Guidelines for Investigation of Congenital Infections in Babies and Children with Permanent Childhood Hearing Loss

2018
INTRODUCTION

The aim of these guidelines is to provide an evidence based approach to the investigation of congenital infection as a cause of permanent childhood hearing loss

Section 1: Congenital Cytomegalovirus
Section 2: Rubella, Toxoplasma, Syphilis

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Section 1: Congenital; Cytomegalovirus

NBHSW Medical Carepathway - Investigation for Cytomegalovirus Infection*

Child with SNHL

Test maternal CMV IgG

Over 3/52 but Under 1yr

Positive

Over 1yr

Child CMV IgG

Positive

Dried Blood Spot Card CMV PCR**

CMV PCR Negative

CMV PCR Positive

CMV PCR Negative

1xCMV PCR Negative

Congenital CMV unlikely

CMV PCR Positive

1xCMV PCR

Urine CMV PCR

CMV PCR Positive

Congenital CMV

CMV PCR Negative

Congenital CMV Excluded

NBHSW Medical Carepathway - Investigation for Cytomegalovirus Infection*
NBHSW Medical Carepathway-Investigation for Cytomegalovirus Infection* cont.

*This pathway is for the investigation of cCMV infection in immunocompetent children.

** Written permission from parents is required to authorise release of Dried Blood Spot Card for CMV PCR. Enquiries should be directed to the appropriate Regional Newborn Screening Laboratory.

This pathway has been developed with Consultant Virologists Dr Nicky Price and Dr Rachel Jones, Public Health Wales and Dr Jennifer Evans Consultant Paediatrician Cardiff and Vale University Health Board.

Acknowledgement: Dr Simone Walter: Flowchart for investigating CMV as a cause of sensorineural hearing impairment.


Reference: Is saliva as reliable as urine for detection of cytomegalovirus DNA for neonatal screening of congenital CMV infection?
**Section 2: Rubella, Toxoplasma, Syphilis**

### Testing for Congenital Infections in Older Children (>3 months) Presenting with Sensorineural Hearing Loss*

<table>
<thead>
<tr>
<th>CMV</th>
<th>Rubella</th>
<th>Toxoplasma</th>
<th>Syphilis</th>
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<tbody>
<tr>
<td><strong>Test if:</strong></td>
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<tr>
<td>Mother or child from abroad</td>
<td>Chorioretinitis</td>
<td>Bone abnormalities (frontal bossing, saddle nose, sabre shins, shortened maxilla)</td>
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<tr>
<td>Mother travelled abroad during pregnancy</td>
<td>Features of congenital toxoplasma</td>
<td>Teeth Abnormalities (Hutchinson’s incisors, mulberry molars) (see Fiumara and Lessel 1983)</td>
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<tr>
<td>Mother didn’t receive 2MMRs prior to pregnancy</td>
<td>Hydrocephalus, Intracranial calcifications, Microcephaly, Microphthalmia, Seizures, Hepatosplenomegaly, IUGR, Jaundice, Purpura</td>
<td>Interstitial keratitis</td>
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<td>Signs of Congenital Rubella Syndrome (2 from A or 1 from A + 1 from B)</td>
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<td>More usual as a late presentation, after 2 years of age. Prior to this age, test if suspicious of congenital syphilis.</td>
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<td><strong>Group A:</strong> Cataracts, Congenital Glaucoma, Congenital Heart Disease, Hearing Loss, Pigmentary Retinopathy</td>
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<td><strong>Group B:</strong> Purpura, Splenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent Bone Density, Jaundice</td>
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**Test maternal IgG**

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<th>Positive</th>
<th>Negative</th>
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<tr>
<td>Test child IgG</td>
<td>Exclude</td>
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<tr>
<td>Positive</td>
<td>Negative</td>
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<td></td>
<td>Exclude</td>
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<tr>
<td>Ask if child received MMR Seek further advice (?IgM tests, ?booking blood, ?serial IgG/avidity tests)</td>
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**Test maternal IgG**

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**Maternal syphilis antibodies**

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*Please note this testing is only for immunocompetent children/adults*

Version 2/2018
# Testing for Congenital Infections in Children under 3 months of Age with Sensorineural Hearing Loss*

<table>
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<tr>
<th>CMV</th>
<th>Rubella</th>
<th>Toxoplasma</th>
<th>Syphilis/HSV</th>
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</table>
| Test if:  
• Mother or child from abroad  
• Mother travelled abroad during pregnancy  
• Mother didn’t receive 2MMRs prior to pregnancy  
• Signs of Congenital Rubella Syndrome (2 from A or 1 from A + 1 from B)  
• **Group A:** Cataracts, Congenital Glaucoma, Congenital Heart Disease, Hearing Loss, Pigmentary Retinopathy  
• **Group B:** Purpura, Splenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent bone density, Jaundice  
| Test current maternal IgG  
Positive ➔ Negative  
Excluding  
Test in parallel with booking sample for IgM/IgG and discuss with Consultant Virologist  
If unable to exclude maternal primary infection then test serum from baby for IgM (a negative result will exclude Congenital Rubella at this age)  
| Test current maternal IgG  
Positive ➔ Negative  
Excluding  
Send to reference laboratory with booking sample for IgM/IgG  
If unable to exclude maternal primary infection then seek specialist advice and send samples from the baby (serum, EDTA, CSF)  
| Test if:  
• Known maternal syphilis infection (should be part of birth plan if UK-born)  
(See ASW infections and Rashes in Pregnancy 2016)  
• Features of congenital Syphilis Rash  
Haemorrhagic rhinitis  
Generalised Lymphadenopathy  
Hepatosplenomegaly  
Skeletal Abnormalities  
(See BASHH UK Guidelines on Management of Syphilis 2015 for testing strategy)  

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*Please note this testing is only for immunocompetent children/adults*

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Version 2/2018

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See existing pathway
References

- Wales Neonatal Network Guidelines on Congenital Infections: diagnosis and management by Morris et al
- British Paediatric Surveillance Unit (BPSU) report 2008-2009: 17 rubella cases over ten years
- The stigmata of late congenital syphilis: an analysis of 100 patients. Fiumara and Lessel, Sex Transm Dis 1983; 10: 126
- Antenatal Screening Wales Infections and Rashes in Pregnancy
- BASHH UK National Guidelines on Management of Syphilis 2015