

Guidelines on Hearing assessment and surveillance of children diagnosed with congenital Cytomegalovirus

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Aim: To set out a protocol for hearing assessment and surveillance of children referred following diagnosis of congenital Cytomegalovirus (cCMV)

Objectives

- To identify the referral pathway into Audiology for hearing assessment following diagnosis of cCMV
- To define hearing assessment procedures for these children
- Set out management guidelines when a hearing loss is diagnosed.
- Define surveillance procedures when initial assessments are satisfactory.

Introduction

Cytomegalovirus (CMV) is a common virus that is part of the herpes family of viruses. Congenital CMV if contracted in the womb can cause significant harm to the foetus, including permanent and progressive sensorineural hearing loss, visual impairment and neurological damage. It accounts for approximately 25% of permanent hearing loss in children under the age of 4 years¹. It is estimated that 1-2 babies in every 200 will be born with congenital CMV in the UK⁴ although not all of these children will be symptomatic.

Referral Pathway into Hearing and Balance

Referrals with a diagnosis of cCMV need to be triaged as urgent. If a hearing loss is detected, this will guide medical management of cCMV.

- **Referral into Audiology:** Referral to audiology is typically via Paediatrics following diagnosis of CMV/cCMV. Babies with confirmed congenital cytomegalovirus (cCMV) are referred directly to Audiology and do not undergo the Newborn hearing screen. This exclusion is because the risk of hearing loss is very high and the window of opportunity for treatment is short. These babies should be referred by the paediatrician to audiology for an early hearing assessment. The urgency is related to the short window of opportunity for anti-viral treatment.
- **Patients already under Audiology service:** In Audiology, children identified with a significant hearing loss following diagnostic assessment at birth are referred for Aetiological investigations into the cause of the hearing loss. Testing for the cCMV virus forms part of these investigations. Should the test be positive for cCMV, that information is conveyed back to Audiology as there is a risk of the hearing loss being progressive in the first few years of life¹. The cCMV protocol for assessment/review is then followed.

Nature of Hearing Loss due to congenital CMV

The association between cCMV infection and sensorineural hearing Loss (SNHL) has been known for a while, although the mechanism by which the virus causes hearing impairment in some children and not others is still not fully understood today. Approximately 30-50% of children with clinically apparent disease (symptomatic) and 8-12% of children without clinically apparent (asymptomatic) cCMV infection will develop SNHL. Hearing loss due to cCMV infection does not have a characteristic audiometric configuration and is variable in the severity of loss. Unilateral and bilateral hearing losses may occur in these children, with loss varying from unilateral high frequency losses (4-8 k Hz frequencies only) to profound bilateral loss. Progression and fluctuation of hearing loss have been observed with 50% of SNHL due to cCMV infection being of late onset loss. The fact that the infection can only be confirmed in the new born period has made it difficult to estimate the proportion of SNHL that is attributable to cCMV infection in childhood populations².

Assessment procedure

Ear-specific and frequency-specific information is needed as unilateral and partial hearing loss can occur. Children can also have complex developmental problems following cCMV. As the presence of a hearing loss can influence medical treatment decisions, outcomes of assessment must be communicated to the medical team responsible for the child as soon as possible.

- BSA guidance⁷ is that in babies (under the age where behavioural testing can be completed) ABR testing should be used and thresholds assessed down to 20 dB e HL at 1 and 4 kHz in both ears. This is irrespective of whether the baby has been screened and irrespective of the screen result.
- Where the patient is developmentally able for behavioural testing, age appropriate behavioural tests are carried out to test both ears across the speech frequencies. Patient continues to be seen at Hearing and Balance clinic till this is achieved due to the time required to complete these tests.
- If OAEs are present following initial ABR and behavioural tests, OAEs can be used to monitor cochlear function going forward. The child can be referred across to CDC once ear specific and frequency specific testing down to satisfactory levels is achieved.
- Due to the high risk of later onset of hearing loss, further hearing assessment is recommended every 3-6 months up to age 1 year, every 6 months up to age 3 and annually up to age 6 as a minimum. Parents can also refer themselves back to Audiology if there is concern regarding hearing following discharge; please inform parents about this on discharge verbally and on discharge letter.
- Please set up a journal alert on Audit base to identify patients with cCMV for routine review and to facilitate self-referral back to the clinic.

Management of hearing loss

Should a sensorineural hearing loss be diagnosed in one or both ears, management will be in line with other causes for a hearing loss.

References

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6. NDCS booklet 'Congenital cytomegalovirus and deafness' www.ndcs.org.uk
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