Brainchild

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香港兒童腦科及體智發展學會

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香港兒童腦科及體智發展學會

The Council

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Cover

The cover picture is a watercolour drawing from a hearing impaired individual. It is a fine example of the talents in the keen observation of shapes and colours so often exhibited by them.

The Hong Kong Society of Child Neurology & Developmental Paediatrics
Brainchild - January 2003 issue

Message from the President

Dr. CHAN Chok Wan

Since its inauguration in December 2000, Brainchild provides effective platform for excellent scientific papers on child neurology and developmental paediatrics with contributors from overseas and the local Hong Kong community. In order to provide more in-depth discussions and more comprehensive coverage on specific clinical problems, the Editorial Board resolves to modify the format of the Education Section with focus on special themes for each issue to effect maximal efficiency of the publication.

We are pleased to witness the first issue of Brainchild consequent to the modification themed on *Hearing and its Related Problems in Childhood*. Appreciation is being expressed to Dr. Ada Yung, Guest Editor, for her dedicated effort in preparing this Issue, and to all authors for their excellent papers which cover important topics including *Auditory and Speech Rehabilitation of Profoundly Deaf Children in Hong Kong Using Multichannel Cochlear Implant, Newborn Hearing Screening, The Role of the Child Assessment Centre in the Identification and Assessment of Childhood Hearing Loss in Hong Kong, Central Auditory Processing Disorders (CAPD), and Services Provided for Hearing Impaired Children by the Education Department*. These presentations highlight different facets of the disorder and provide effective solutions to tackle the problems: an excellent illustration of multidisciplinary, interdisciplinary and transdisciplinary approach to childhood developmental disorders. I am confident readers will find the articles informative, useful and handy for their daily practice.

At the Society level, the Hong Kong Society of Child Neurology and Developmental Paediatrics has been very active and enthusiastic on scientific and professional affairs related to our subspecialties. The Working Party on Specific Learning Disabilities (SLD) has hosted a Special Workshop for Teachers at the Hong Kong City Hall on 12th May 2002 providing basic information, concept and clinical features on SLD for frontline workers at school. We have organized joint meeting with the Equal Opportunity Commission of Hong Kong on 22nd June 2002 at the Hong Kong Convention and Exhibition Centre on education rights of children with SLD. We also proactively had meetings with the Hong Kong Examination and Assessment Authority on 29th July 2003 advocating accommodation for children with SLD at public examinations and with Heads of the Special

Education Service of Education Department of the Hong Kong SAR Government on 7th October 2003 on service provisions at school for SLD. All meetings yielded constructive results and we are pleased to witness fruitful actions consequent to the meetings from the parties concerned. Climax of these endeavours is the Summit Meeting on 19th October 2003 between the Working Party with Mrs. Fanny Law, Permanent Secretary for Education and Manpower, Dr. Margaret Chan, Director of Health, Professor CK Leong and Dr. Connie Ho, local experts on the subject, and colleagues from the Special Education Service of Education Department on 3rd October 2003 whereby all participants engaged in friendly, sincere, serious and critical discussions about the problem in Hong Kong. We are pleased to witness two significant projects being spinned off from the meeting. A 30-Hour Course for 800 Primary School Teachers and a 7-Hour Course for 1,200 School Principals (both Primary and Secondary) will be organized by the Society to arouse awareness and provide basic information for understanding the problems and management for children with Special Educational Needs (SEN) at school. The Society was also empowered to submit an official proposal on principles to tackle the problems of SLD in Hong Kong. The Society is very encouraged with these achievements which signify official recognition of the problems of SLD and commitment of our SAR government towards solution of the disabilities. The Society will endeavour to do our utmost to effect both projects so as to bring maximal beneficial effect to our children with SLD whose welfare and rights have been our main concern all along.

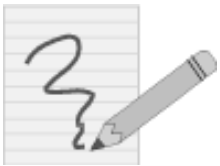
Our Society is proud to have successfully hosted the International Conference 2002 on *Dyslexia in Children Using the Chinese Language* on 26-28th October 2002 in Hong Kong with focus on Functional MRI (fMRI) and Advocacy Issues. We are honoured to have Mr. Emerson Dickman, an eminent US attorney at law specialized on advocacy for children with SLD, and Professor CK Leong, world expert on dyslexia in children using the Chinese language, as our keynote speakers. These scholars together with eight overseas speakers from the United States, Singapore, Taiwan and the Mainland of China as well as 20 local speakers contributed successfully to a Conference comprising of two keynote lectures, four seminars, one open forum, one open lecture and dinner, and a satellite symposium hosted jointly by our Society and the Support Group on Integrated Education on the topic of *Towards Better Integrated Education*. The attractive and comprehensive programme attracted active participation from more than 450 participants covering a large number of professionals including medical, nursing, teachers, educational and clinical psychologists, speech and hearing pathologist, social workers, administrators, policy makers, legislators, parents and others. We are very fortunate to have Miss Anna Wu, Chairperson for the Equal Opportunity Commission of Hong Kong, addressing the audience at the Opening Ceremony on advocacy issues for children with SLD in Hong Kong. Her speech as always was both enlightening and forceful while her words of wisdom did shine light onto future direction for our work in the area of SLD. Post-meeting evaluation confirmed that the Conference has indeed achieved its goals and objectives of promoting knowledge in the fundamental nature of, and the professional, social and political issues required

for addressing the subject. It sets a platform for exchange of experience, sharing of knowledge, collaborating of research and cooperating of future professional activities and meetings. It has cast a monumental milestone for scholars and professional working for dyslexia in the Chinese language to converge and open dialogues for cooperation. It is extremely encouraging to witness eager and sincere commitment of local and international participants for engaging in future similar meetings and how they treasured the fraternity and friendship cultivated via this Conference. I would like to take this opportunity to express our heartfelt gratitude to members of the Organizing Committee, notably Dr. Catherine Lam, Professor CK Leong, Dr. Philomena Tse, Dr. Becky Chiu, Dr. Wu Shun Ping, Dr. Tsui Kwing Wan, Dr. Sharon Cherk, and Miss Anciently Chan, to Society Council Members for their dedicated effort, and to all Members for their ever-unfailing support. Most important of all, we are indebted to Wyeth for sponsoring this important Conference which facilitated realization of the visions and missions of our Society.

Other than the area of SLD, our Society has also been very active with other areas contributed by the Working Party on Epilepsy Surgery, convened by Dr. Dawson Fong and Dr. Sharon Cherk, and the Working Party on Physically Impairment, convened by Dr. Sophelia Chan and Dr. Tsui Kwing Wan. Both have dedicatedly worked within their designated duties and achieved outstanding results. We thank them all for their effort. The Annual Scientific Meeting 2003 will be on Neurological Intensive Care by Dr. Robert Tasker from Cambridge UK provisionally scheduled in October 2003 and we can promise professionals in Hong Kong another quality meeting of top professional standard on the subject.

In conclusion, we have achieved a significant plethora of projects over the past few months. In view of the small membership of our society and the limited resources available, I have to pay tribute to all Members and the Society Council for their hardwork and dedication. We together serve to illustrate the omnipotency of child neurologists and developmental paediatricians for our being able to effectively play multiple key roles as professionals, clinicians, academics, administrators and organizers. The Society is proud to have you all and look forward to your continuing contribution in the future. We need your support to bring our activities into new horizon of attainments!

Editor-in-Chief, Brainchild
President, HK Society of Child Neurology & Developmental Paediatrics



Education Section

Auditory and Speech Rehabilitation of Profoundly Deaf Children in Hong Kong Using Multichannel Cochlear Implant

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Introduction

The provision of cochlear implantation signifies an important milestone in the history of rehabilitation of hearing-impaired children. Normal hearing is a prerequisite for the development of speech perception, speech production and language competence. Speech production and perception are seriously affected without an intact auditory feedback loop.[1] In young mammals including humans, an adequate sensory experience is critical to the developing nervous system for the expression and maintenance of certain sensory functions. Animal studies on auditory deprivation with induced deafness a few days after birth showed significant subsequent morphological and physiological changes in the central auditory system.[2-6] These studies gave rise to the concept of a 'critical period'. The critical period refers to certain points in times where the sensory experience and environment exert a major influence in the development of neural systems or their organization.[7,8] Auditory deprivation during the critical period will lead to atrophy or underdevelopment of structures related to the processing of auditory inputs. Areas of the auditory cortex will also be usurped by other modalities.[7,8] Although there is no consensus on the upper age limit of the critical period, it is generally agreed that auditory deprivation during the first 3 years of life may lead to irreversible audition and language retardation.[9] Therefore, it is important to detect hearing impairment in children early enough so that rehabilitation through various hearing prosthesis can be provided to ameliorate the devastating effects from early auditory deprivation.

Although early cochlear implantation is advocated for young profoundly hearing-impaired children, the small dimension and shallowness of the skull require special attention and meticulous manipulation. Children for receive cochlear implants (CIs) are getting increasingly younger. The National Institutes of Health[10] has recommended selection criteria for cochlear implantation in children. In general, the selection criteria are: (a) 18 months to 17 years old, (b) bilateral profound deafness and an inability to benefit from appropriate conventional hearing aids, (c) no medical contraindication to implantation, (d) support and realistic expectations from family members and significant others, (e) willingness to participate in post-operative evaluation and training, and (f) enrolment in an educational setting with a strong auditory component. Children younger than 18 months of age have also received CIs. The main indication for these patients is meningitis, which can lead to cochlear ossification and consequently prevent the electrode from being successfully inserted into the scalar tympani.

CIs have been demonstrated to aid the successful rehabilitation of children whose native spoken languages are of Latin origin.[11,12] Very few studies have reported on the speech perception performance of cochlear-implanted children in Hong Kong.[13,14] In these studies, the age of the implanted children ranged from 2 years to 12 years old. Speech perception and the linguistic skills of children below 3 years of age are very limited compared with those of older children. A tailor-made test battery to assess the auditory and speech perception abilities in congenitally deafened young children is required.

In 1994, the CI team of the University of Hong Kong Medical Centre at Queen Mary Hospital performed the first paediatric cochlear implantation in a 5-year-old profoundly deaf child whose mother tongue was Cantonese. As of 2001, 82 hearing-impaired children had undergone cochlear implantation in our centre. This study examines the pre- and postoperative auditory and speech perception results of young cochlear-implanted children after 1 year of intense post-operative training.

Methods

From 1994 to 2001, 14 prelingually deafened children (7 boys, 7 girls) underwent multichannel cochlear implantation at Queen Mary Hospital. The age of the patients ranged from 1.3 years to 3 years (median: 2.2 years). Table 1 lists the causes of deafness of the subjects.

Table 1. Distribution of various causes of deafness of the 14 hearing-impaired prelingually deafened children

Causes of deafness	Number of children	Percentage
Unknown	5	36
Deformed cochlea	3	26
Ototoxicity	2	14
Heredity	2	14
Large vestibular aqueduct	1	7
German measles	1	7
Meningitis	1	7

In paediatric cochlear implantation, a team approach was adopted to manage the medical, rehabilitative, social and academic needs of the child. The CI team in Queen Mary Hospital comprised of paediatric ENT surgeons, radiologists, audiologists, speech therapists, social workers, teachers of the deaf and parents. The teachers of the deaf could provide expertise in evaluating and observing the pre- and post-operative academic performance of the implanted children. Parents of the hearing-impaired children received also training in auditory and speech perception skills so that they could provide everyday training to their children in an accustomed home environment.

The candidacy for cochlear implantation of these young children was evaluated by conducting age-appropriate hearing tests such as behavioural observation audiometry, play audiometry and impedance audiometry. The reliability of the obtained puretone thresholds was verified by electrophysiological tests that included auditory brainstem response audiometry and otoacoustic emissions. When bilateral profound sensorineural hearing loss was confirmed, the children then underwent a 3-month hearing-aid trial to observe whether they could benefit from hearing amplification. At the end of the hearing-

aid trial period, auditory and speech perception abilities were re-evaluated with a battery of tests. The results from these speech tests set the preoperative baseline for comparison with post-operative performance.

Medical assessments included otologic history and physical examinations. An otologically stable ear without ongoing infection is important to avoid infective complications such as meningitis. Radiologic evaluation was important to determine the presence of any congenital deformation of the cochleas. MRI was performed to evaluate the structure of the cochlea and to confirm the presence of auditory nerves. CT scans were carried out to evaluate intracochlear obliteration as well as the shape and size of the internal auditory meatus (IAM).[15] When abnormal IAM narrowing was suspected, an electrical auditory brainstem response auditory was performed to evaluate the integrity and electro-conduction of the auditory nerves. Congenital malformation and obliteration of the cochleas were not factors that contraindicated implantation. Specially designed short or split electrode arrays could be inserted surgically to circumvent such abnormalities. However, radiological evidence of cochlear agenesis or an absence of cochlear nerves is a contraindication for cochlear implantation.

Postauricular skin incision was designed in order to access the mastoid process. A bony recess is drilled in the temporal bone to accommodate the receiver. The very thin skull bone thickness in young children (often 2-3 mm) makes this part of the surgery difficult. The cochlea was approached through the mastoid cavity and facial recess, created surgically after performing mastoidectomy and posterior tympanometry. During the process, special attention has to be paid to the facial nerve, which is located very closely to the posterior tympanometry site. An antero-inferior small fenestration leading to the basal turn of the cochlea was then created in the round window niche for the insertion of the electrode array to the scala tympani (Figure 1).

The first mapping of the electrodes usually took place 4 to 6 weeks after the operation. The purpose of the mapping was to set the electrical current level at each electrode for stimulation of the auditory nerves at the most comfortable and just noticeable loudness levels. Once these levels were identified and stabilised, the implanted children could begin auditory and speech training from the speech

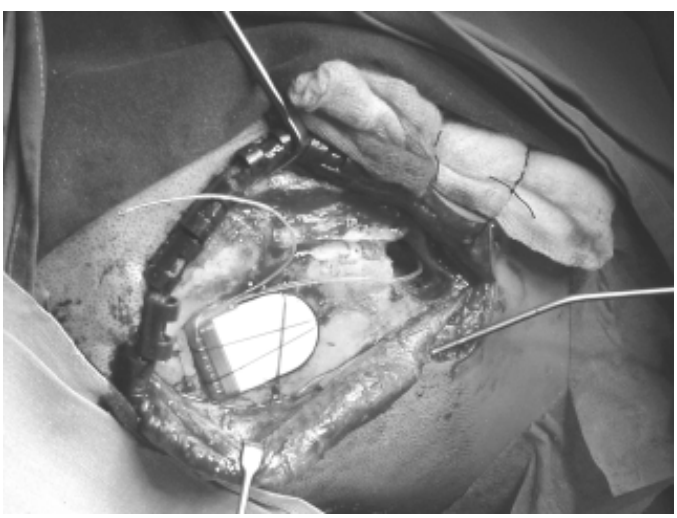

Figure 1. Operative photo showing the receiver anchored in the skull bone recess, the active electrode in the mastoid cavity and the ground electrode.

therapist. Different development stages of the implanted children required different training material and skills. Once the expected level of auditory and speech perception performance was reached, the children returned at different time intervals to continue their evaluation.

Results

None of the 14 cochlear-implanted children had complications during the surgery and all recovered uneventfully. The 3 children with deformed cochleas had reduced cochlear turns and they received partial insertion of electrode arrays. The child who had a large vestibular aqueduct did not show significant vestibular problems and was well and about the day after the operation. Table 2 shows the various CI devices used by the children.

Table 2. Brands of cochlear implant systems used by the 14 implanted children

Cochlear implant system	Number used
MED-EL	7
Nucleus 24	4
Clarion	2
Nucleus 22	1

The unaided pure tone thresholds, averaged at 500 Hz, 1K Hz and 2K Hz, in the left and right ear were, 112 dB HL and 114 dB HL, respectively. Figure 2 shows the aided functional hearing thresholds across the speech frequencies from preoperative binaural hearing aids and postoperative CIs. All the aided functional thresholds from CIs fell within the spectrum for all frequencies while binaural hearing aids could not.

Table 3 shows the auditory and speech perception performance before and 1 year after cochlear-implantation. After 1 year of intensive rehabilitation, the implanted children obtained significantly better auditory and speech perception scores than preoperative results.

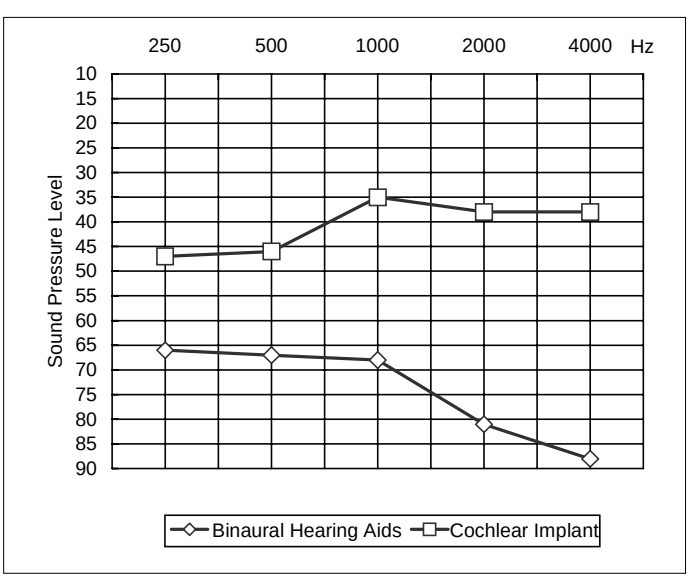

Figure 2. Aided functional hearing thresholds across speech frequencies from preoperative binaural hearing aids and postoperative cochlear implants.

Table 3. Auditory and speech perception performance before and one year after operation

Auditory and speech tests	Implantation		*P*-value
	Before (%)	After 1 year (%)	
Response to environmental sounds	45	100	<0.05
Response to voice	45	100	<0.05
Identification of environmental sound	30	85	<0.05
Identification of own name	40	95	<0.05
Discrimination between long/short sounds	35	95	<0.05
Discrimination between loud/quiet sounds	45	95	<0.05
Discrimination of Ling's 5 sounds	25	65	<0.05
Discrimination between tones	20	75	<0.05
Story comprehension	0	60	<0.05

Discussion

The overall results from the young cochlear-implanted children were encouraging. After implantation, the implant-aided functional thresholds were ameliorated to the mild hearing loss range with which they could detect all frequencies of speech sounds within the speech spectrum. These results could not be achieved even with most powerful binaural hearing aids. Improved audition is the first step of rehabilitation for normal speech and language development.

A distinction has to be made between 'chronological' and 'hearing' age. If a child receives cochlear implantation at 2 years old, his/her hearing age is '0-year old'. After one year of training, although the chronological age will then be 3, the hearing age would only be 1-year-old. Evaluations of the auditory and speech perception skills should therefore take into account the hearing age instead of the chronological age. The ability to respond to environmental sounds and voice indicated that the young implanted children could detect and were aware of different sounds. In the identification of environmental sounds and their own name, the children showed not only that they could detect but also the ability to distinguish and differentiate between different sounds. Long/short and loud/soft sounds are the supra-segmental aspects of speech. The young implanted children achieved higher scores in these tests. This indicated that they were equipped with the basic auditory skills to meet the demand of future linguistc development. The Ling's 5 sounds has five phonemes (ah, ee, oo, sh and s). Good auditory and speech perception skills are required to discriminate the different formant frequencies among these sounds. This group of implanted children could detect, discriminate and comprehend the differences between the five phonemes. Cantonese has six lexical tones that differ from each other by the variation patterns of the vocal cord. Good temporal and spectral auditory abilities are required to discriminate the different tones. The children in this study also obtained higher scores after receiving the implant compared with the preoperative results. Story comprehension stresses sophisticated linguistic and cognitive skills. The higher postoperative scores achieved by these children showed that they were able to comprehend the sequence and semantics of running speech in daily life.

Early cochlear implantation is extremely important in children who do not benefit from conventional hearing aids. Studies had shown that implantation received early in life, could evoke auditory potentials that are similar to their normal hearing counterparts. These results reflected the plasticity of the

central auditory pathway, and which was minimally affected through early acoustic stimulation.[16,17] The adverse effects of complete auditory deprivation are not reversible even after prolonged period of auditory input.[18,19] The results of this study indicate that cochlear implantation can benefit congenitally deafened young Chinese children. The CI devices used in this study are also effective in processing the unique properties of the Cantonese tones. Early implantation should be considered in light of the disruption to the child's linguistic, cognitive and emotional development. As professionals working for hearing-impaired children, we advocate the motif - 'Early detection and early rehabilitation'.

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Newborn Hearing Screening

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Introduction

Significant Hearing Impairment (HI) is an important handicap affecting 1 to 3 per 1000 live births and 2 to 4 per 100 neonatal intensive care infants.[1-4] Based on an annual birth rate of 55,000 in Hong Kong, about 165 infants were born with significant HI annually.

Children with HI are deprived of an important source of sensorial input. Despite recent advances in hearing aid technology, improved educational techniques and intensive intervention services available to the deaf children, there has been little advancement in the academic performance of this group of children.[5,6] In the study by Yoshinago-Itano, the age of identification was the only significant variable identified to affect the development of language skill.[7] Children whose hearing losses were identified by 6 months of age demonstrated significantly better language scores than children identified after 6 months. Hence the issue of newborn hearing screening had received a lot of attention both locally and internationally. Over the past years, different models of newborn hearing screening programs have been developed in United Kingdom, European countries and United States.

Hearing Screening in Hong Kong

Hong Kong, like many other countries, did realize the importance of hearing screening. For the past decades, hearing screening had been performed as part of the comprehensive observation scheme by using the behavioral distraction test. The test was performed at 6-9 months of age for children attending the Maternal and Children Health Centre (MCHC). This test has a poor sensitivity and low specificity. Infant who fails the first test usually needs to be re-tested several weeks later. Many of the hearing impaired children are either not detected or if detected, not soon enough. Hence, with this program, the usual age at diagnosis especially for those without risk factors is at 2 years and most infants received treatment and rehabilitation after 2 years.

High Risk Newborn Hearing Screening

The America Academy of Paediatrics Joint Committee on Infants Hearing 1994 Position Statement recommended to maintain a role for high risk indicators associated with sensorineural and/or conductive hearing loss in newborns.[8] For infants with risk factors, most Hospital Authority birthing hospitals performed some form of hearing test, usually by brainstem auditory evoked potential (BAEP) or other screening device e.g. Otoacoustic Emission (OAE) tests. However, the screening programs were usually not well coordinated with a formal tracking and follow up system. Experience from studies also showed that high risk screening could only identify less than 50% of all cases of congenital hearing impairment,[9,10] it appears that a universal newborn hearing screening is a more logical approach aiming at identifying most, if not all babies with congenital hearing loss.

Common Devices Used for Universal Newborn Hearing Screening

The conventional brainstem auditory evoked potential (BAEP) was the most reliable method and the gold standard for evaluating the peripheral auditory function of newborn.[11,12] However, this is mainly used as a confirmatory test. At present, the choice of device for newborn hearing screening is between Otoacoustic emission (OAE) and automated brainstem response (ABR), or some combination of these two.

Two forms of OAE technology have been used for newborn hearing screening. These were known as 'transient or click-evoked Otoacoustic Emissions' (TAOAE) and 'Distortion product Otoacoustic Emissions' (DPOAE). Both OAEs have the advantages of being a simple and quick test, with minimal disturbance to the babies. The false positive rate was high when the tests is performed in the first few days of life because of presence of debris in the external ear canal and middle ear collection in newborn babies.[13,14] Furthermore, the device can only test up to the level of cochlea; it does not provide information on retrocochlear pathology in the auditory nerve, brain stem and auditory cortex. Hence babies with neurological disorders associated retrocochlear pathology may be missed by OAE testing.

The Automated Auditory brainstem response (AABR) is a simplified version of Auditory Brainstem Response (ABR). This is a quick and simple test. It can be performed reliably even in newborns under 24 hours of life, and hence is best for a hospital based screening program. The refer rate for further diagnostic test was also lower than the OAE response. However, the currently available AABR machine and the consumables are much more expensive than the OAE device.

The recent introductions of equipment that incorporate the OAE and ABR functions provide an additional option for the choice of screening methods.

Universal Newborn Hearing Screening

The year 2000 Joint Committee on Infant Hearing (JCIH) position statements endorses early detection of, and intervention for infants with hearing loss through integrated, interdisciplinary state and national systems of universal newborn hearing screening, evaluation, and family centered intervention. The Academy also recommended five essential elements to an effective universal newborn hearing screening program (UNHSP) including screening, tracking and follow up, identification, intervention and evaluation.[15,16] Hence, regardless of which device or program we adopted, it is important to emphasis that newborn screening is just the beginning of dealing with the problem of hearing loss in children. The screening should be followed by evaluation, diagnostic work up and a multidisciplinary intervention and rehabilitation program. It is a multidisciplinary program involving paediatricians, audiologist, ENT surgeon, nurses, speech therapist, community health care workers and education specialist.

As for Hong Kong, while waiting for more solid evidence on the cost effectiveness of newborn hearing screening program, we should work towards the direction of developing a collaborative, multi-disciplinary, cost effective and sustainable universal newborn hearing screening and early intervention program for our next generation of hearing impaired children.

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Central Auditory Processing Disorders (CAPD)

Louciana LAU, Audiology
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The broad definition of auditory perception or auditory processing encompasses various functions at all levels of the auditory system from the external ear to the cortex. With good peripheral hearing, sound energy is transmitted efficiently from the external ear, via the middle ear to the inner ear. Beyond that, central auditory processing provides us the most important characteristic of human by affording us with a means of verbal communication, i.e., it makes meaning out of auditory signals.

In our daily communication, the mature and normal central auditory system allows us to assign priority to certain sounds, words, and sound/word combinations that are useful to us. It is the brain's function to assign meaning to auditory stimuli which the ears receive. This occurs while the system also suppresses and/or blocks undesirable or unwanted auditory stimuli. This process prevents all sounds from running together which results in a "listening chaos".

There are instances reported where peripheral hearing is normal, but some higher level disorders do exist. In testing peripheral hearing, tests involved are well standardized and relatively straightforward, such as pure tone audiometry, otoacoustic emission and tympanometry. If present, the degree and nature of hearing loss can then be accurately identified. Unlike peripheral hearing, central auditory processing can give professionals challenges for various reasons. The highly complex structure of central auditory nervous system (CANS) and its linkages with more global cognitive and linguistic functions are not completely understood. Its many different functions are not adequately defined. Also, the effects of CANS disorders are often quite subtle, and test results may be highly variable.

In 1995, the American Speech Language Hearing Association (ASHA) task force (1996, American Journal of Audiology, 5(2), 41-54) on central auditory processing consensus development met to define central auditory processing and its disorders. Central auditory processing disorders (CAPD) are not a unitary disease entity, but a description of functional deficits. It can be observed in a variety of clinical populations, including those with known lesions or pathology of the central nervous system, neuro-developmental disorder, or neurological changes resulting from aging process.

The ASHA consensus statement defines central auditory processes as the auditory system mechanisms and processes responsible for the following behavioral phenomena. Central auditory processing disorder (CAPD) is an observed deficiency in one or combinations of them (Table 1).

Table 1. Central auditory processing domains.

Sound localization and lateralization
Auditory discrimination auditory pattern recognition
Temporal aspects of audition
Temporal resolution
Temporal masking
Temporal ordering
Temporal integration
Auditory performance decrements with competing acoustic signals
Auditory performance decrements with degraded acoustic signals

Assessment

I) It is possible that clients with CAPD may also have general dysfunctions. It is not uncommon to have a child who is inattentive and easily distracted or has poor academic performance, apart from having hearing problems in background noise, following oral instructions, having poor listening skills, or having poor auditory association skills. Since the issue of overlap with other disorders remains to be resolved, it is recommended that investigation should be made in psychological, behavioural or learning aspect when concerns arise. Thorough developmental, medical history and careful observation should be obtained in the following areas:

- Auditory processing, language and learning problems that exist in the family
- Speech and language development
- Hearing and auditory behaviour
- Educational progress
- General behaviour and social-emotional development

II) *Tests:*

Basic audiologic evaluation must precede any assessment of central auditory function. Measurements of pure tone audiometry, tympanometry, otoacoustic emission and speech audiometry (in quiet) provide information on peripheral hearing status and that helps to rule out any conductive or sensorineural hearing problem.

Behavioural tests

Battery of tests is essential to locate areas of deficits, for example, temporal ordering tests, binaural interaction tests and degraded speech audiometry, etc. CANS is challenged by reducing the acoustic redundancy of the stimuli and increasing the loading of the system under various testing conditions. It is important to have norms on an adequate sample pool of individuals with appropriate age of whom testing is intended. Validity and reliability information should be available for reference.

Electrophysiological test

The use of electrophysiological tests has gained much interest and popularity in CAPD. They have added an additional dimension to the central auditory test battery and helped to understand the neurophysiological substrata of CAPD.

Auditory brainstem evoked response (ABR) reflects the sequence of activity of the auditory nerve and nerve tracts and nuclei of the ascending auditory pathway, and hence is valuable in the measurement of its integrity.

Late auditory evoked responses, such as N1, P2, P300 and mismatch negativity, are found to be consisted of perceptual and cognitive processes. Responses are generally related to the acoustic features of the stimulus and subjects' preconscious discrimination ability towards them.

Management

There is no absolute agreement about therapy approaches for CAPD. Some basic strategies are as follows:

Perceptual training

- with the belief in neuro-plasticity, training addresses the temporal aspects of audition may help the perception of fast and brief speech signals.

Compensatory techniques

- teaching of auditory skills, for example, phonemic synthesis and speech sound discrimination.
- cognitive training to teach clients to actively monitor or self-regulate their message comprehension skills, and develop new problem-solving and organizational skills.

Management of the environment

- improving room acoustics to reduce noise and reverberation, use of amplification to enhance signal to noise ratio.

Further development and systemization of the assessment and management of CAPD is definitely needed. Interdisciplinary approach is highly preferable to tackle CAPD.

CAPD testing in Hong Kong

Since many language specific CAPD tests are not available in Cantonese, local test tools are rather limited. Development of Cantonese materials has begun with the Cantonese Hearing In Noise Test (CHINT) in which sentences are presented in noise (Wong and Soli, in press). Child Assessment Centres will try out another speech in noise test with spondees as stimuli which is more applicable to younger children. Norm data of some non-linguistic tests will also be collected for local reference, i.e. Pitch Pattern Sequence, Duration Pattern Sequence and Auditory Fusion Test.

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The Role of Child Assessment Centres in the Identification and Assessment of Childhood Hearing Loss in Hong Kong

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A total of seven Child Assessment Centres (CAC) have been set up in Hong Kong. One of them (the Duchess of Kent CAC), under the Hospital Authority, is located in Pokfulam. The other six, under the Department of Health, are located respectively in Central Kowloon, Mongkok (Arran Street), Kwun Tong, Shatin, Tuen Mun and Ha Kwai Chung. The Department of Health will provide an additional CAC in Fanling next year to meet the future service demand.

The role of CAC in the identification and assessment of childhood hearing loss in Hong Kong will be discussed in this paper.

Identification of Hearing Loss by Screening at the Maternal and Child Health Centres

The Maternal and Child Health Centres (MCHC), under the Department of Health, have provided mass developmental screening and medical check up, known as the Comprehensive Observation Scheme,[1] for Hong Kong children since 1978. Under this scheme, the child will be seen at the age of ten weeks, nine months and three years for screening of vision, hearing, cognition, and speech and motor development. When impairment or delay is suspected, a referral will be made to the CAC for further comprehensive assessment.

In the past, the hearing screening used to include a behavioural distraction test at the age of ten weeks and nine months, and a behavioural word discrimination test at the age of three years. If the child failed the test on the first occasion, he would be re-tested within a few weeks. If failed again, he would be referred to the CAC for further assessment. As there has been growing evidence that the sensitivity of the distraction test used at very young age would be unacceptably low,[2] the hearing screening at the age of ten weeks was scrapped in early 2000. In replacement, the parents are now given a checklist[3] of some general signs of hearing loss suspicion to look at. Examples of the checklist items include: "Will your child quieten or smile to the sound of your voice even when he cannot see you at the age of 4 months?", and "Will your child turn immediately to your voice across the room if he is not too occupied with other things?"

While the behavioural tests scheduled for the age of nine months and three years remain, an additional assessment for high-risk indicators is now in operation. These high-risk indicators are generally in line with those adopted by the Joint Committee on Infant Hearing.[4,5] Children known to have these high-risk factors will be referred to the CAC for hearing screening and assessment.

Parents may also consult the MCHC at any time when they suspect their child of hearing problems from birth to five years old.

Hearing Assessment at the CAC

A multidisciplinary approach is adopted by the CAC. The team includes paediatricians, nurses, clinical psychologists, speech therapists, physiotherapists, occupational therapists, medical social workers, optometrists (or orthoptists) and audiologists. After receiving the referral from the MCHC, the team will provide a comprehensive assessment of the child's physical, cognitive, psychological and social aspects of development together with his vision and hearing. If appropriate, the team will provide interim therapy for the child, support and counseling for his parents, and co-ordination of rehabilitation services (such as training, education and care) to meet the individual needs of him and his family.

The audiologists at the CAC will in particular help to assess, confirm and make diagnosis of the type of hearing loss present in terms of its nature, severity and site of lesion. A battery of hearing tests, both behavioural and physiological, will be used in assessment, and the selection of that is dependent on the mental age and ability of the child. Referrals to the ENT doctors for further consultation and to the Education Department for amplification will be made as soon as hearing impairment is confirmed. Habilitation programs with appropriate education placement will start soon as well. For children diagnosed with bilateral profound sensorineural hearing loss, cochlear implantation may be considered.

Targeted population at the CAC

Apart from those who fail the hearing screening at the MCHC, infants and children with high-risk indicators for congenital hearing impairment and indicators for progressive or delayed-onset sensorineural hearing loss and acquired conductive hearing loss[5] altogether form the major caseload of the CAC. These infants and children with high-risk indicators require regular hearing screening and assessment at the CAC.

The high-risk indicators for congenital hearing impairment include: (a) an illness requiring admission to a neonatal intensive care unit, (b) stigmata associated with a syndrome known to include a hearing loss, (c) family history of permanent childhood sensorineural hearing loss, (d) craniofacial anomalies and (e) in-utero infection such as cytomegalovirus or rubella.

In addition to the above, the following may render the children a high risk of developing progressive or delayed-onset sensorineural hearing loss, and acquired conductive hearing loss: (a) speech and language delay, (b) postnatal infections associated with sensorineural hearing loss including bacterial meningitis, (c) syndromes associated with progressive hearing loss such as neurofibromatosis, (d) neurodegenerative disorders such as Hunter syndrome, (e) head trauma, (f) neonatal indicators such as hyperbilirubinemia at a serum level requiring exchange transfusion and persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and (g) recurrent or persistent otitis media with effusion (OME).

Among these high-risk indicators, children with recurrent and persistent OME are highly prevalent in the CAC population. It is estimated that each year up to one third of the children diagnosed with hearing impairment in the Duchess of Kent CAC, for instance, suffer from conductive hearing loss associated with OME.

The impact of OME and its associated conductive hearing loss to the intellectual development of affected individuals is controversial. Roush[6] summarizes that the findings about the effects of OME on speech, language, and learning have been mixed. Some studies have indicated a relationship between persistent OME and reduced language skills or school performance. Others have found no significant association or one that is related only indirectly. But yet, in the end, he concludes it is widely recognized that persistent OME has potentially detrimental long-term consequences, especially for children already experiencing communicative disorders related to learning disabilities or developmental delays.

Identification of Hearing Loss by Screening at the Student Health Service Centres

According to the data published in the Hong Kong Rehabilitation Program Plan,[1] slightly over 90% of all newborns in Hong Kong have attended the Comprehensive Observation Scheme run by the MCHC in 1998. In other words, if this figure also holds true for other years, about 10% of the newborns each year in Hong Kong will never go through hearing screening during their infancy and early childhood. In addition, each year there may also be another group of children moving in to Hong Kong from the Mainland China and other countries which do not have neonatal or early childhood hearing screening programs. The hearing screening service provided by the Student Health Services Centres (SHSC), under the Department of Health, may thus act as the last defensive measure for identification of childhood hearing loss in Hong Kong.

The SHSC provide a comprehensive health program for all primary and secondary day school students in Hong Kong.[1] The program includes health education and screening of hearing, vision, spinal curvature, psycho-social health and growth status. Among these, hearing screening is compulsory for all primary one students and those who attend the SHSC for the first time. Hearing screening can also be arranged at any time upon request by the students.

For students suspected of hearing impairment, they will be followed up at a Special Assessment Centre for further assessment and diagnosis. It has been reported that 1.64%, adjusted for under ascertainment, of the primary one students screened from 2000 to 2001 were found to have hearing impairment (hearing loss $\geqq$ 26dBHL), of which 56% were sensorineural in nature.[7]

The Role of CAC and Future Development

A cost-effective system of early identification of hearing impairment in infants and children together with appropriate timely intervention services requires the collaborative efforts of many health professionals and educators.

By closely working together, the CAC serve to assess, confirm and diagnose the hearing loss for infants and children who fail the hearing screening at the MCHC. Regular hearing screening and assessment are also provided by the CAC for children presenting with high-risk indicators of congenital hearing impairment and indicators for progressive or delayed-onset sensorineural hearing loss and acquired conductive hearing loss. The CAC also serve to co-ordinate the habilitation services.

The SHSC, on the other hand, may help to capture those children with congenital or acquired hearing loss who have by whatever reasons missed the hearing screening at the MCHC, and who move in to Hong Kong later in their childhood.

Identification of childhood hearing loss sometimes may also take place at the ENT clinics and private hearing test centres, especially for hearing loss associated with ear diseases. At present, there is no effective tracking system established for exchange of medical and audiological information in Hong Kong. The development of such a system in future among all the service providers, both public and private, may worth be explored.

There is growing evidence that early identification of hearing loss followed by early intervention results in better long-term developmental outcomes for children and their families.[8] According to the recommendations made by the Joint Committee on Infant Hearing in 1994, all infants with hearing loss should be identified before 3 months of age and should receive intervention by 6 months of age.[9] Certain hospitals, under the Hospital Authority, and the MCHC in Hong Kong are at the moment conducting research studies on the feasibility of screening newborns by otoacoustic emissions and automated auditory brainstem responses. The outcome of these studies may give new direction to the existing system of childhood hearing loss identification and assessment.

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Services Provided for Hearing Impaired Children by the Education Department

Jenny CY CHAN
Audiological Services Section, Education Department

Hearing loss is typically a silent and hidden impairment: especially so in children. Hearing impairment itself does not carry any visible abnormal sign, and the affected children cannot tell us that they are not hearing well. However, if undetected and untreated, hearing impairment can lead to delayed or impaired speech and language development. Language is the key to the door by which we express our thoughts, needs, and feelings and by which we receive and comprehend the thoughts, needs, and feelings from others.[1] Children with hearing impairment may therefore be confronted with a life of language difficulties, educational struggles, underachievement, social and emotional problems.[2]

Role of the Education Department

To ensure that all aspects of children's hearing and learning are maximized, the Education Department (ED) started to support preschool-age and school-age children with hearing impairment as early as 1960's. Its Audiological Services Section (AS Section) provides comprehensive services, which include prevention, assessment, recommendation for amplification and (re)habilitation, providing follow-up and monitoring, parents guidance, review and recommendation for school placement.

Referral to Services of AS Section

It is through early detection and early intervention that many of the negative effects of hearing impairment on a child's development can be prevented or lessened. Close collaboration between ASS and different medical settings are therefore always maintained to streamline this two-fold process. ENTs in various hospitals, paediatricians in Child Assessment Centres and private ENTs make up a large portion of referrals to AS Section each year. ED started universal audiological screening for P.1 children in 1968 to advocate hearing conservation and to identify any hearing problem in early school life. In the year 2000, this service was handed over to the Student Health Service under the Department of Health and screened children who need follow-up are referred back to AS Section.

School personnel and parents are also in a good position to identify their children's hearing difficulties. Direct referrals from schools and from parents to AS Section are playing an important role in helping these children.

Audiological Assessment and Diagnosis

In AS Section, audiological assessment is arranged for individual children who are referred. About 3800 children were audiologically assessed in the academic year 2001-2002. Among them, about 2000 are active cases that need hearing device recommendation and follow-up (re)habilitation programs.

Amplification and (re)habilitation

For children with identified hearing impairment, no other device is more fundamental to their education and ability to learn speech than the properly fitted hearing aids.[3] To satisfy this basic need of hearing and education, ED issues a free hearing aid and custom-made earmoulds to each child assessed to be in need of them. Based on individual educational needs, FM system will also be loaned to them. Children in need of other hearing devices (e.g. Cochlear implant) are referred to the appropriate settings.

Apart from providing hearing devices as the "hardware", "software" like parent guidance, design of (re)habilitation program and review of school placement also need to be focused. AS Section set up two teams: School Support and Educational Services (Formerly known as Pre-school Advisory and Training Service, and Peripatetic Advisory Service) (SSES) and Visiting Audiological Service (VAS) to serve these purposes. The former takes care of pre-school age children and school age children studying in ordinary schools, while the latter looks after children studying in special schools and those with additional disabilities.

School Support and Educational Services (SSES)

Training of hearing impaired children should start as early as they are identified but not wait until they reach school age. This is evident by the fact that the most important period for language and speech development is the first 3 years of life.[4] Recent studies even point to the first 6 months of life as the significant period of language development.[5,6] With this belief in mind, SSES provides individual and group speech and auditory training service, parent guidance, advisory service to pre-school teachers and guidance on proper school placement to hearing impaired pre-schoolers.

According to the Hong Kong Rehabilitation Plan 1994,[7] school age hearing impaired children should receive education according to their special needs caused by their different degree of disabilities. As far as possible, hearing impaired children are encouraged to integrate and receive education in ordinary schools. To guarantee that they are not "dumped and forgotten" in the regular classroom, support services must be provided. SSES takes up the roles of advising school personnel, discussing remedial educational service and coordinating different services.

Visiting Audiological Service (VAS)

For those who need more intensive training in language and communication skills, they are placed in one of the four special schools for hearing impaired children, or the special class for hearing impaired children. VAS team is responsible to keep close contact with these schools and visit the children regularly. It aims at coordinating with the schools in maximizing the use of children's residual hearing.

In case other disabilities occur in combination with hearing impairment, "additional learning problems" will occur which significantly add to the complexity of educating these children.[3] In view of this, VAS service also serves hearing impaired children with additional disabilities and provides support to the relevant special schools, special child care centres and early education and training centres.

Professional Support

AS Section is soundly staffed with professionals like audiologists, technicians, inspectors and teachers for the deaf. With their joint effort, we hope all children with hearing loss will achieve their full potentials and attain the best future possible.

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News of Society

The International Conference on Developmental Dyslexia in Children using the Chinese Language - fMRI and Advocacy
26 - 28 October 2002

The Council of the HKCNDP and the organizers welcome the invited speakers from overseas at a dinner.

Government officials, concerned parties and legislators exchanged their views on dyslexia in a forum held on 28 October 2002.

Mr and Mrs Emerson Dickman, Professor Leong Che Kan, Legislative Councillor Mr Leung Yiu Chung and Mr Thomas West met the press. Dr Chan Chok Wan, the President of HKCNDP, presided.

Professor CK Leong (on the left) is the mastermind behind this International Conference.

Mr Emerson Dickman III, Esq. received a token of gratitude from Dr CW Chan during the closing evening lecture. Mr Dickman is the Secretary of the Board of Directors of the International Dyslexia Association.

Welcome New Member

A very warm welcome to the following new members:

Associate Members:	Dr Yau Kin Cheong Eric
Affiliate Members:	Ms Christina FK Wong
	Ms Clara SC Lee

Photographs and Memorabilia of the Past Events

Our Society has entered her eighth year. We have held a large number of activities in the past years and many of them are very memorable to a lot of members. Do you have any photographs or memorabilia that can bring back the fond memory of these events? If you do, would you be so kind to share them with your fellow members? Our Society would like to build a digital collection of these photos. Your photos can be sent to our Vice President Dr Lau Wai Hung at Parkes Medical Centre, 2/F Parkes Commercial Centre, Tsim Sha Tsui, Kowloon. The photos will be scanned to digital format and returned to you. As an alternative you can scan your pictures into digital format and send to Dr Lau via e-mail at hkcndp@hongkong.com. Your kind contribution will be very highly appreciated.

Manual of Child Neurology and Manual of Developmental Paediatrics

The Society has a number of copies of the two manuals. They are good reference for clinicians and allied health professionals. As the Society has recently received requests from members asking for these manuals, the Council has resolved to give them away to all members who are interested free of charge. You can write to the Honorary Secretary at Department of Paediatrics, Queen Elizabeth Hospital, 30 Gascoigne Road, Kowloon or by e-mail at hkcndp@hongkong.com stating your requests for these manuals. We will write to you about the arrangements of collecting them in person. The give-away will continue as long as stock lasts.

Working Party on Cerebral Palsy

Dr Sophelia Chan, one of the convenors of the Working Party, will give a presentation on the surveillance project on the community wide prevalence of cerebral palsy in the upcoming Neuro-developmental Conference in February 2003. The surveillance will collect information at the schools for children with physical and intellectual disabilities. At the presentation Dr Chan will discuss the details of data collection and the rationale behind her surveillance. Please mark your calendar for that special conference on 16 February 2003.

Joint Meeting at Macau 2003

At the kind invitation of the paediatricians in Macau, our Society will organize a joint meeting of general paediatricians, developmental paediatricians and child neurologists of Hong Kong, Macau and Mainland China. The meeting is scheduled for 11 - 12 October 2003. It will be held at Macau.

Our Society will be responsible for arranging the scientific program of the meeting. An entourage of 30 representatives from Hong Kong will be formed. If you are interested in delivering a topic presentation on certain important issues in child neurology or developmental paediatrics, please kindly contact the Honorary Secretary.

Annual Subscription

The annual subscription of all members falls due on 1st January 2003. The amount payable is as follows:

Affiliate Members:	HK$100
Associate Members:	HK$200
Full Members:	HK$300

Please send a cheque payable to "The Hong Kong Society of Child Neurology and Developmental Paediatrics" of the appropriate amount to Dr Sharon Cherk, Honorary Treasurer, at Department of Paediatrics, Kwong Wah Hospital, Kowloon. Do send in the cheque today!

Annual Scientific Meeting 2003

" Neurological Intensive Care"

This coming September 2003, Queen Elizabeth Hospital

Watch for future Announcements