 30% of children with symptomatic cCMV will have a degree of deafness.[2]
For more information on ADHD visit **www.adhdfoundation.org.uk.** For more information on autism visit **www.autism.org.uk**.

Asymptomatic cCMV

About 90% of babies with cCMV are born without any symptoms or signs of the virus.[1, 2] This is called asymptomatic infection. Most children with cCMV are never diagnosed and so little is known about their progress.

Many children with asymptomatic cCMV do not appear to be affected, however, one in ten of these children will have or develop some degree of deafness.[1, 2] The hearing loss may be unilateral, bilateral, fluctuating or progressive. It is rare for these children to go on to have developmental or learning difficulties.

How is cCMV infection diagnosed?

Sometimes there are signs of cCMV before a baby is born, for example on the ultrasound scan, and tests for the infection may be offered to the mother.

After birth, cCMV may be suspected when a child has sensori-neural hearing loss, with or without other symptoms, or if they have other symptoms of cCMV with normal hearing. The following tests may be offered to provide evidence of cCMV infection.

Detecting the virus itself

Testing a sample of urine, saliva or blood within the first three weeks of a child's life is the standard way of confirming cCMV. Tests using saliva samples should be taken one hour after the baby has been breastfed. If a sample collected from a child older than three weeks is positive for CMV, it will not be clear if the child became infected before or after birth. Newborn blood spot testing is advised to help find out the timing of the infection.

The newborn blood spot test (commonly called the Guthrie card and dried blood spot (DBS))

All newborn babies in the UK are offered a newborn blood spot test to check for some rare illnesses (such as an underactive thyroid gland). The test involves taking four drops of blood and placing them on a special card when the baby is five to seven days old.

A positive blood spot test for CMV helps diagnose cCMV infection because it has been taken within the first three weeks of the baby's life. However, failing to find the CMV virus does not completely rule it out because CMV can't always be detected in the bloodspot from a newborn baby.

Antibody tests

A blood test to identify the CMV IgG antibody is the most reliable way of testing someone over a year old. If the antibody is not found, it suggests that the child has not had CMV. If the child's mother does not have the antibody, this suggests that she has not had the virus and therefore, that her child has not had CMV.

A positive antibody test only shows that the child has come into contact with CMV at some time in their life. It cannot tell us whether this happened before birth (causing cCMV) or afterwards, so examination of the newborn blood spot test (Guthrie card) is advised.

The age of the child and CMV tests

- Urine and saliva tests are more helpful than antibody tests for children under a year of age. This is because, if a mother has had CMV, she passes IgG antibodies to her unborn baby through the placenta to protect him or her. So a positive test for the CMV antibody in a baby aged up to one year of age may reflect the mother's exposure to CMV, rather than the baby's.
- IgG antibody tests are more helpful than urine or saliva tests in children over one year of age. This is because virus excretion (shedding) due to cCMV infection usually stops sometime during ages one to four years. A negative urine test does not help exclude cCMV.

When might deafness happen?

Half of all children with deafness caused by cCMV have a degree of deafness at birth and this may be picked up following newborn hearing screening.[1] The other half go on to develop deafness after birth and will have had no problems identified on their newborn hearing screen.

Most deafness caused by cCMV develops during the first three years of life and so may affect speech and language development. Some children born with cCMV may develop deafness later in childhood.[4] It is important that children with cCMV have regular hearing tests so that any changes in hearing are picked up early and help is offered quickly.

What type of deafness does cCMV cause?

cCMV causes sensori-neural deafness, which affects the cochlea. This is the organ in the inner ear that changes sound vibrations into nerve impulses to be sent to the brain – or the hearing nerve from the inner ear to the brain. This type of deafness is permanent and can affect one or both ears. Half of the children whose deafness is caused by cCMV will have progressive or late onset deafness (becoming worse over time).[5]

cCMV may cause unilateral deafness (deafness in one ear). This is more common in children with asymptomatic cCMV, and some may develop deafness in the other ear later. Sometimes a child's hearing may go up and down (fluctuate). Research is being carried out to try to understand how CMV affects hearing and how this damage might be prevented.

cCMV may also affect the balance organs in a child's inner ear, their hearing nerve, or more rarely their ability to interpret sound (auditory processing disorder (APD)). Children with cCMV may also experience glue ear, which causes temporary conductive deafness. For further information see our resource *Glue Ear: A guide for parents* at **www.ndcs.org.uk/glueear**. There are lots of documents on this site. To get you started, we recommend that you look first at the most relevant guidance leaflet and appeal forms.

Why find out if cCMV has caused the deafness?

It is important to diagnose cCMV soon after a child is found to have deafness because of the higher than usual risk of the deafness being progressive or of unilateral deafness progressing to deafness in both ears. Parents and doctors will know to monitor the child's hearing, vision and balance carefully, and assess their progress so that any problems are dealt with early on.

Finding out that a child is deaf because of cCMV makes other causes of deafness such as inherited (genetic) causes less likely. This means it is less likely that any future children in the family will be deaf.

Treatment for cCMV

Babies less than four weeks old showing signs of symptomatic cCMV and babies with cCMV who have been identified as deaf, may be offered one of two antiviral drugs to stop the virus from multiplying. Ganciclovir is injected into a vein twice a day for about six weeks, and valganciclovir is a liquid taken orally. One research trial so far has shown that ganciclovir may improve hearing or stop deafness from getting worse.[2] This treatment may have other benefits too but not enough children have been studied for us to know for sure.

The baby will need to be closely monitored for side effects of ganciclovir, the most common being a reduction in platelets (blood count) and/or white blood cells, which can weaken the immune system and kidney function, and cause anaemia. There may be unexpected long-term effects of treatment, especially on growth and development, and this issue is currently being studied.[2]

Valganciclovir is also being used to treat children and adults who contract CMV during transplant surgery. Research trials are taking place to find out more about the benefits of valganciclovir medicine for newborn babies with symptoms of cCMV.

Audiological care

A child may not have noticeable problems with their hearing initially, especially if the deafness is affecting one ear. It is recommended that all children with cCMV should have hearing tests regularly in early childhood to detect deafness. For example, every three to six months in the first year, every six to nine months until age three years, and then every year until they are at least six years old. If the child is not likely to report a change in hearing themselves or parents or professionals are concerned, these tests should take place up to secondary school age. The family and child should be informed of the very small possibility of the onset of deafness in teenage years, so they can get help quickly if there are any concerns about their child's hearing.

cCMV is quite a common condition but there is a very small chance a child's deafness may have been caused by something else. Children with cCMV and sensori-neural hearing loss should be offered tests and investigations to look for another cause. For further information see our resource *Understanding Your Child's Hearing Tests at* **www.ndcs.org.uk/hearingtests**.

Knowing about a child's deafness means that help and support can be offered straight away. Early action is very important. A team of people who work within educational, medical, social and voluntary organisations can then support the deaf child and their family. The team may include a Teacher of the Deaf, an audiologist, a speech and language therapist and a doctor who specialises in deafness in children.

Some children with profound hearing loss who do not benefit from conventional hearing aids may benefit from cochlear implants. See our resource *Cochlear Implants: A guide for families* at **www.ndcs.org.uk/cochlearimplants**.

Balance problems are thought to be common in children with cCMV and vestibular function assessment (balance function testing) should be considered for children diagnosed with cCMV and deafness. Safety advice and specialised physiotherapy may be helpful for children with balance problems (known as 'vestibular dysfunction') due to cCMV. For further information see our resource *Balance and Balance Disorders* at **www.ndcs.org.uk/balance**.

Precautions and prevention

CMV infection is common in toddlers and young children. Women of childbearing age, pregnant women and people with low immunity may be able to avoid CMV infection through good hygiene. This is possible through:

- wearing gloves while changing nappies, and washing hands with soap and water after changing a nappy or coming into contact with a child's saliva
- not sharing food, cutlery, drinks, dummies, spoons, cups or toothbrushes with young children
- not kissing young children on the mouth. Kiss them instead on the top of the head and give them a big, long hug
- washing toys often with soapy water and then rinsing them
- washing hands before preparing food
- using condoms even after conception.

Information for nursery staff

It is important to realise that excretion of CMV in body fluids is common in many young children attending day care nurseries and schools, many of whom will have no symptoms or signs of infection. Staff who have had CMV infection in the past may also excrete the virus. Children with cCMV should not be isolated from other children or from staff. Instead, it is recommended that staff take the precautions listed above.

Future pregnancies

There is little information about the risk of having a second baby with cCMV and while this risk is extremely small, parents who already have a baby with cCMV are advised to wait a year before having another baby.

Other help and support

Some families with children affected by cCMV may be able to get Disability Living Allowance. Information is available at **www.ndcs.org.uk/dla**.

It is important for families affected by cCMV to know that they are not alone. Families and their friends, teachers, and health professionals can get information from CMV Action. A parent-to-parent support network has also been set up.

For more information contact:
CMV Action
0808 802 0030
info@cmvaction.org.uk
www.cmvaction.org.uk

Families with children with hearing and sight problems can get support from:
Sense
0300 330 9250 020 7520 0999
0300 330 9252 (textphone)
info@sense.org.uk
www.sense.org.uk

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4. Dahle, AJ (2000). Longitudinal investigation of hearing disorders in children with congenital cytomegalovirus. J Am Acad Audiol. 11, 283-290.
5. Fowler, KB. (1999). Newborn hearing screening: will children with hearing loss due to congenital cytomegalovirus infection be missed? J Paediatrics. 135 (1), 60-64.

Useful National Deaf Children's Society resources

Glue Ear: A guide for parents
Understanding Your Child's Hearing Tests.
Cochlear Implants: A guide for families
Balance and Balance Disorders

Available at **www.ndcs.org.uk/publications**.

Recommended reading

Griffiths, P. D. (2012). *The Stealth Virus*. CreateSpace Independent Publishing Platform.

This information can be requested in large print or as a text file.
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For resource references or to give us your feedback email **informationteam@ndcs.org.uk**.

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