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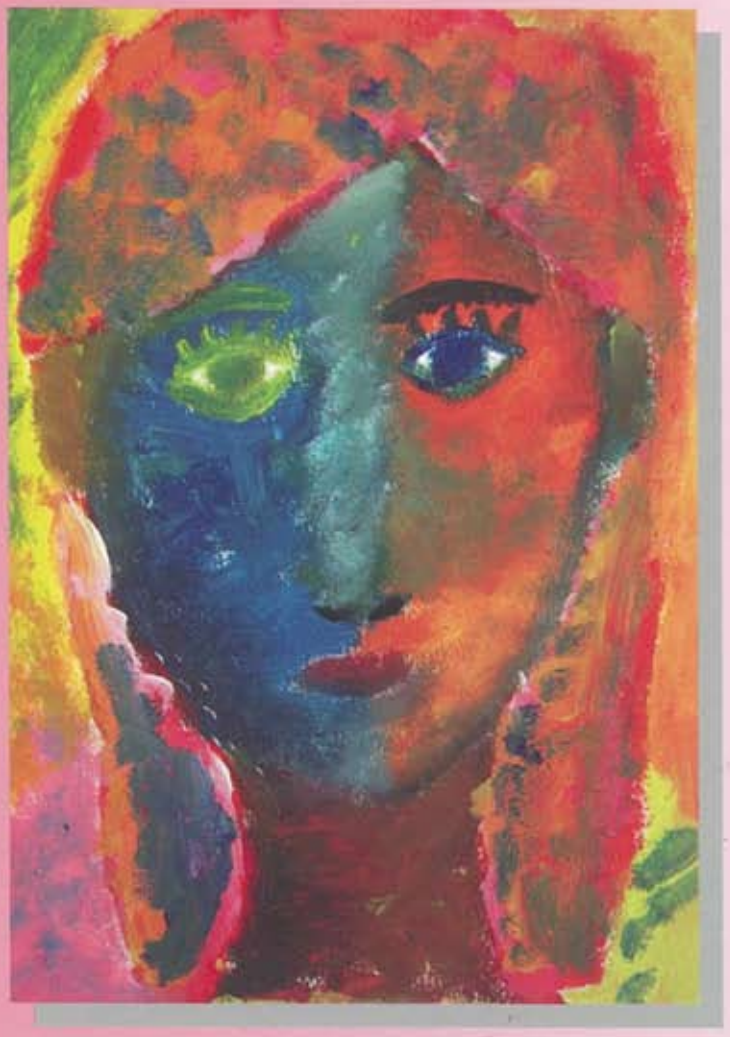
BRAINCHILD

The Official Publication of HKCNDP
Special Issue on Hearing Impairment



香港兒童腦科及體智發展學會
The Hong Kong Society of Child Neurology and Developmental Paediatrics

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SPECIAL ISSUE ON HEARING IMPAIRMENT

CONTENTS

On the Cover
Almost Matisse: a Portrait by a 16 year old girl

The Hong Kong Society of Child Neurology & Developmental Paediatrics

BRAINCHILD – SEPTEMBER 2007 ISSUE

"Hearing Impairment"

Message from the President

The current issue of Brainchild is devoted to Hearing Impairment (HI). This is consequent to the successful Joint Meeting on Developmental Paediatrics in Beijing in October 2005 whereby experts from Hong Kong, Macau and the Mainland of China met and exchanged expertise and knowledge on modern management of children with Hearing Impairment as well as most up-to-date concepts and skills in the rehabilitation and education of children with this disability. In order to share with readers in Hong Kong, the Editorial Board invited local experts to share with us on the Hong Kong scene. Thanks to the effort of Dr. Sylvia Doo and Dr. Catherine Lam of the Child Assessment Services of the Department of Health, we are able to present several outstanding papers covering the subjects of *Auditory Neuropathy – what do we know so far?* (Sylvia Doo), *Balance and Motor Skills in Children with Hearing Impairment* (Candice Poon), R*ecent Advances in Cochlear Implants* (Eddie C M Wong and H C Yu), *Serving hearing impaired children in Hong Kong* (Catherine Lam) and *Universal Infant Hearing Screening and Diagnosis in Hong Kong – Experience at Child Assessment Service, Department of Health, Hong Kong* (Sylvia Doo and Catherine Lam). The papers explore different facets of the problem in Hong Kong and throws light for future direction in the management of our children with HI. We indeed have excelled ourselves in the issue but there is still room for improvement in the care of our children with HI.

The Hong Kong Society of Child Neurology and Developmental Paediatrics (HKCNDP) is pleased to witness successful review of the Rehabilitation Programme Plan (RPP) under the Rehabilitation Advisory Council (RAC) of Hong Kong. We would like to congratulate members of the RPP Committee for their hard work over the past two years. We are especially impressed by some enlightened members of the Committee together with the great leadership of Dr. York Chow, Secretary for Health and Food of the Hong Kong SAR Government, in finally including Specific Learning Disabilities (SLD) and Attention Deficit/Hyperactivity Orders (ADHD) into the category of disabilities which enables our children with these disorders henceforth to have official recognition and resources support at the family, school and the community. On behalf of our children we would like to salute all those who has dedicated to the success of this noble mission!

The RPP Report 2007 discussed the way forward with short and long-term deliverables and objectives outlined for key areas, with specific measures proposed for various objectives. This is different from previous RPP's where service needs and provisions for the period covered were documented and enumerated. The purpose for this new approach was supposedly to

allow flexibility of development over the period covered (of say 5 years), rather than pledge precise funds and provisions for each area. It is good for policy and dynamic in principles but is extremely difficult to perform end-of-term efficacy assessment. It thus follows that the principles as proposed should be accompanied by practical guidelines (or code of practice) so that objective parameters are available to monitor outcome performance of the new concept, which is one of the vital components in modern management.

Our Society takes pride in presenting the Position Paper on SLD in February 2006 and again the Position Paper on ADHD on 5th July 2007 as fulfillment of our pledge to RPP and as our humble contribution to the community. These Position Papers are prepared by Hong Kong professionals from different disciplines and transectoral domains based on local literature, data and information with the key targets to assist the HKSAR government, policy makers, professionals, service providers and the general publics in the understanding of the nature of the conditions, their etiologic bases, mode of inherence, clinical features, early identification, diagnosis and intervention, cutting edge management regimes, current confronting problems and their solutions in Hong Kong. We sincerely hope that both of these Position Papers will serve to achieve their goals and to attain their roles and missions.

In order to consolidate our work in ADHD, the Society's Working Party on ADHD met on 2nd August 2007 and established three Committees to further our mission on the subject: namely, a **Scientific Committee** with membership of Luk Siu Luen (Chair), Patrick Leung, Tatia Lee, Shiu Ling Po, Hung Se Fong, Daniel Shek, Stephenie Liu, Lee Chi Chiu and others; **Professional Development Committee** with Joseph Lau (Chair), Patrick Leung, Shiu Ling Po, Cheng Pui Wan, Hung Se Fong, Daniel Shek, Catherine Lam, Tsui Kwing Wan, Sin Kuen Fung and others; and **Advocacy Committee** consisting of Chan Chok Wan (Chair), Daniel Shek, Ho Lok Sang, Catherine Lam, Fernando Cheung Chiu Hung, Leung Yiu Chung, Kwok Ka Ki and others. These are all local experts on the subject. With such a robust infrastructure and powerful manpower input, we are ready to strive for the best services for our children with ADHD in Hong Kong. Our deepest appreciation to all professionals for their dedications towards this worthwhile endeavour!

As for our work on SLD/Dyslexia, the Society will host a Summit on SLD 2007 on 19th November 2007 at the Queen Elizabeth Hospital, Hong Kong with the goal to review work achievement on SLD/Dyslexia over the past ten years in Hong Kong, study current situations, identify confronting challenges in management, collect opinions from stakeholders, and plan the way ahead with strategic recommendations. We have already invited two world experts on the subject Professor Doris Johnson from Chicago and Professor Leong Che Kan from Canada as keynote speakers who together with local experts and stakeholders on the subject comprising of Legislators and Policy Makers, Bureau Heads and Government Officials, Academicians (tertiary Institutions), EOC, Ombudsman, ASLD (parent organization), Hong Kong Jockey Club Charitable Foundation and NGO's currently providing services to our children with SLD will promise all participants a fruitful meeting in Hong Kong. Please stay tune!

The Annual Scientific Meeting 2007 for our Society focuses on "Mitochondrial Diseases" with Professor Ingrid Tein from the Toronto Sick Children Hospital as Course Director. Professor Tein is the world expert on the subject and we are anticipating another good scientific platform for local professionals specialized in neurosciences. Thanks to the effort of Dr. Tsui Kwing Wan and Dr. Catherine Lam, the scientific programme is ready and we strongly recommend all colleagues to register early so as not to miss this good learning opportunity.

The Joint Meeting on Developmental Paediatrics has been the annual scientific activities of developmental paediatricians and allied health professionals in the Chinese speaking regions from Hong Kong, Macau, Taiwan, the Mainland and Singapore since our Inaugural Meeting on Developmental Paediatrics (DP) in 2003 in Macau. This year we are pleased to host the Meeting in Chengdu, Sichuan on the timely subject of "Update on Childhood Visual Impairment" on 11-14th October 2007. Members and professionals are welcome to join us at this upcoming meeting of minds, which is now one of the popular events for professionals engaged in the subspecialty of Developmental Paediatrics within our region.

HKCNDP has been commended as one of the most productive subspecialty societies with diversified range of acitivities over the years in Hong Kong. Judging from the above-mentioned programmes, we are expecting another year of success for 2007. On behalf of the Society, I would like to thank all colleagues once again for your ever-unfailing patronage to our Society and I wish you all reading pleasure!

Dr. Chan Chok Wan
Editor-in-Chief, Brainchild, Official publication of HKCNDP
President, The HK Society of Child Neurology & Developmental Paediatrics (HKCNDP)

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Serving Hearing Impaired Children in Hong Kong

Catherine CC LAM
Child Assessment Service, Department of Health

The understanding of hearing impairment in adults began over a century ago with the clearer appreciation of the ear's anatomy, physiology, and related pathologies, instead of focusing on the deafness itself. With the introduction of antibiotics and otological surgery, disabling hearing loss was markedly reduced. The physiology of hearing was extensively studied in past decades, when efforts in managing deafness turned to reconstruction of hearing mechanisms, including repair of tympanic membrane perforations, treatment of otosclerosis, myringotomy and grommet tube insertion for middle ear effusion, and prevention of chronic ear disease. The widespread use of vaccines against common viral childhood illnesses, including the introduction of vaccination against rubella virus, removed many viral etiologies as a cause of hearing impairment. The inclusion of neurosurgical approaches to ear surgery laid the groundwork for cochlear implant. Early identification of hearing impairment in infants, especially those with high risk of sensori-neural hearing loss was highlighted by the Joint Commission on Infant Hearing (Joint Commission on Infant Hearing, 1990, 2000). With early identification, evidence based and timely intervention for managing the hearing loss, supporting speech, language, learning and psychosocial development of affected children became expected practices in developed societies.

In Hong Kong, the prevalence of all grades of loss according to the Census & Statistical Department in 2000 was 0.2% under 15 years, and the Central Registry for Rehabilitation 2004 (voluntary enrollment) for moderate hearing loss or worse was 0.06% under 15 years. While the incidence for bilateral permanent childhood hearing impairment of moderate or greater degrees has been shown to be around 1.2 per 1,000 live births (Davis A et al. 1997), population figures for Hong Kong are not yet available.

Hearing impairment has been included as a category of disability from the inception of Hong Kong's first policy paper on rehabilitation in 1977. The Rehabilitation Policy in Hong Kong aims to promote and provide comprehensive measures to prevent disability, through enabling persons with significant biological and functional limitations to develop to their fullest physical, mental and social capabilities. Contributing to these efforts are public and community services, with prevention, early identification, medical, education and social rehabilitation programmes. The Department of Health provides genetic counseling for hereditary conditions with hearing impairment, universal new born screening and hearing surveillance through the family health and student health services and diagnostic evaluations at the child assessment centres. The hospital authority ENT departments provide medical treatment, audiological evaluations and cochlear implants for indicated clients, while the Education Bureau provides hearing aid prescription, and special education support in special and mainstream schools. Non-government organizations and community centres also provide the range of services from diagnosis to rehabilitation.

It is clear that children with significant hearing impairment face a complex range of medical, developmental, learning and social special needs. These are provided by the wide range of services and the multidisciplinary teams therein. In order that families with these children may access necessary advice and services effectively, strong collaboration and teamwork are essential. Parent groups are invaluable for providing support to new members and each other as their children grow and new needs emerge.

Finally, the importance of fostering language development in children with significant hearing impairment from the earliest age possible cannot be over-emphasized. With progress in early identification and intervention, including cochlear implant, it has now become possible for these children to look forward to reasonable hearing after treatment. However, language development in young children comes with critical periods which may have passed before adequate hearing amplification is achieved, the consequence of which are serious for language, learning, social development. Natural signing is a complete language (unlike gestures that parents may use with hearing impaired children), through which babies and young children with no or limited hearing may develop language networks in the brain within the critical growth period, with all of the vocabulary, grammar and pragmatic elements. Coupled with simultaneous speech and language input, communication ability and intent in these children will be strengthened and ensured. As sounds become more available to them after auditory rehabilitation, verbal language will gradually and naturally take over, by which time, these children would have enjoyed and acquired the richness of language potential as in all other children with adequate language exposure. This concept is still new to and against the practice of many, who in past decades did not condone signing in therapy and schools, for fear that hearing impaired children would "take the easy way out and not speak in future". Despite strong efforts with oralism in education, learning and academic standards did not improve. Today, the number of Hong Kong's hearing impaired students with otherwise solid intellectual abilities who could enter local universities is abysmal. In fact, most were unable to even continue with matriculation studies locally because of weak language subject performances. Hopefully clearer understanding of the neuro-linguistic nature of child language development by all concerned will allow the subject of language acquisition in hearing impaired children to be thoroughly discussed, to inform practice for the benefit of these children in future.

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Universal Infant Hearing Screening and Diagnosis in Hong Kong – Experience at Child Assessment Service, Department of Health, Hong Kong

Sylvia Doo, Catherine Lam
Child Assessment Service, Department of Health

Introduction

Bilateral congenital permanent hearing impairment (PCHI) of moderate (40dB) or greater degree can expected to lead to significant negative impact on the social, emotional, and intellectual development of affected individuals as well as society as a whole [1]. It is estimated that prevalence ranges from 1 to 3 per 1000 newborns [2]. Early identification of hearing impairment allows timely intervention to prevent significant speech and language deficits [3].

In 1993, the National Institutes of Health Consensus Development Conference on Early Identification of Hearing Loss in Infants and Young Children recommended that all newborns should be screened for hearing loss within the first 3 months of life, preferably prior to hospital discharge [4]. In 1994, the Joint Committee on Infant Hearing (JCIH) also recommended that all infants with hearing loss be identified before the age of 3 months, and receive intervention by the age of 6 months [5]. In their 2000 position statement, the principles and guidelines of implementing the screening programme with appropriate intervention were stressed [6]. Universal neonatal hearing screening is preferred over screening using a high-risk register, which can only identify around 50% or less of infants with hearing loss [7].

In Hong Kong, universal infant hearing screening in Maternal and Child Health Centres (MCHC) of the Department of Health's Family Health Service (FHS) using distortion product otoacoustic emission (DPOAE) was launched in August 2003. Being a major referral centre for diagnostic confirmation, experience of the Child Assessment Service (CAS) of the Department of Health is shared in this article.

Child Assessment Service Data

In a one-year period (July 2005 to June 2006), 1294 children were referred to the 7 Child Assessment Centers (CAC) of CAS from various sources with the chief complaints of suspected hearing problem. Among these, 1142 babies (88.25%) were referred because of failed OAE screening in Maternal & Child Health Center (MCHC). Age at referral ranged from 0.33 month to 13.1 months with a mean 3.09 months. There were 670 (59%) boys and 472 (41%) girls with a male to female ratio of 3:2. At the time of this report, confirmatory tests were completed in 1076 of these children. The age of diagnosis ranged from 0.33 months to 15.8 months with a mean of 4.77 months(Table 1).

Table 1. Age at referral and diagnosis

	Age at referral (month)	Time taken for diagnosis (month)	Age at diagnosis (month)
Maximum	13.1	13.40	15.8
Minimum	0.33	0.00	0.33
Mean	3.09	1.67	4.77

For these 1294 children referred by various sources for suspected hearing problems, the majority were seen within two months (mean 1.67 months). Diagnostic OAE (TEOAE) was used in 94.8% of children as the first audiological assessment. Most of those who failed TEOAE were evaluated with the Auditory Brainstem Response test (99.26%). A small proportion (9.7%) of children old enough to demonstrate head turn will be given the Distraction Test as the follow-up assessment.

Among the 1076 children with evaluations completed, 106 babies (9.85%) were confirmed to have some degree of hearing loss (Figure 1). Sixteen (1.49%) were classified as having significant hearing loss, i.e. hearing loss at moderate grade or worse in the better ear: six with moderate grade loss (hearing threshold 41-55dB), three with moderately severe grade loss (hearing threshold 56-70 dB), three with severe grade loss (hearing threshold 71-90 dB) and four with profound grade loss (hearing threshold >90dB). All of these were of the sensorineural type of hearing loss.

According to the statistics from the Family Health Service during the same period, a total of 36,075 babies were screened at their MCHC for hearing impairment. 1228 babies (3.40%) failed screening and were referred for confirmatory assessments at CAS or Hospital Authority ENT Departments. As mentioned, 1142 referrals to CAS were received from MCHC (93%). 16 of these babies were confirmed to have significant hearing loss. The yield of screening with CAS data alone was 0.44/1000 (16/36,075), and positive predictive value for the cohort referred to CAS of 1.4% (16/1142).

Discussion

Through the implementation of universal neonatal hearing screening in MCHC and strong collaboration with Child Assessment Service, we aim to identify children with congenital hearing loss no later than the age of 3 months with intervention by the age of 6 months, as recommended by the Join Committee on Infant Hearing (JCIH).

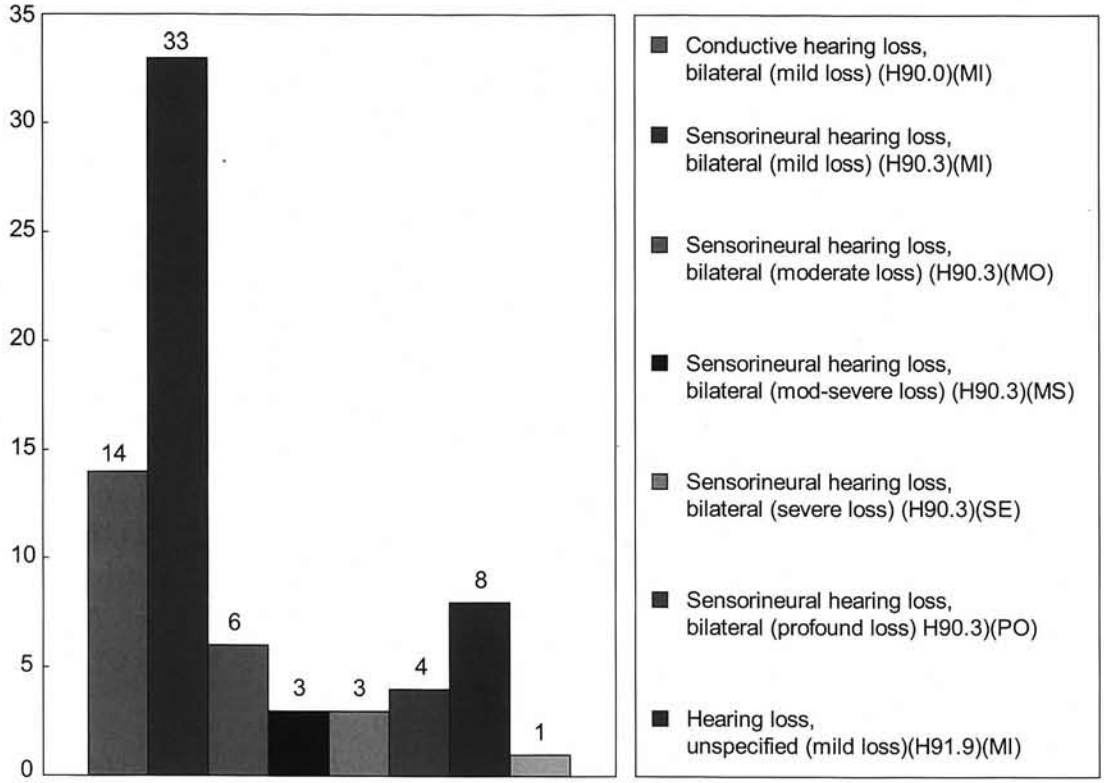

Figure 1. Number of children with different levels of hearing loss

During this report period, CAS received 93% of cases who failed hearing screening at MCHC. The present statistics hence provides good representation of the effectiveness of this program, with the screening yield of 0.44/1000 through diagnosis at CAS only. This has to be added to data from other ENT departments where audiological confirmation tests were also done. Furthermore, there are still a number of babies not registered at MCHC at birth (albeit small with MCHC coverage of over 90%), and hence not included in the screening program.

From February 2007, universal newborn hearing screening was implemented in all obstetric units under the Hospital Authority, where babies are to be screened by automated ABR within 3 days of birth. Moreover, some private hospitals are also including hearing screening as part of the routine examination for newborn babies. With the increase awareness of early screening and cooperation of various services from both public and private sectors, it is hoped that early diagnosis and intervention can really be achieved for children with significant hearing loss in Hong Kong.

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Recent Advances in Cochlear Implants

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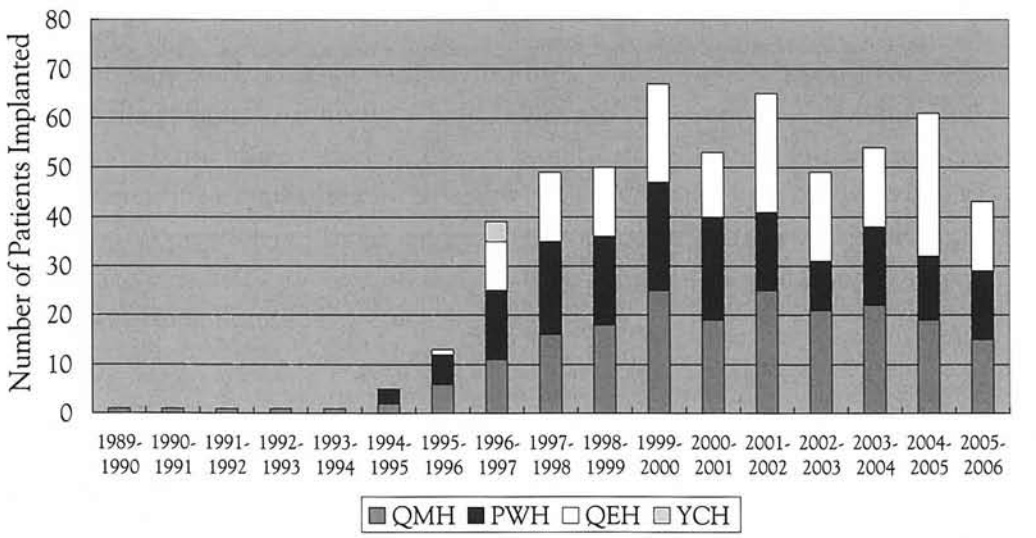
Introduction

Hearing impairment affects approximately one in a thousand newborns and the percentage of the population affected by hearing impairment increases dramatically with age; with at least 30% of individuals over 65 years of age having certain degrees of hearing impairment [1]. For hearing impaired persons with profound deafness, conventional hearing aids provide little or limited benefit and cochlear implantation has been shown to be a safe and reliable option in the management of profoundly deaf adults and children. Cochlear implant is an electronic device capable of substituting for the external, middle and inner ears, transforming sound vibration into electrical signals that stimulate remaining auditory nerve fibres in a manner that the brain can perceive in a meaningful manner.

History of Cochlear Implant in Hong Kong

The first multi-channel cochlear implant was performed in 1989 and by today, more than 500 adults and children have been implanted. There are three cochlear implant centres in Hong Kong, at the Queen Mary Hospital, Prince of Wales Hospital and Queen Elizabeth Hospital. Statistics on cochlear implants performed in Hong Kong are summarized in Figure 1.

Fig. 1: Statistics on Cochlear Implants in Hong Kong



The Selection Criteria of Paediatric Patients for Cochlear Implant

In Hong Kong, the selection criteria of paediatric cochlear implant patients are based on the recommendations issued by the Food and Drug Administration (U.S.A.). The age of patients should preferably be twelve months or older and the patients should suffer from profound deafness bilaterally, with thresholds greater than 90dBHL for 1kHz and above. The patients should have bilateral hearing aid fitting for at least 3 months. Detailed hearing aid evaluation should be performed and the aided functional thresholds from above 1kHz should be out of the speech spectrum. CT scans of the cochlea should be arranged to rule out the abnormality of inner ear. The patient should be fit for surgery, with no psychological or neuropsychological contraindications.

The Cochlear Implant Operation

Figure 2 shows a modern cochlear implant system. The microphone on the speech processor picks up the sound, analyzes and digitizes the sound into coded signals which are sent via a transmitting coil across the skin to the receiver–stimulator of the internal implant. These signals are sent to electrodes that stimulate the remaining nerve fibres, to be then recognized as sounds by the brain, producing a hearing sensation.

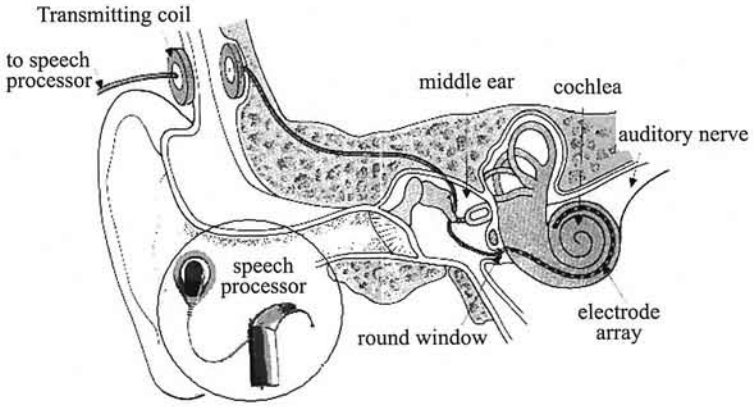


Figure 2: Block diagram of a cochlear implant system

Medical Precautions and Contraindications

The cochlear implant surgery incurs the general risks associated with surgery under general anesthesia such as bleeding and wound infection, potential disturbance or damage of the facial nerve, stiffness or numbness around the ear, disturbance of taste or balance, or noticeable change in the tinnitus. Special precaution should be taken in future Magnetic Resonance Imaging (MRI). Surgical diathermy and electroconvulsive therapy need to be handled with caution. Head trauma in the area of the implant area may damage the internal device and result in its failure.

Recent advances in Cochlear Implants

The recent advances in cochlear implant include new technologies in electrode and speech processor designs, speech coding strategy, neural response telemetry, magnetic resonance imaging compatibility, combined electric and acoustic stimulation and bilateral cochlear implant.

Electrode Design

The soft tip electrode design can protect the delicate cochlear structures from damage during cochlear implant surgery so as to preserve the residual hearing. The pre-curved electrode array enables the close proximity of the electrodes to the auditory nerve, with the benefits of focused stimulation and increased power efficiency.

Speech Processor

Modular design of using both body-worn and behind-the-ear speech processors gives the cochlear implant recipients wearing options. Moreover, the splash proof speech processor can withstand rain, sweat and moisture especially for children.

Speech Coding Strategy

High rate stimulation in speech coding strategy improves temporal information [2] and current-steering technology can provide up to 120 virtual channels. By varying the proportion of currents delivered to adjacent electrodes simultaneously in each electrode pair, the increased spectral information can possibly provide better tonal perception in cochlear implant recipients of tonal languages.

Neural Response Telemetry

Most modern cochlear implants have telemetry capability which allows bi-directional communication link. Neural response telemetry (NRT) is a technique in recording the near field electrically evoked compound action potential (ECAP) using the intracochlear electrodes of the computer implant system [3]. It provide clinicians valuable information for the programming of the cochlear implant device especially young children.[4].

Magnetic Resonance Imaging (MRI) compatibility

The magnetic and electrical fields during MRI examination can potentially cause hazard to cochlear implant recipient and damage the device. The removable magnet by minor surgery provides safe magnetic resonance imaging procedure up to 3.0 Tesla. [5]

Combined Electric and Acoustic Stimulation

The Combined Electric and Acoustic stimulation (EAS) is the concept of using hearing aid and cochlear implant technology together in the same ear [6]. The hearing aid acoustically amplifies low frequencies and the cochlear implant electrically stimulates the middle and high frequencies. The combined stimulation can be achieved by using a specially designed short cochlear implant electrode. The short electrode can preserve the low-frequency hearing in the apex of the same cochlea. Combined acoustic and electric stimulation can provide better speech intelligibility in noise.

Bilateral Cochlear Implants

Traditionally, cochlear implant surgery has only been routinely performed in one ear due to loss of residual hearing following cochlear implantation, the belief that one ear should be preserved in order to benefit from future technologies or for re-implantation, and the cost-benefit issues associated with a second device. However with the success of unilateral cochlear implantation and the numerous benefits demonstrated by bilateral hearing aid fitting [7], many cochlear implant recipients and their clinicians have started to question whether significantly enhanced speech understanding and better sound localization may be achieved through bilateral cochlear implantation.

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Auditory Neuropathy – what do we know so far?

Sylvia Doo
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Introduction

Auditory neuropathy (AN) is a term used to describe a condition in which the patient displays auditory characteristics consistent with normal outer hair cell function and abnormal neural function at the level of the VIIIth (vestibulo-cochlear) nerve. This clinical syndrome is characterized by the presence of otoacoustic emission (OAE) and/or cochlear microphonics (CM) in conjunction with absent or grossly abnormal auditory brainstem response (ABR)[1].

Though more discussion was only made in the recent decade, AN was first reported in late 1970's as paradoxical findings because of discrepancy between absent ABR and the presence of hearing thresholds (although with some reduction)[2]. Only with the advance of technology and use of OAE can we now identify the disorder. Different names were used to refer to this disorder: central auditory dysfunction, auditory neural synchrony disorder, auditory neuropathy and most recently, auditory dys-synchrony.

While any disorder of the auditory neural pathways from to VIIIth nerve to the cortex might be defined as an auditory neuropathy, the current use of the term relates specifically to more peripheral portions of the auditory pathways in the area between the outer hair cells and brainstem. These patients are distinguished from patients with space-occupying lesions, such as VIIIth nerve tumors, or multiple sclerosis, in that radiological evaluation yields normal results and even the most peripheral responses from the VIIIth nerve are absent[3]. Patients with auditory neuropathy require a different management approach to their auditory and communication problems from approaches used in patients with usual peripheral hearing losses.

Epidemiology

The prevalence is presently unknown with estimates varying from 1/200 to 15% of patients with sensorineural hearing loss[4,5]. The frequency of occurrence of AN in children attending schools for the hearing impaired in Hong Kong was estimated to be 2.44% (3 out of 123 children screened)[6].

Etiology and risk factors

The characteristics of auditory neuropathy most likely reflect more than a single etiology. Possible sites of auditory neuropathy include inner hair cells, the tectorial membrane, the synaptic juncture between the inner hair cells, auditory neurons in the spiral ganglion,

and the VIIIth nerve fibers, or any combination of the above. Neural problems may be axonal or demyelinating. Afferent as well as efferent pathways may be involved. The problem might also be related to a biochemical abnormality involving neurotransmitter release[3]. The specific sites and mechanisms of auditory neuropathy have yet to be determined.

Several factors have been linked to auditory neuropathy in children. However, a clear cause and effect relationship have not been proven. Some children who have been diagnosed with auditory neuropathy experienced certain health problems as newborns, or during / shortly before birth. These problems include jaundice, premature birth, low birth weight, and an inadequate supply of oxygen to the unborn baby.

Auditory neuropathy runs in some families, which suggests that genetic factors may be involved in some cases[7-9]. Some people with auditory neuropathy have neurological disorders that also cause problems outside of the hearing system. Examples of such disorders are Charcot-Marie-Tooth syndrome, Friedreich's ataxia and hereditary neuropathy[1].

Clinical features

Patients with AN can present with variable pure-tone thresholds that can range from normal to severe or profound hearing loss ranges. Some patients show rising or unusual configurations and threshold responses may or may not be symmetric between ears. In many but not all patients with auditory neuropathy, word recognition in quiet surroundings is poorer than one would predict from their pure-tone average[1]. Furthermore, some patients who show some word recognition ability in quiet surroundings have great difficulty understanding speech, even sentences, when there is even a small amount of background noise. This difficulty may be related to neural timing problems that may limit the ability to follow rapid transitions of normal speech[3].

Patients with either some residual hearing ability or later-onset progressive auditory neuropathies tend to rely heavily on lip-reading to supplement whatever auditory information is available to them. While reception of speech is difficult, patients generally have normal sounding speech and vocal qualities, suggesting an intact monitoring system.

While patients may display characteristics of central auditory processing problems (e.g., inattention, missing of some information, inconsistencies in responses, etc.), peripheral measures such as middle-ear muscle reflexes and the ABR are abnormal in auditory neuropathy, while function at the brainstem level is more often normal in patients with classic central auditory processing disorders.

Auditory neuropathy may also affect vestibular function causing dizziness and problem in balancing[10].

Management

Identification of auditory neuropathy presents a particular diagnostic problem in infants and children where the incidence of otitis media is higher than in older children and adults. If middle ear problems prevent evaluation of otoacoustic emissions, it may be necessary to evaluate outer hair cell function using cochlear microphonics measurement since this response appears less vulnerable to mild middle-ear problems than are OAEs. In addition to the complicating factor of middle ear problems, patient could have a co-existing peripheral hearing loss that could affect the ability to measure otoacoustic emissions.

Until the underlying etiologies of AN are better understood, the appropriateness of using hearing aids and cochlear implants is difficult to determine. Adult patients with auditory neuropathy generally report that hearing aids are of little or no benefit. Some patients find FM systems helpful in situations where enhancement of signal-to-noise ratios allows use of residual hearing for speech understanding.

Hearing aids are being tried to a limited extent in some children with AN. Generally, high quality, low gain, wide dynamic range compression hearing aids are recommended. This approach is intended to minimize any deleterious effects of amplification on otoacoustic emissions. If hearing aids are tried, frequent monitoring of otoacoustic emissions for either temporary or permanent effects on OAEs should be part of the management program.

The potential benefit of cochlear implants is still an open question. If the underlying etiology of the auditory neuropathy in a particular patient is cochlear in origin (i.e., the inner hair cells and/or the hair cell-nerve junction) and neural function is intact, then a cochlear implant may be potentially beneficial. In cases where the underlying etiology involves neural function, then the anticipated results with a cochlear implant may be less predictable. However, no tests are currently available to determine whether an individual with AN might benefit from a hearing aid or cochlear implant.

Another major dilemma involves the development of communication abilities in infants and young children identified with AN. Since input to the auditory system and processing of auditory stimuli is most likely compromised, alternative input methods may be most helpful. Most hearing health experts agree that parents should work with a team of professionals who consider the situation and options for each child as well as the child's family members and caregivers. Most also agree that parents and caregivers should interact often with infants who have AN by holding, facing, smiling at, and responding to the child.

There are two main philosophies of how to teach infants and children with AN to communicate. One favors using sign language as the child's first language. The second encourages the use of listening skills and skills in spoken language together with technologies such as hearing aids and cochlear implants. A combination of these two approaches can also be used. Some health professionals believe it may be especially difficult for children with AN to learn to communicate only through spoken language because their ability to understand speech is often greatly impaired. Adults with AN and older children who have already developed spoken language may benefit from learning how to speech read (also known as lip reading).

Prognosis

In adult patients, hearing generally seems to remain stable. However, it may also show fluctuation (as in cases of temperature sensitivity or auto-immune disorders), or progressively worsen (as in some patients with HMSN).

In infants, both decline in hearing and improvements in auditory function have been observed. Some newborns may show improvement if neuromaturation is the underlying problem. Other cases have been reported where auditory function, reflected in development of speech and language, develops over a longer period of time. Still other infants and young children have shown a progressive decrease in auditory responsiveness[3].

Until the etiologies underlying auditory neuropathy can be identified and distinguished clinically, it will be impossible to make accurate predictions about changes in auditory ability. For now, changes – be they improvement or decline – can be ascertained only through long-term follow-up.

Otoacoustic emissions (OAE) are faint sounds emitted by the cochlea, either spontaneously or in response to an acoustic signal. These emissions are thought to be generated by active movements of the outer hair cells and are left after severance of the auditory nerve. OAE are used as a test of outer hair cell function in patients with hearing loss or as an objective test of the integrity of the cochlear in patient unable to make behavioural responses for an audiogram (e.g. infants) [1].

Cochlear microphonics (CM) is an electrical potential generated by the inner and outer hair cells in response to sound stimulation. This potential is generally only recorded in response to high level sound and is often obscured by the N1 response of the auditory nerve (wave I of ABR) when it is present. CM may be visible inpatients having AN because of the lack of time-lock activity from the auditory nerve in that condition which would otherwise hide the CM [3].

OAE sometimes diminish or disappear in children with AN, for reasons that are unclear, while the CM has been more stable. When OAEs disappear, the CM would suffice as evidence of good hair cell function for purpose of AN diagnosis [3].

Auditory brainstem response/potential (ABR) are the far-field reflection of electrical activity of the VIII nerve and auditory brainstem pathway that can be detected with scalp electrodes. Waves I and II reflect activity of the distal and proximal portions of the VIII nerve, respectively; while wave III, IV and V reflect activity in central portions of the brainstem auditory pathway. Judicious use of ABR can assist in localizing lesions to particular portions of the auditory pathway [1].

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Balance and Motor Skills in Children with Hearing Impairment

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Introduction

With congenital hearing loss being diagnosed early in life today, early intervention focused on management of the hearing impairment and support for communication skills are much emphasized in identified young children. However, in observing this group of children as they grow up, teachers and parents sometimes report incoordination, clumsiness, and balance deficits which may affect their ability to function optimally within the environment and to interact with their peers (Butterfield and Ersing, 1986; Rine et al, 1996). Balance disorder in children is very often difficult to recognize. Children are not able to describe symptoms of their coordination problems, and simply look awkward and clumsy to peers and adult. Early identification and remediation of motor and balance deficits are important, both for training of motor skills, as well as for avoiding adverse social as a consequence.

Background information

Balance is maintained through visual, proprioceptive and vestibular signals. Damage to any of these systems or an abnormality in the central nervous system (CNS) that coordinates impulses from these three sensory systems can cause balance problems (Casselbrant & Mandel, 2005). Balance skills and normal motor performance require an intact postural control system which depends on sensory components, as well as motor and integrative components (Woollacott et al, 1987).

The functional vestibular system is comprised of vestibule-spinal and vestibulo-ocular systems. The vestibule-spinal system contributes to the postural tone necessary for the emergence of early motor milestones, such as rolling, sitting and standing. The vestibulo-ocular system contributes to visual stabilization, acuity, and the development of visual spatial and perception abilities (Montgomery, 1985).

Reports showed that vestibular dysfunction is common in children with sensorineural hearing loss and morphological changes are found in the labyrinths of these children (Crowe and Horak, 1988; Rine et al, 1996; Rine et al, 2000). Children with hearing impairment and normal vestibular function perform normally on balance and dynamic motor tasks, while children with hearing impairment and concurrent vestibular dysfunction present with balance problems (Crowe and Horak, 1988; Horak et al, 1988). Researchers postulate that insult to vestibular structures early in life may result in morphological alterations in the vestibular system, with consequent aberration of balance and motor proficiency (Horak, Shumway-Cook, Crowe et al, 1988; Siegel et al, 1991).

Symptoms of Vestibular Dysfunction in Children (Rine, 2000)

Peripheral Disorders

1. Persistent, obvious nystagmus (longer than 1 second) on head movement in the light or dark
2. Visual instability on head movement, complaints of blurring or double vision
3. Below age level balance abilities, e.g. tandem, single leg stand
4. Complaints of spinning sensation or dizziness
5. Below age level vestibular ratios on posturography testing, unable to maintain upright on conditions 5 and 6 (Figure 1)
6. Hypo- or hyperactive responses on post-rotary nystagmus test
7. May or may not have hearing loss or tinnitus
8. May be fearful of movement activities, or crave it
9. Complaint of incoordination

Central Disorders

1. Delay or below age level performance on gross motor tasks
2. Delay or below age level performance on visual motor, visual perception tests
3. Persistence of tonic reflexes
4. Visual instability, particularly with head movement
5. Normal, hypo-, or hyperactive responses on post-rotary nystagmus test, although majority either hypo- or hyperactive
6. Below age level on vision and vestibular ratios on posturography testing
7. Below age level performance on dynamic posturography, with stepping or loss of balance on conditions 4 to 6 (Figure 1)
8. Possible sensory integrative dysfunction

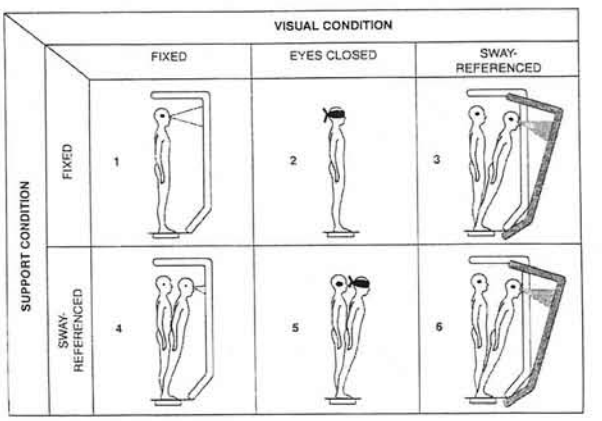

Figure 1. The six sensory testing conditions of the dynamic posturography platform (Casselbrant & Mandel, 2005).

Assessment of Vestibular Dysfunction in Children

Assessment Tool	Test type	Vestibular system	Age Group
Peabody Developmental Motor Scales	Balance and motor development Visual perception	VO and Vsp	0 to 6 years
Bruininks Oseretsky Tests of Motor Proficiency	Balance and gross motor ability coordination	VO and Vsp	4 to 14 years
The Sensory Integration and Praxis Tests	Sensory integration and motor planning ability, visual perception	VO and Vsp	4 to 8 years
Southern California Post Rotary Nystagmus Test	Vestibular ocular system test	VO	5 to 11 years
Functional Reach	Balance ability in standing	Vsp	5 to 15 years
Pediatric Clinical Test of Sensory Integration for Balance	Balance under varying sensory conditions	Vsp	4 to 9 years
Dynamic Posturography Testing	Functional test of balance Effectiveness of vestibular, visual and somatosensory systems	Vsp	3 to adult
Timed Up and Go Test	Functional stand and walk test	Vsp	3 to adult
Vestibular Auto-Rotation Test	Visual stability	VO	

VO – vestibulo-ocular system

Vsp – vestibulospinal system

(Rine, 2000)

Results of various studies

Studies reporting the motor and balance deficits in children with hearing impairment have been inconsistent in their findings. Boyd (1967) reported that many, although not all of the 90 children between 8 to 10 years of age with severe hearing impairment of various etiologies, presented with impaired static balance, e.g. single leg stand. Wiegersma & Van der Velde (1983) reported impaired dynamic coordination, e.g. balance beam walking, skipping, hopping and lateral hopping in 55 children 6 to 10 years of age with idiopathic hearing impairment. Dummer et al. (1996) examined 200 children with hearing impairment of varying etiology, and reported deficits in those below 10 years of age, including on object control, e.g. ball skills, and locomotor tasks, e.g. balance beam. Butterfield & Ersing (1986) reported that although no impairment of most motor skills was evident in 132 children with moderate to severe hearing impairment between 3 to 14 years old, impairment of static and dynamic balance skills, e.g. kicking, jumping and hopping was evident in the youngest children. Rine et al. (1996) examined 7 children 4 to 5 years of age with profound sensorineural hearing loss and reported immature responses in static and dynamic balance, with a lack of proximal stability and delayed, less mature equilibrium responses. All these inconsistent results could be explained by the fact that different components of balance are tested within the different tests, and there are different variables such as etiology, vestibular dysfunction and age at the time of testing (Bundy et al, 1987).

Age is a confounding factor in developmental test results and may contribute to different test results. The critical period for the development of postural control is between the ages of 3 and 7 years old (Shumway-Cook & Woollacott, 1985; Woollacott et al, 1987). During this time, sensory integrative capacity is developing, with a visually dominant pattern seen in children 3 years of age, and mature responses evident by the age of 7. Therefore, it may be proposed that programs to identify and address postural control impairment in children should occur between the ages of 3 and 7 years old (Rine et al, 1996).

Various studies had been done to investigate the effects of remediation programs on hearing impaired children with balance deficits. Some reported that the exercise intervention focused on substitution strategies may halt the progression of motor development delay and enhance the postural control abilities in children with sensorineural hearing loss and concurrent vestibular impairment (Rine et al, 2004; Herdman & Clendaniel, 2000). Some reported significant improvement in dynamic balance ability in 17 children with hearing impairment, who received a 16-week rhythmic gymnastic program at a frequency of 3 lessons per week, for 40 minutes (Fotiadou et al, 2002). Others reported that after a 10-day exercise program consisting of static balance practice, single leg stance time improved in a group of 7 to 11 years old children with severe or profound hearing impairment, but the postural sway in double or single leg stance, with eyes open or closed did not (Effgen, 1981).

Conclusion

Motor and balance deficits in young children with sensorineural hearing impairment and evidence of vestibular dysfunction have been reported in different studies. The postural control in young children with sensorineural impairment deviates from the norm and is reflected in less skilled motor ability. The development of appropriate intervention is dependent upon an accurate and comprehensive evaluation and analysis of postural control and vestibular system function. Based on the interpretation of the evaluation results, intervention program should begin with activities to facilitate balance and vestibular training, sensory integration, and visual stabilization exercises.

Despite the above findings, the question of balance and motor competencies in children with hearing impairment is complex, and the results derived from various studies are still inconsistent, owing to the use of different measures of balance, lack of control of age of subjects, limited subject size in some studies, and the omission of measures of vestibulo-ocular function. Hence further research should be considered to ensure a more comprehensive understanding on the issue, and the provision of optimal intervention.

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Predictive and Diagnostic Genetic Testing for Deafness

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Approximately 1-3 in 1000 children is affected by severe or profound hearing loss at birth or during early childhood (prelingual deafness). In China, there are more than 20 million people suffered from the hearing impairment and more than 30 thousand children with profound hearing loss were born every year. At least 50% of congenital profound hearing loss is attributed to genetic defects. This proportion is continually increasing as public health conditions improve, leading to a decrease in the prevalence of hearing loss resulting from infections. Genetic deafness is divided into syndromic forms, in which hearing loss is associated with a variety of other anomalies, and in non-syndromic forms. The non-syndromic forms of deafness account for most part (70%) of deafness with genetic etiology. More than sixty auditory genes have been identified up to now. State-wide universal newborn hearing screening programs and characterization of auditory performance and intervention strategies following neonatal screening in both the U.S.A and China become a very important part in clinic management for hereditary hearing impairment. Specifically, infants who are hard of hearing and deaf who receive intervention before 6 months of age maintain language development commensurate with their cognitive abilities through the age of 5 years. Some infants with hearing loss will pass the newborn hearing screening. Both ABR and OAE technology can show false-negative findings due to the fact that newborn hearing screening can't predict late onset of hearing loss such as cases suffered from Enlargement of Vestibule Aqueduct (closely related to the mutations in SLC26 gene) or non-syndromic hearing loss caused by mitochondrial DNA mutations.

We are now establishing a standard genetic testing lab for deafness and setting up the protocol for genetic diagnosis and screening for hereditary hearing loss in China. The data from our lab showed that the genetic background of hearing loss in children is very strong. An epidemiologic investigation for molecular etiology of congenital and early onset hearing loss in different area of China was initiated one year ago. The preliminary data showed that the GJB2 related deafness is most important type of inherited deafness. Among 1244 deaf children, 101 children were found to have homozygous GJB2 235 del C mutation and 120 children have heterozygous GJB2 235 del C mutation. Sequencing analysis showed that most single GJB2 235 del Cs were accompanied by another mutation like 299-300del AT or even 35delG or 35insG which are never reported in Chinese before. Among the same group, 37 deaf children were found to carry mitochondrial DNA A1555G, most of them have received the treatment of aminoglycoside antibiotics. Because the very low penetrance of deafness in these family, the other member of these family are benefit from the genetic screening for this mutation. Among the 38 deaf patients with enlarged vestibular aqueduct(EVA), thirteen patients have homozygous mutations, nine have compound mutations, thirteen have heterozygous mutations in SLC26A4 gene. The data provide very strong evidence for that the EVA syndrome is genetic disease.

The significance of predictive and diagnostic genetic testing for deafness in China is to provide a tool to uncover the exact etiology of congenital deafness and introduce suitable treatment, preventive methods and prenatal testing for birth quality guarantee. The counseling for hereditary hearing loss will be essentially improved on the basis of the precise result of deaf gene diagnosis. In 4 Chinese family which have first child with congenital severe neurosensory hearing loss, prenatal genetic testing was provided for parents for their second pregnancy. Two new born babies passed new born hearing screening test and another two fetus were proved to be GJB2 heterozygous mutation and PDS heterozygous mutation carrier respectively which. Additionally the fact that mitochondrial DNA A1555G mutation are frequently detected in the pedigrees and sporadic sensory-neural hearing loss induced by aminoglycoside antibiotics in China reveals the importance of genetic screening for this mutation before the usage of aminoglycoside antibiotics.

The Hong Kong Society of Child Neurology and Developmental Paediatrics
Annual Scientific Meeting 2007
on
"Energy Crisis of Nervous System"
16 – 19 November 2007

First Announcement

Dear colleagues,

We are pleased to announce that our Annual Scientific Meeting for this year will be held on 16 – 19 November (Friday – Monday) at the Queen Elizabeth Hospital and the Marco Polo Hong Kong Hotel. The theme for the meeting is "Energy Crisis of Nervous System". We are privileged to have Dr. Ingrid Tein from Canada as our course director to deliver lectures covering important aspects of this condition.

Dr. Ingrid Tein is an Associate Professor of the Department of Paediatrics at the University of Toronto. She is also the Staff Neurologist and the Senior Scientist in the Research Institute at the Hospital for Sick Children. Dr. Tein directs the Neuro-metabolic Clinic and Neuro-investigational Unit for the investigation, management and treatment of children with a variety of neuro-metabolic diseases including mitochondrial disorders, fatty acid oxidation defects, peroxismal disorders, glycolytic/ glycogenolytic defects, organic acidurias, etc. Her research interests are fatty acid oxidation and molecular genetics. In future, she will focus on the development of Paediatric Metabolic Exercise Physiology Unit.

In addition to Dr. Tein, we will also be hosting local experts from the fields of neurology, paediatrics and pathology, to share with us their latest research, both basic and clinical, and the most up-to-date information on this area, which will be of interest to specialists and professionals alike.

Your attendance will no doubt prove to be a valuable and enjoyable experience. We sincerely invite you to make plans now to join with other colleagues to exchange new ideas and share your experiences and knowledge.

You are invited to submit abstracts for the free paper presentation session as well. Any topics related to Child Neurology and Developmental Paediatrics are welcomed. For detailed information, please refer to the attached "Call for Abstracts" submission form.

The registration fee will be waived for members of our society. For non-members, it is $200 per person.

Should you have any enquiry about this meeting, please do not hesitate to contact our meeting secretariat at tel: (852) 2155 8557 or e-mail: meeting.hk@asia.cmpmedica.com or visit our Society's website at www.fmshk.com.hk/hkcndp/index.htm

Look forward to seeing you in the meeting.

Yours sincerely,

Dr. Chok-wan Chan
President
The Hong Kong Society of Child Neurology and Developmental Paediatrics

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Call for Abstract

Participants are invited to submit abstracts for the free paper session of the meeting. This is to provide a platform for academic and clinical workers, especially frontline colleagues, to share their experience in areas covering ANY aspects of Child Neurology and Developmental Paediatrics. Accepted abstracts will be published in the programme book. Our course director and local experts will be invited as the judges, and prizes will be awarded to the winner of the presentations. Presenters are allowed to submit more than one abstract.

Submission Guidelines:

1. Abstract must be written in English.
2. The content should include: i) Background & Objectives, ii) Methods iii) Results and iv) Conclusion. However, it is not necessary to organize the abstract under different headings.
3. Please clearly type the title of the abstract, author name(s) and institution name(s) on top of the abstract. The name of the presenting author should be underlined.
4. Type the abstract using a computer-generated, Times New Roman, 10-point font, with single-line spacing. The abstract should not exceed 200 words.
5. The deadline for abstract submission is 15 September 2007, and notification of acceptance will be sent out by 6 October 2007.
6. Once submitted, abstracts will not be returned.
7. All abstracts must be sent or faxed to the meeting secretariat by completing the abstract submission form. A soft copy of the abstract must either be saved on a CD-rom and sent to the meeting secretariat or directly e-mailed to meeting.hk@asia.cmpmedica.com
8. All free paper presenters are required to register with the meeting for presenting their works.

Meeting Secretariat:
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Preliminary Scientific Programme

Date	Time	Session	Topic	Speaker
16 Nov 2007 (FRI)	1800 - 2000	*Registration and Light Buffet Dinner*		
	2000 - 2200	Seminar I	Approach to the Diagnosis and Management of Fatty Acid Oxidation Disorders	Dr. Ingrid Tein
17 Nov 2007 (SAT)	1230 - 1400	*Registration and Light Buffet Lunch*		
	1400 - 1500	Seminar II	Primary and Sceondary Disorders of Carnitine Matabolism	Dr. Ingrid Tein
	1500 - 1530	*Tea Break*		
	1530 - 1600	Local Presentation	Local Scenario of Neurometabolic Diseases - Experience of a Tertiary Referral Center	Dr. Cheuk-wing Fung
	1600 - 1700	Seminar III	Approach to the Diagnosis and Management of Mitochondrial Disorders Part I – mtDNA Encoded Defects	Dr. Ingrid Tein
18 Nov 2007 (SUN)	0900 - 0930	*Registration*		
	0930 - 1030	Seminar IV	Approach to the Diagnosis and Management of Mitochondrial Disorders Part II – nuclear DNA Encoded Defects	Dr. Ingrid Tein
	1030 - 1130	Free Paper Session	Oral Presentations of Free Papers	
	1130 - 1200	*Tea break*		
	1200 - 1300	Local Presentation II	Local Laboratory Support on Neurometabolic Diseases Genetic study of CNS Diseases	Professor Nelson Tang Dr. Ivan Lo
	1300 - 1430	*Light Buffet Lunch*		
	1430 - 1600	Case Presentation	Cases Discussion from Various Hospital	
	1600 - 1630	*Tea Break*		
	1630 - 1710	Seminar V	Application of Exercise Physiology to the Diagnosis of Metabolic Myopathies	Dr. Ingrid Tein
19 Nov 2007 (MON)	1830 - 1900	*Registration*		
	1900 - 2000	Keynote Lecture	Approach to the Diagnosis and Management of Muscle Cramps, Exercise Intolerance and Recurrent Childhood Myoglobinuria	Dr. Ingrid Tein
	2000 - 2200	*Chinese Banquet*		

Venues:

16 November 2007: Lecture Theatre, 12/F., Block R, Queen Elizabeth Hospital, Jordon, Hong Kong
17 – 18 November 2007: Lecture Theatre, G/F., Block M, Queen Elizabeth Hospital, Jordon, Hong Kong
19 November 2007: Centenary Ballroom, G/F., Marco Polo Hong Kong Hotel, Tsim Sha Tsui, Hong Kong

The Hong Kong Society of Child Neurology and Developmental Paediatrics
Annual Scientific Meeting 2007
on
"Energy Crisis of Nervous System"
16 – 19 November 2007

Registration Form

(Deadline: 12 October 2007)

Name: Professor / Dr. / Mr. / Ms. _____ (_____)
(Please use block letters) Chinese Name

Job Title: _____

Department: _____

Hospital / Clinic: _____

Address: _____

Tel No.: _____ Fax No.: _____ E-mail: _____

Registration:

- ☐ I am a member of the HKCNDP*. Fee: Waived
- ☐ I am a non-member. Fee: HK$200 per person

*HKCNDP – The Hong Kong Society of Child Neurology and Developmental Paediatrics.

Please indicate your attendance by placing a "√" in the appropriate box(es) below:

- ☐ 16 November 2007 (FRI) – Buffet Dinner at Queen Elizabeth Hospital
- ☐ 16 November 2007 (FRI) – Evening Lecture at Queen Elizabeth Hospital
- ☐ 17 November 2007 (SAT) – Buffet Lunch at Queen Elizabeth Hospital
- ☐ 17 November 2007 (SAT) – Afternoon Sessions at Queen Elizabeth Hospital
- ☐ 18 November 2007 (SUN) – Morning Sessions at Queen Elizabeth Hospital
- ☐ 18 November 2007 (SUN) – Buffet Lunch at Queen Elizabeth Hospital
- ☐ 18 November 2007 (SUN) – Afternoon Sessions at Queen Elizabeth Hospital
- ☐ 19 November 2007 (MON) – Keynote Lecture at Marco Polo Hong Kong Hotel
- ☐ 19 November 2006 (MON) – Chinese Banquet at Marco Polo Hong Kong Hotel

Please complete and send registration form with appropriate registration fee (if applicable) by a crossed cheque made payable to "*The Hong Kong Society of Child Neurology & Developmental Paediatrics*":

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Abstract Submission Form

(Deadline: 15 October 2007)

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