

# **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

### **Contributors:**

Dr Nicola Price, Consultant Virologist Public Health Wales.

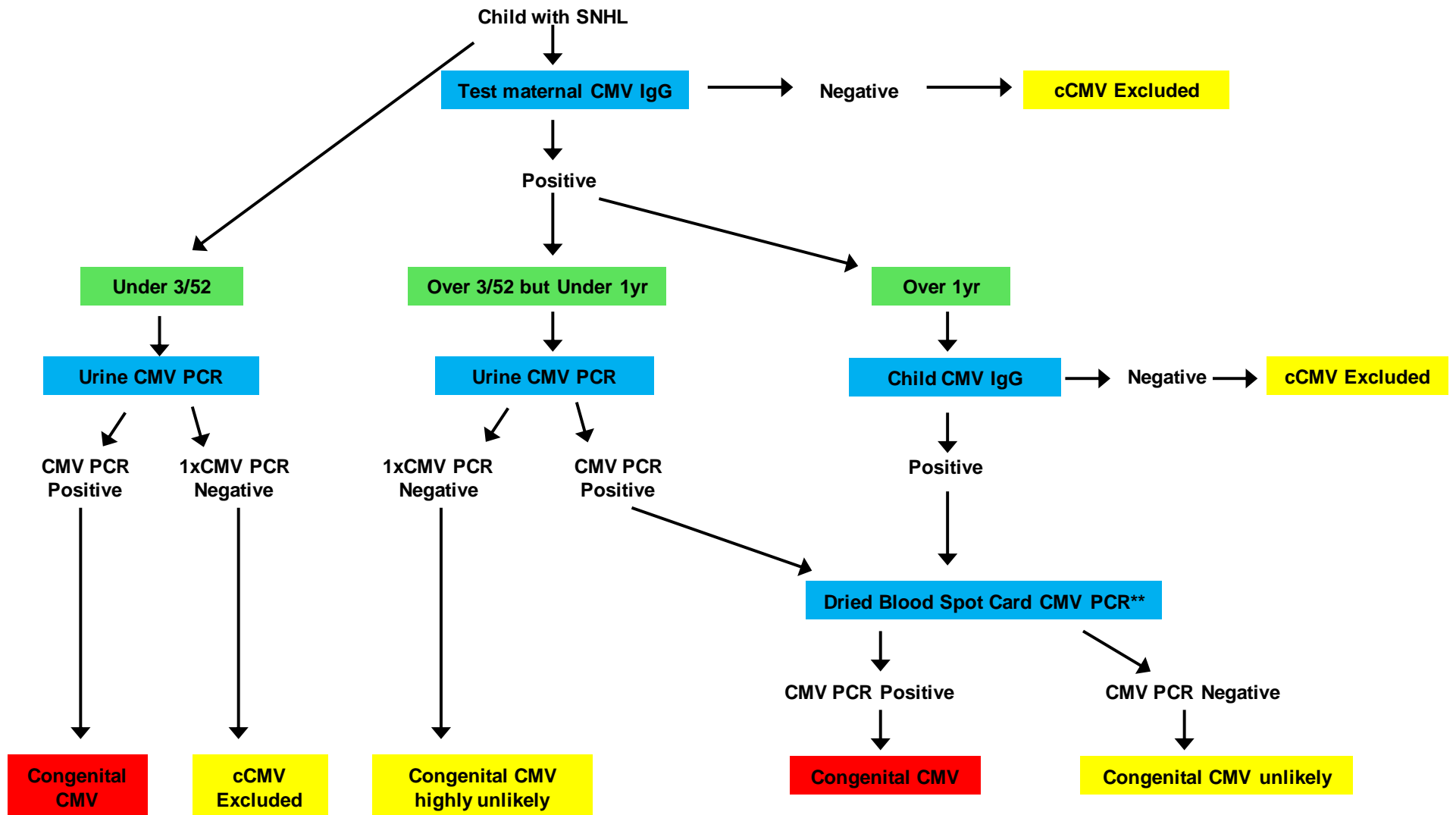
Dr Amanda Roberts, Programme Coordinator Newborn Hearing Screening Wales.

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\*



## **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: Evidence based management guidelines for the detection and treatment of congenital CMV.

S. Kadambari, E.J. Williams, S. Luck, P.D. Griffiths, M. Sharland. Early Human Development 87 (2011) 723–728.

Reference: Is saliva as reliable as urine for detection of cytomegalovirus DNA for neonatal screening of congenital CMV infection?

Aparecida Yulie Yamamoto, Marisa Marcia Mussi-Pinhata, Lauro Juliano Marin, Rosangela Moura Brito, Patricia Frizzo Carvalho Oliveira, Thalita Bonadio Coelho. Journal of Clinical Virology 36 (2006) 228–230

## Section 2: Rubella , Toxoplasma, Syphilis

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

CMV	Rubella	Toxoplasma	Syphilis
<p>See existing pathway  <a href="http://www.wales.nhs.uk/sitesplus/documents/980/NBHSW%20Medical%20Carepathway%20-%20flow%20chart%20finalJuly2013.pdf">http://www.wales.nhs.uk/sitesplus/documents/980/NBHSW%20Medical%20Carepathway%20-%20flow%20chart%20finalJuly2013.pdf</a></p>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Mother or child from abroad</li> <li>• Mother travelled abroad during pregnancy</li> <li>• Mother didn't receive 2MMRs prior to pregnancy</li> <li>• Signs of Congenital Rubella Syndrome (2 from A or 1 from A + 1 from B)</li> </ul> <p>➤ <b>Group A:</b> Cataracts, Congenital Glaucoma, Congenital Heart Disease, Hearing Loss, Pigmentary Retinopathy</p> <p>➤ <b>Group B:</b> Purpura, Splenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent Bone Density, Jaundice</p>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Chorioretinitis</li> <li>• Features of congenital toxoplasma Hydrocephalus, Intracranial calcifications, Microcephaly, Microphthalmia, Seizures, Hepatosplenomegaly, IUGR, Jaundice, Purpura</li> </ul>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Bone abnormalities (frontal bossing, saddle nose, sabre shins, shortened maxilla)</li> <li>• Teeth Abnormalities (Hutchinson's incisors, mulberry molars) (see Fiumara and Lessel 1983)</li> <li>• Interstitial keratitis</li> </ul> <p>More usual as a late presentation, after 2 years of age. Prior to this age, test if suspicious of congenital syphilis.</p>
	<p>Test maternal IgG</p> <pre> graph TD     A[Test maternal IgG] --&gt; B[Positive]     A --&gt; C[Negative]     B --&gt; D[Test child IgG]     C --&gt; E[Exclude]     D --&gt; F[Positive]     D --&gt; G[Negative]     F --&gt; H[Ask if child received MMR Seek further advice (?IgM tests, ?booking blood, ?serial IgG/avidity tests)]     G --&gt; I[Exclude]         </pre>	<p>Test maternal IgG</p> <pre> graph TD     A[Test maternal IgG] --&gt; B[Positive]     A --&gt; C[Negative]     B --&gt; D[Test child IgG]     C --&gt; E[Exclude]     D --&gt; F[Positive]     D --&gt; G[Negative]     F --&gt; H[Further reference laboratory testing performed]     G --&gt; I[Exclude]         </pre>	<p>Maternal syphilis antibodies</p> <pre> graph TD     A[Maternal syphilis antibodies] --&gt; B[Positive]     A --&gt; C[Negative]     B --&gt; D[Test child Elisa/RPR/VDRL/IgM]     C --&gt; E[Exclude]     D --&gt; F[Positive]     D --&gt; G[Negative]     F --&gt; H[Refer to Paediatrician for further testing/specialist advice]     G --&gt; I[Exclude]         </pre>

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

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

CMV	Rubella	Toxoplasma	Syphilis/HSV
<p>See existing pathway  <a href="http://www.wales.nhs.uk/sitesplus/documents/980/NBHSW%20Medical%20Carepathway%20-%20flow%20chartfinalJuly2013.pdf">http://www.wales.nhs.uk/sitesplus/documents/980/NBHSW%20Medical%20Carepathway%20-%20flow%20chartfinalJuly2013.pdf</a></p>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Mother or child from abroad</li> <li>• Mother travelled abroad during pregnancy</li> <li>• Mother didn't receive 2MMRs prior to pregnancy</li> <li>• Signs of Congenital Rubella Syndrome (2 from A or 1 from A + 1 from B)</li> </ul> <p>➤ <b>Group A:</b> Cataracts, Congenital Glaucoma, Congenital Heart Disease, Hearing Loss, Pigmentary Retinopathy</p> <p>➤ <b>Group B:</b> Purpura, Splenomegaly, Microcephaly, Mental Retardation, Meningoencephalitis, Radiolucent bone density, Jaundice</p> <p style="text-align: center;">Test current maternal IgG</p> <p style="text-align: center;">Positive ←————→ Negative</p> <p style="text-align: center;">↓ Exclude</p> <p>Test in parallel with booking sample for IgM/IgG and discuss with Consultant Virologist</p> <p>↓</p> <p>If unable to exclude maternal primary infection then test serum from baby for IgM (a negative result will exclude Congenital Rubella at this age)</p>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Known or suspected maternal Toxoplasma infection acquired during pregnancy</li> <li>• Features of congenital toxoplasmosis</li> </ul> <p>Hydrocephalus            Intracranial calcifications            Chorioretinitis            Microcephaly            Microphthalmia            Seizures            Hepatosplenomegaly            IUGR            Jaundice            Purpura</p> <p style="text-align: center;">Test current maternal IgG</p> <p style="text-align: center;">Positive ←————→ Negative</p> <p style="text-align: center;">↓ Exclude</p> <p>Send to reference laboratory with booking sample for IgM/IgG</p> <p>↓</p> <p>If unable to exclude maternal primary infection then seek specialist advice and send samples from the baby (serum, EDTA, CSF)</p>	<p>Test if:</p> <ul style="list-style-type: none"> <li>• Known maternal syphilis infection (should be part of birth plan if UK-born) (See ASW infections and Rashes in Pregnancy 2016)</li> <li>• Features of congenital Syphilis</li> </ul> <p>Rash            Haemorrhagic rhinitis            Generalised Lymphadenopathy            Hepatosplenomegaly            Skeletal Abnormalities            (See BASHH UK Guidelines on Management of Syphilis 2015 for testing strategy)</p> <p>Test if:</p> <ul style="list-style-type: none"> <li>• Features of neonatal HSV infection</li> </ul>

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

## 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
- Newborn hearing screening programme in Belgium: a consensus recommendation on risk factors. Vos et al, BMC Pediatr. 2015;15:160
- Hearing loss in congenital toxoplasmosis detected by Newborn Screening. Andrade et al, Braz J Otorhinolaryngol 2008; 74:21-28
- A systematic review of neonatal toxoplasmosis exposure and sensorineural hearing loss. Brown et al, Int J Pediatr Otorhinolaryngol 2009; 73:707-711
- Maternal infection with toxoplasma gondii in pregnancy and the risk of hearing loss in the offspring. Austeng et al, Int J Audio 2010;49:65-68
- The stigmata of late congenital syphilis: an analysis of 100 patients. Fiumara and Lessel, Sex Transm Dis 1983; 10: 126
- A systematic review of the incidence of sensorineural hearing loss in neonates exposed to Herpes simplex virus (HSV). Westerberg et al, Int J Pediatr Otorhinolaryngol 2008; 72:931-937
- Antenatal Screening Wales Infections and Rashes in Pregnancy  
<http://www.antenatalscreening.wales.nhs.uk/sitesplus/documents/989/V2a-Final%20Infections%20in%20Pregnancy%202016.pdf>
- BASHH UK National Guidelines on Management of Syphilis 2015  
<https://www.bashhguidelines.org/media/1053/syphilis-2015.pdf>