Summary of guidelines for aetiological investigation into progressive permanent childhood hearing impairment

**Level 1 investigations**: Recommended for all children with progressive PCHL

1) **Clinical history**: [Recommendation D]

2) **Clinical examination**: [Recommendation D]

3) **MRI IAM & brain / CT Petrous Temporal**: [Recommendation C]

4) **CMV testing**: [14, 24-34] [Recommendation B]

5) **Family audiograms**: [14, 35,36] [Recommendation D]

6) **Ophthalmic assessment**: [Recommendation B]

7) **Urine examination (labstix) for microscopic haematuria and proteinuria**: [Recommendation D]

8) **Genetic tests**: GJB2/GJB6 [Connexin 26/30] and for m.1555A>G [Recommendation B]

9) **Serology for other infections** [Recommendation C/D]
   - Congenital toxoplasmosis
   - Congenital Rubella
   - Congenital Syphilis
   - Congenital HIV

10) **Investigation for autoimmune diseases** [Recommendation C/D]

**Level 2 investigations**: for selected children depending on clinical features

1) **Haematology and Biochemistry**: [Recommendation D]

2) **Renal ultrasound**: [Recommendation D]

3) **Metabolic Screen on blood and urine**: [Recommendation D]

4) **Lyme disease serology**: [Recommendation D]

6) **CGH microarray**: [Recommendation D]

7) **Further genetic testing**: [SLC26A4, BOR, Usher, gene panel, whole genome sequencing] [Recommendation C/D]

8) **Referral to Clinical Geneticist**

9) **Vestibular investigations**: [Recommendation C/D]

10) **Referral to Paediatric Rheumatologist/Immunologist**