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Contemporary Practice

Hong Kong Universal Newborn Hearing Screening (UNHS) Care Path Protocol under Joint Committee on UNHS

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HA-DH Joint Committee on Care Path for Universal Newborn Hearing Screening

Abstract The Joint Committee on Universal Newborn Hearing Screening (UNHS) was formed to ensure the effective implementation of UNHS in Hong Kong and to achieve effective communication among various services in Hospital Authority and Department of Health. Meetings with multidisciplinary professionals had been conducted during the years of 2013 and 2017. A consensus on a common care path for infants identified to have hearing impairment had been reached.

Key words *Hearing; Newborn; Screening*

Background

The Joint Committee on Universal Newborn Hearing Screening (UNHS) was formed to ensure the effective implementation of UNHS in Hong Kong and to achieve effective communication among various services in Hospital Authority (HA) and Department of Health (DH). Meetings with multidisciplinary professionals had been conducted during the years from 2013 to 2017. A consensus on a common care path for infants identified to have hearing impairment had been reached (Appendix I).

Children with hearing impairment are usually classified according to the degree and nature of the impairment. Hearing impairment can be conductive, sensorineural, or mixed in nature. Hearing is measured in decibels hearing level (dB HL) with the threshold of 0 dB for each frequency denoting the value at which normal young adults perceive a pure tone of a given intensity and frequency 50% of the time. The table below shows one of the more commonly used classification systems for the degree of hearing impairment in Hong Kong.

Degree of hearing impairment	Hearing impairment range (dB HL)
Mild grade	26 to 40
Moderate grade	41 to 55
Moderately severe grade	56 to 70
Severe grade	71 to 90
Profound grade	>90

Goodman A. Reference zero levels for pure tone audiometers. ASHA 1965; 7:262-3.[1]

Children with hearing impairment (HI) greater than or equal to moderate grade are classified as having significant hearing impairment. Early identification, diagnosis, and intervention are crucial to the successful management of these children to alleviate the possible adverse outcome on the language and communication, cognition, reading, and social-emotional development.

Over 5% of the world population that is 360 million people has significant hearing impairment (328 million adults and 32 million children).[2] The international prevalence of permanent childhood hearing impairment (PCHI) of moderate or greater degrees is about 1 to 3 per 1,000 children, based on screening and/or medical records.[3,4] The number increases to 1 in every 40 for infants who require care in the Neonatal Intensive Care Unit.[5] In Hong Kong, for every 1,000 children under the age of 15 years, 1.3 of them were registered with significant HI in the Central Registry for Rehabilitation in 2014. At the Child

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Assessment Service (CAS) of the Hong Kong DH, new cases received with PCHI of moderate or greater degrees remained relatively stable over the past decade, with 61 new cases in 2007 and 49 in 2016. With the universal screening programme in place and referring to the mean number of births per year in Hong Kong over the period and CAS's catchment coverage of over 85% of the population, an incidence of about 0.14% of children having PCHI is estimated, comparable to prevalence of developed regions.

The rate of the coexistence of additional disabilities or medical conditions with hearing impairment in children is high. Most estimates suggest that between 30% and 40% of children with hearing impairment have one or more additional disabilities.[6] Studies have reported that the frequency of additional disabilities is similar across all levels of hearing impairment, pointing to the need for children with mild or unilateral impairment, as well as those with more severe impairment, to receive thorough medical and developmental evaluations.[7] Among those with additional disabilities, the commonest ones are developmental delay (33%), cerebral palsy (20%), vision problem (7.6%) and autism spectrum disorder (7.6%).[8]

Children with significant hearing impairment are at risk of growing up with deficits in language, communicative abilities, cognition and literacy, psychosocial functioning and possible problems in balance and gross motor proficiency.[9]

Many in developed countries changed from using the term "Universal Newborn Hearing Screening" to "Early Hearing Detection and Intervention (EHDI)" programs. The change underscores that successfully identifying and serving infants and young children with PCHI, requires going beyond screening to address issue related to audiological diagnosis, appropriate medical and educational intervention, and coordination with the child's family.[10]

Hearing Screening for Children in Hong Kong: Universal Hearing Screening After Birth in HA Hospitals

HA hospitals would provide universal newborn hearing screening and make initial diagnosis for patients with congenital or delayed hearing loss.

Screening Methods and Confirmatory Tests

Currently 2-stage Automated Auditory Brainstem Response (AABR) screening program (or alternative acceptable screening program in future) would be offered to all neonates born in HA hospitals or outborn babies transferred in from other hospitals. Babies who had failed these screening tests (with AABR threshold > 35 dB) would be referred for confirmatory Brainstem Auditory Evoked Potential (BAEP) test. Initial counselling would be offered to their parents and information sheet would be given to them. In keeping with the latest American Academy of Pediatrics Guidelines for newborn hearing screening, BAEP should preferably be performed before 4 months of age to facilitate early detection and subsequent interventions for babies with congenital hearing loss. Babies who failed confirmatory BAEP test would be referred to Ear, Nose and Throat (ENT) Clinic; at which comprehensive hearing assessment by otolaryngologist (ENT specialists) would be performed. These babies would also be referred to paediatric clinics for follow-up and investigations.

Infants with Risk Factors

For infants with risk factors associated with permanent congenital, delayed-onset or progressive hearing impairment in childhood, follow-up at HA Paediatric Clinics should be considered.

These risk factors include:

1. Family history of permanent childhood hearing impairment
2. Neonatal intensive care for more than 5 days
3. Extracorporeal membrane oxygenation, assisted ventilation, exposure to ototoxic drugs (which toxic level was detected on drug monitoring*)
4. Severe neonatal jaundice requiring exchange transfusion
5. In utero infections, such as cytomegalovirus (CMV), herpes, rubella, syphilis and toxoplasmosis
6. Head trauma, especially fracture base of skull and fracture of temporal bone
7. Caregivers' concern regarding hearing loss
8. Significant craniofacial anomalies
9. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
10. Syndromes associated with hearing loss or progressive or late-onset hearing loss such as neurofibromatosis, Pendred syndrome and Jervell and Lange Nielson syndrome
11. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis

** For hospitals in which drug level service is not available, they might offer repeat hearing test a few months after discharge.*

Screening for Congenital CMV Infection

Urine CMV screening (within 3 weeks of age) should be considered for infants who have failed the Newborn Hearing screening twice. Congenital CMV infection is the most frequent non-hereditary cause of sensorineural hearing impairment worldwide. Studies had demonstrated early administration of antiviral treatment in symptomatically infected infants with CMV infection prevents hearing deterioration at 6 months and may potentially prevent hearing deterioration at > or =1 year. [11,12]

Supplementary Screening in Maternal & Child Health Centres (MCHCs) of Family Health Service

MCHCs would provide screening for those infants who had not been screened by their birthing hospitals (e.g. private hospitals), currently by 2-stage Automated Otoacoustic Emission (AOAE) method. The procedure and potential limitations of AOAE test would be explained to parents before the screening.

For those infants who failed AABR screening once, parents would be advised to return to their birth hospitals for completion of the AABR test. If the parents refuse to return to the hospitals (where initial AABR was performed), their babies would be referred to HA ENT for further management of failed AABR. These infants would be screened irrespective of their risk status except those with absent auditory canal. AOAE screening would not be provided for those who had already been referred for audiological assessment or babies more than 4 months of age.

First AOAE screening would be performed for infants aged between 2 weeks to 4 months. Preferably this first screening would be performed within 2 weeks after the first MCHC registration. Second AOAE test would be offered to those infants who failed the initial AOAE test, preferably within one week after the first AOAE. For those who failed AOAE test twice, they would be referred to HA ENT or audiologist services for confirmatory testing.

Routine hearing surveillance and developmental surveillance would be provided to all children even for those who had passed both AOAE or who had passed the UNHS program in HA or private hospitals. If they fail to achieve the pre-determined developmental milestones or if there are parental concerns on possible hearing impairments, these children would be referred to HA ENT or audiology services for further evaluations if indicated.

Management of Children with Confirmed Hearing Impairment

Referral to Other Services After Identification of Hearing Loss

Referrals may be made to CAS, The Duchess of Kent Children's Hospital at Sandy Bay Child Assessment Centre (DKCAC), Clinical Genetic Service (CGS), ophthalmologists or other specialties as clinically indicated. Patients with mild grade hearing impairment would be followed-up at HA ENT Clinics. Behavioural tests (paediatric audiometry) would be performed for babies with hearing impairment at around 6-9 months of age for better delineation on the degree of hearing impairments for these babies. For babies with severe or profound hearing impairment, early imaging studies (CT and MRI) would be performed to look for any structural anomalies and to plan for cochlear implantations. These babies should also be referred to ophthalmologist for assessment in view of the high rate of associated visual abnormalities. For babies with persistent HI, referral will be made to appropriate service for hearing aids arrangements.

CAS and DKCAC

CAS or DKCAC would receive referral from HA hospitals, private hospitals, Education Bureau and MCHC for further workup and management for patients with hearing impairment. After identification of hearing impairment, CAS or DKCAC would take up the co-ordination role in multi-disciplinary management of these children.

Referral could be made to CAS or DKCAC by the ENT surgeons for infants with the following criteria, after ENT assessment:

1. Significant hearing impairment cases (permanent hearing impairment of moderate degree or worse in the better ear)
2. Unilateral severe to profound sensorineural hearing impairment
3. Bilateral severe to profound high frequency hearing impairment

Children with mild sensorineural hearing loss could be followed-up by ENT. Patients with suspected syndromal diagnosis or high risk of developmental delay could also be directly referred to CAS or DKCAC by the HA hospitals Paediatrics Department for follow-up and detailed evaluation.

After receiving referral for significant hearing impairment, nurse interview would be conducted for brief history taking at CAS. Audiologists would provide information on available community resources and counselling to the parents. Audiologists in CAS or DKCAC may perform hearing assessment again if the child is older than 6 months of age. School placement for Early Education and Training Centre (EETC) (deaf) or EETC and Special Child Care Centre (deaf) would be arranged for them.

In CAS, when the hearing impairment is diagnosed at 3 months to 6 months of age, the child would be assessed by paediatric doctor for a comprehensive physical and developmental assessment within 2 months after the referral. A team conference with paediatric doctor, audiologist and medical social worker would be conducted to formulate the final management plan for the patient. If the child is aged more than 6 months, CAS would provide physical and developmental assessment by paediatric doctor, interim language support by speech therapist, and a team conference with paediatric doctor, speech therapist, audiologist and medical social worker would be arranged for management plan and counselling. The child may be referred to HA specialist for further investigation and management if necessary.

In DKCAC, when the hearing impairment is diagnosed, the child would be assessed by the paediatric doctors for a comprehensive assessment and an aetiology work-up within 2 months. Subsequently, the child would go through a multi-disciplinary assessment with paediatric doctors, audiologists, speech therapists and medical social workers to formulate his/her final management plan. The child may also be referred to other HA specialists for further investigation and management when necessary.

Assessment of the child's functioning in the aspects of hearing, communication and language, cognition, literacy and motor development helps to determine what type of training the child needs. Together with audiologists, developmental paediatricians take comprehensive medical and family histories to identify possible risk factors and aetiology. Detailed physical examinations are performed for possible aetiology and any associated physical abnormalities. Developmental paediatricians assess the strengths and weaknesses of the child in all developmental areas and evaluate the presence of any associated comorbidities. A thorough evaluation of the child's level of communication, speech and language development by the speech therapist is important and should include assessment on verbal comprehension, verbal expression, speech production, and speech perception. Assessment of spoken language for these children should follow similar procedures as those for the hearing children but adequate visual cues should be noticed. Phonological errors and error patterns should be analysed. Speech perception tests include assessment of the discrimination and identification of suprasegmental and segmental features should be included to reflect the degree of communication handicap in everyday situation.

Audiologists at CAS would perform yearly phone follow-up for children with hearing aids until these children enter primary school. They would follow-up on problems of using hearing aid, any further audiological results, language development and difficulties in school coping, etc.

Paediatric doctors at CAS would review these children at 18 to 24 months of age. This aims to assess for any improvement after application of hearing aids or cochlear implant and review of placement. If parents decline referral for preschool services for children with hearing impairment, speech therapists would review their language progress at 24 to 30 months of age. For children with significant hearing impairment, a combined multi-disciplinary assessment by paediatric doctor (+/- clinical psychologist), audiologist and speech therapist would be arranged before the children go to primary school. Gross motor screening would also be performed for this group of children to look for potential motor balance problems. Advice on school placement, accommodation, continual auditory and language training would be given.

Paediatric doctors at DKCAC would also review these patients at 18 to 24 months of age for developmental assessment and review of replacement. Speech therapists would then review them at 24 to 30 months of age for speech and language progress. Prior to their admission to primary school, an evaluation by paediatric doctors would be conducted. Gross motor screening would also be performed for them to look for potential motor balance problems.

CGS

CGS would provide genetic assessment, testing and counselling for patients with hearing impairments and suspected genetic diseases.

Referral criteria for children with hearing impairment to CGS:

1. Suspected genetic or syndromic cause of hearing impairment
2. Positive family history of congenital hearing loss, especially among first degree relatives
3. Specific inner ear malformations or agenesis and/or coexisting abnormalities involving other organ systems
4. Parenteral consanguinity
5. Parental request, especially for family planning purpose
6. For interpretation and counselling of relevant genetic testing results
7. Hearing impairment in a child exposed to aminoglycoside antibiotics

Please note that referral could be made irrespective of the severity of hearing impairment. Patients without syndromic features could also be referred to CGS if there was parental consanguinity or there was parental request, especially for family planning purposes.

Concerning non-syndromic-related hearing impairments, only 3 gene tests are available for the time being (GJB-2 gene, GJB-6 gene and mitochondrial DNA 155C2A gene). These genetic testing had low pickup rates of only around 30%.

Declaration of Interests

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Endorsement

This protocol had been endorsed by COC (Paed) in November 2017.

This protocol had been presented to COC (ENT) in November 2017.

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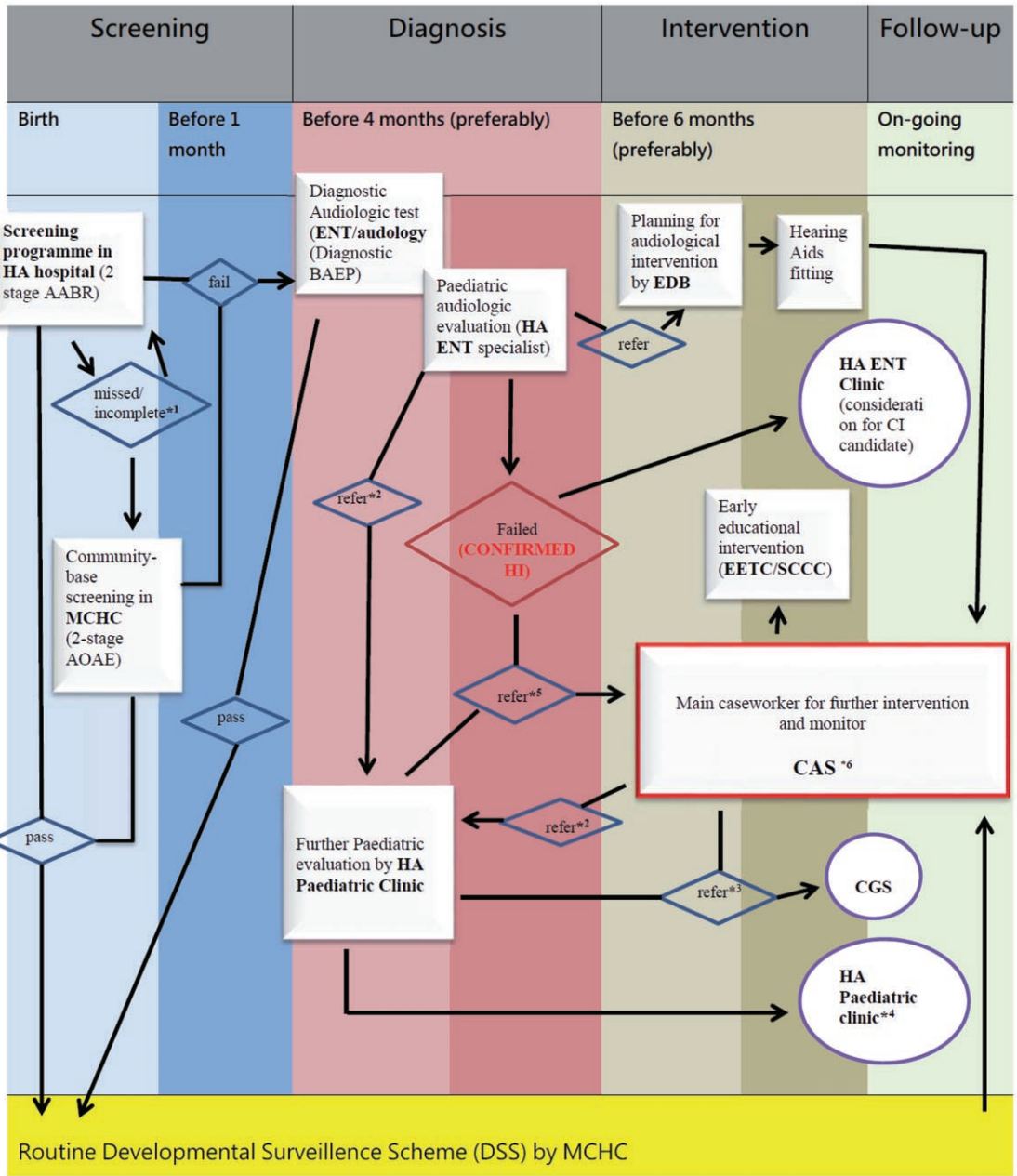
Appendix I. Algorithm for universal newborn hearing screening in Hong Kong

Universal Newborn Hearing Screening, Diagnosis, Intervention and Follow-up



Notes:

*1 For those infants who did not complete the hospital base AABR programme, would be advised to return to their birthing hospital for completion of the test

*2 Patient should be referred to paediatric clinic for further evaluation if not previously done

*3 Please refer to section on Role of CGS for referred criteria

*4 Please refer to section on infants with Risk factors

*5 Please refer to section on Role of CAS for referral criteria

*6 For infants born in Queen Mary Hospital (QMH), they will be referred to DKCAC (under HA) for further management

Appendix II. Members of the HA-DH Joint Committee on Care Path for UNHS

Hospital Authority

Dr. William Wong, Con(Paed), PWH (Chairperson)
Dr. Wilson Yeung, COS(Paed), PYNEH (Chairperson)
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