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

2017
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

Contributors:
Dr Nicola Price, Consultant Virologist Public Health Wales.

Dr Amanda Roberts, Programme Coordinator Newborn Hearing Screening Wales.

Dr Jennifer Evans, Consultant Paediatrician in Communicable and Infectious Diseases, Cardiff and Vale University Health Board

Professor Edward Guy, Toxoplasma Reference Unit Swansea, Public Health Wales
Section 1: Congenital; Cytomegalovirus

NBHSW Medical Carepathway-Investigation for Cytomegalovirus Infection*

Child with SNHL

Test maternal CMV IgG

Negative → cCMV Excluded

Positive

Under 3/52

Over 3/52 but Under 1 yr

Over 1 yr

Urine CMV PCR

Urine CMV PCR

Child CMV IgG

Negative → cCMV Excluded

Positive

CMV PCR

2xCMV PCR

2x CMV PCR

CMV PCR

Positive

Negative

Negative

Positive

Dried Blood Spot Card CMV PCR**

CMV PCR Positive

CMV PCR Negative

Congenital CMV

c CMV Excluded

Congenital CMV highly unlikely

Congenital CMV

Congenital CMV unlikely

Final Version

15.07.13

Version 1/2017
NBHSW Medical Carepathway-Investigation for Cytomegalovirus Infection* cont.

*This pathway is for the investigation of CMV 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.

# Section 2: Rubella, Toxoplasma, Syphilis

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

<table>
<thead>
<tr>
<th>CMV</th>
<th>Rubella</th>
<th>Toxoplasma</th>
<th>Syphilis/HSV</th>
</tr>
</thead>
</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, Spleenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent bone density, Jaundice | Test current maternal IgG  
- Positive  
  - 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)  
- Negative  
  - Exclude | Test current maternal IgG  
- Positive  
  - 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)  
- Negative  
  - Exclude | Test Syphilis 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) |

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

---

See existing pathway  

---

Version 1/2017
# 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>
</tr>
</thead>
<tbody>
<tr>
<td><strong>See existing pathway</strong>&lt;br&gt;<a href="http://www.wales.nhs.uk/sitesplus/documents/980/NBHSHSW%20Medical%20Carepathway%20-%20flow%20chartfinalJuly2013.pdf">http://www.wales.nhs.uk/sitesplus/documents/980/NBHSHSW%20Medical%20Carepathway%20-%20flow%20chartfinalJuly2013.pdf</a></td>
<td><strong>Test if:</strong>&lt;br&gt;- Mother or child from abroad&lt;br&gt;- Mother travelled abroad during pregnancy&lt;br&gt;- Mother didn’t receive 2MMRs prior to pregnancy&lt;br&gt;- Signs of Congenital Rubella Syndrome (2 from A or 1 from A + 1 from B)&lt;br&gt;   - <strong>Group A:</strong> Cataracts, Congenital Glaucoma, Congenital Heart Disease, Hearing Loss, Pigmentary Retinopathy&lt;br&gt;   - <strong>Group B:</strong> Purpura, Splenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent Bone Density, Jaundice</td>
<td><strong>Test if:</strong>&lt;br&gt;- Chorioretinitis&lt;br&gt;- Features of congenital toxoplasma Hydrocephalus, Intracranial calcifications, Microcephaly, Microphthalmia, Seizures, Hepatosplenomegaly, IUGR, Jaundice, Purpura</td>
<td><strong>Test if:</strong>&lt;br&gt;- Bone abnormalities (frontal bossing, saddle nose, sabre shins, shortened maxilla)&lt;br&gt;- Teeth Abnormalities (Hutchinson’s incisors, mulberry molars) (see Fiumara and Lessel 1983)&lt;br&gt;- Interstitial keratitis</td>
</tr>
</tbody>
</table>

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

Version 1/2017
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
• Hearing loss in congenital toxplasmosis detected by Newborn Screening. Andrade et al, Braz J Otorhinolaryngol 2008; 74:21-28
• 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 [Link]
• BASHH UK National Guidelines on Management of Syphilis 2015 [Link]