Brainchild

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香港兒童腦科及體智發展學會

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Message from the President

Dr. CHAN Chok Wan

The Hong Kong Society of Child Neurology and Developmental Paediatrics was inaugurated in May 1994 with the vision to advance knowledge of child neurology and developmental paediatrics, to promote relevant services in Hong Kong, to provide public education and to foster comradeship for professionals within the two disciplines and all other colleagues working in the medical and health domains. To achieve these, education at all levels and in different formats is essential to ensure its ultimate fulfillment.

Over the past six years, we are pleased to witness the substantial results we have achieved within our disciplines. Our busy scientific activity schedules included regular bimonthly scientific meetings, neuro-developmental conferences, child neurology conference, case management conferences, the annual scientific meetings with world experts as course directors, adhoc meetings featuring prominent overseas speakers and others. Our Course Director for the Annual Scientific Meeting 2000 is Professor A Galaburda, Emily Fisher Landau Professor of Neurology and Neuroscience, Harvard Medical School who together with Professor Susana Camposano, eminent Consultant Child Neurologist from Chile, will conduct a course on "Language development, learning disorders, and brain plasticity: research and clinical implications" from 8-11th December, 2000 in Hong Kong. Professor Galaburda is a leading scientist and authority on the relationship between neurobiology, cognition and language; his seminal work on cerebral characteristics in reading disorders provides much of the basis of today's understanding on the subject. Through these experts together with our local contributions, I am confident that the meeting is going to be another exciting educational experience for all participants. These academic activities serve the purposes of continuing medical education, exchange of knowledge and experience, enhancement of collaboration for research and for quality assurance in subspecialty practice.

In the meantime, the HKCNDP Working Party on Specific Learning Disabilities and the Working Party on Rett Syndrome continue to thrive actively in organizing multidisciplinary programmes on the understanding, public awareness, local incidence, diagnostic tools, clinical approach, and

management of children suffering from these conditions. These together with the Society's policy to promote active collaboration of research projects amongst members have been successful in creating more opportunities for academic activities in our subspecialties in Hong Kong.

To this date, we are proud to witness the large number of publications that have been produced by our Society: the Manual of Developmental Paediatrics, the Manual of Child Neurology, Seminar Proceedings and Position Monographs by the SLD Working Party, Education Pamphlets on Epilepsy, Febrile Convulsion and EEG, and others. It is most encouraging to us that all the above have been well received by colleagues. The manuals, especially, have become popular references for clinicians.

The Education Bulletin and the Society Newsletter were established soon after the Society's inauguration with the former designated to keep our scientific activities on permanent record for future reference and for the benefit of those unable to attend the meetings, and the latter for communication between members on current events and Council activities. Both have attained excellent results in achieving their goals and missions.

The current inaugural issue ***Brainchild*** is another attempt of the Society to further our efforts by combining the Education Bulletin and Society Newsletter into one publication of wider contents and higher quality of articles and papers. The editorial board under Dr. Philomena Tse, Dr. Wu Shun Ping, Dr. Catherine Lam, Dr. Yam Ka Ling, Dr. Lau Wai Hung, and Dr. Liu Kam Tim are to be congratulated for launching this excellent production to be published triennially. We would also like to express our appreciation to Medcom Ltd, our publisher, for bringing this publication into reality and to Wyeth (Hong Kong) Ltd, our sponsor, for financing the publication.

Finally I would like to thank members and friends of our Society for their ever-unfailing support and their contributions towards our achievement. With your encouragement, we are confident that the Hong Kong Society of Child Neurology and Developmental Paediatrics will further thrive as we enter the new century. I wish you all reading pleasure with this new publication ***Brainchild***.

Editor's Note

Dr. CHAN Chok Wan

Amongst the manifold vision and mission of the Hong Kong Society of Child Neurology and Developmental Paediatrics, one important objective is to advance the knowledge of child neurology and developmental paediatrics, to improve and to promote their services in Hong Kong. Since its inauguration, the Education Bulletin has been proven to be very popular among members of the Society as well as among colleagues in the medical, nursing and allied health professions. For all these, we would like to thank the significant contributions from the authors, the ever-unfailing support from the readers and their constructive comments, and to the outstanding advice and technical input of our publisher Medcom for making the Education Bulletins possible.

Encouraged by this success, we are pleased to present this inaugural issue of ***Brainchild***, the official publication of the Society to which we encompass our past Education Bulletin and the Society Newsletter one, both of which were established since our Society's inauguration in May 1994. The name of the new publication serves to embody both the disciplines of child neurology and development, and also emphasizes the spirit of innovation and momentum which we are keen to promote. I am confident our readers will find this name appropriate and acceptable.

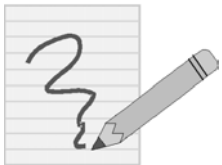
Brainchild, while continues to uphold the already successful history of the Education Bulletin and the Society News, will venture to explore other areas of interest within the domains of child neurology and developmental paediatrics. To this aim, we have added new sections on annotations from local and overseas experts on topics of current interest, reports from international conferences, reports on experience and recommendations after visits to overseas institutions by members, free papers and original articles, special announcements for academic activities, members' news, correspondence column, and others to be added in the course of time. At the beginning, we intend to publish the publication three times per year and to circulate them to members of the Society, local paediatricians, colleagues working within the domain of neurosciences (adult neurologists, neurosurgeons, adult and child psychiatrists etc.), family physicians, allied health professionals engaged in the fields of child neurology and developmental paediatrics. We sincerely hope that the publication will serve to

disseminate the academic information to all colleagues working for the same interest and dedication, and to promote better communication between different disciplines within the important domain of the neurosciences, To achieve this, we welcome comments, criticisms and support from all readers and, most important of all, your kind input of articles to this humble cultural garden of our Society.

I would like to thank key figures who contributed significantly to success of this new publication: Dr. Philomena Tse and Dr. Wu Shun Ping, Associate Editors, as well as Dr. Catherine Lam, Dr. Lau Wai Hung, Dr. Yam Ka Ling and Dr. Liu Kam Tim, members of the Editorial Board for their innovations and dedications in delivering the Brainchild into reality. I also would like to thank our publisher Medcom Ltd. and sponsor Wyeth (Hong Kong) for their support and contribution. Most important of all, I would like to thank all readers for their support and comments: all these are essential for the success of the publication.

Cover

The front cover is a crayon drawing of a 10 year-old girl who suffered from benign partial epilepsy with centrotemporal spikes. She has been exceptionally artistic and generous. She gave her doctor a painting every time she visited him. Her seizures did not return after commencing carbamazepine treatment, and she is looking forward to a withdrawal of drug soon.



Education Section

Pulmonary Rehabilitation in Children

Dr. Winnie GOH
Consultant Paediatrician, Duchess of Kent Children's Hospital, Hong Kong

Paediatric pulmonary rehabilitation has been mainly confined to broncho-pulmonary dsyplasia in the past.

Pulmonary rehabilitation for children with neuromuscular diseases was not a priority in Hong Kong due to perceived poor quality of life. In the past children with Duchenne Muscular Dystrophy would die at a very young age without active intervention. During the later years of their lives, they are usually uncomfortable and depressed due to hypoventilation and hypoxemia.

Ventilator dependent children were institutionalized and unable to enjoy a normal childhood.

The pulmonary rehabilitation program was started after reviewing a group of children with Duchenne Muscular Dystrophy and assisted ventilation were prescribed to those who needed intervention.

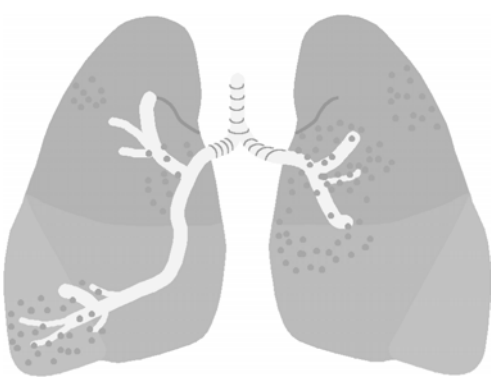
The program has since extended its scope of service to all children with neuromuscular diseases and those with obstructive sleep apnea. Children with developmental disabilities like Down Syndrome, Achondroplasia, mucopolysaccharidosis; congenital cranio-facial malformation and upper airway obstruction had benefited from this program. The main goal of pulmonary rehabilitation program is to provide good quality of life and children who are ventilator dependent be able to attend school and live in community.

Role of Physiotherapist in Respiratory Care of Duchenne Muscular Dystrophy Patient

Ms Annie NG
Physiotherapist I, Duchess of Kent Children's Hospital, Hong Kong

As a natural course of Duchenne Muscular Dystrophy, the pulmonary function deteriorates with time and particularly compromised in critical periods such as post-operation, chest infection and very late stage of the disease. It happens that the most common cause of death in this **muscle** disease is **chest complication**.

This is due to the progressive weakness of the respiratory and skeletal muscles, the lung **ventilation** becomes progressively ineffective. As a result, the lung failed to develop normally and fail to maintain its **compliance**, making it prone to develop **atelectasis**. Together with the weak expiratory muscles and limited chest expansion, the secretion easily **stuck in the lung**. Added with a **collapsing spinal deformity**, the pulmonary function further jeopardized. Knowing the underlying causes, as physiotherapist, we set our **goals** in respiratory rehabilitation aiming at improvement of lung ventilation, maintenance of lung and chest wall **mobility**, improvement of **cough effectiveness**, enhancement of the **coordination of breathing** in daily activities and development of ability to maintain **good bronchial hygiene**. The pulmonary program should includes **assessment**, breathing **exercise**, spine mobilization exercise, application of respiratory **aids and techniques**, **positioning** and **family education**. The physiotherapist's scope of practice starts from outpatient preventive care during the ambulatory stage, hospital-based care such as during acute chest infection, pre- and post-operative care; introduction to the use of various respiratory aids, community based care such as school and home program. We see our role proactively in the pulmonary rehabilitation process starts right **from the beginning** and goes all the way to the very end.



Role of Occupational Therapy in Pulmonary Rehabilitation

Ms. Patsy Pui Sai SIT
OTR, PDOT, Duchess of Kent Children's Hospital, Hong Kong

Occupational Therapy aims at maximize the independence of the children with ventilator support in performing the Activities of Daily Living including ***Self-care, Learning/School*** and ***Leisure*** activities so as to promote their quality of life. Ultimately, we target to let the patients to discharge home to enjoy the family life as soon as possible.

In order to achieve the aims, our treatment provided for these children with ventilator support including assessment and training in positioning and self-care aspects; using environmental control unit and computer access to assist in active learning and leisure cultivation; providing home modification and Community Occupational Therapy for this group of patients.

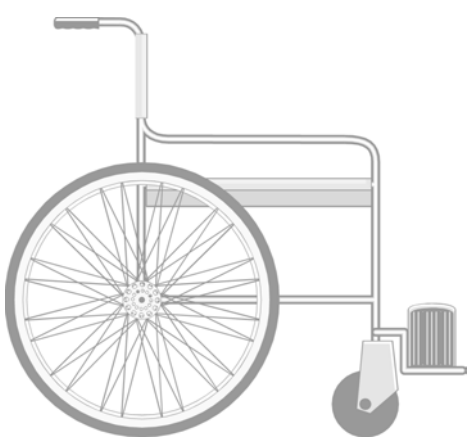
For a bed-bounded or wheel-chair bounded patients, we will provide **positioning** assessment and recommendation. Proper positioning not only minimizes the influence of pathologic forces on body posture but provide a stable base of support for performance and function; thus, it is the key to increased independence and improved self-esteem. In addition, it can prevent pressure sores and other deformities which commonly developed among this group of patients. Furthermore, Occupational Therapist will provide **wheel-chair assessment**, **prescription** as well as **maneuver training** in order to maximize their independence. Concerning about independence, **self-care assessment and training** will be provided as well. We aim at assessing the functional abilities of the patients and then train up their self-care skills with suitable **aids and gadgets**, if needed.

For the patients with ventilator support, **learning** and having their own **interest** will help them to relax, and build up their self-confidence. In this aspects, we assist the patients in learning actively and developing their own interest and leisure activities through assistive devices and technologies. For example, how can the input or the mouse be adapted to suit individual needs when using an ordinary **computer** in reading and writing, playing games, connect to the internet and sending e-mail, etc.? In fact, applications of electronic technology provide persons with disabilities with the

potential for increased control over the environments. Devices with the capacity to regulate aspects of a person's physical surroundings are called environmental control devices. Environmental Control Unit (ECU) consists of five parts, including switch device, control device, target device, connections and feedback device. However, how can these five parts be chosen to meet individuals' demands and can they use the ECU effectively and efficiently?

As most of the patients is severely physically disabled, they may have difficulty in accessing the computer or using the ECU actively. Hence, adaptive switches and mouse may be needed. There are wide varieties of **switches and control interface** available, including single, dual, and multiple switches. The switches may be activated by movement of different body parts, including head, tongue, mouth, jaw, eyelid, chin, neck, arm, finger, hand, leg and foot, depending on the functional ability of the patients. After choosing the suitable switch and control system, on-site practical training sessions by means of suitable software are essential in order to let them familiarize with the system and new acquired skills.

In order to encourage the patients to discharge home, **Community Occupational Therapist** (COT) will provide **home visit and recommendations on environmental modification** in order to empower the patients and their relatives to live at home safely. In addition, **parental education** on caring of the patients and training in using different assistive equipment or ECU in a safe and proper way are essential as well.



Duchenne Muscular Dystrophy: Quality of Life

Ms. Connie LEUNG
Clinical Psychologist, Duchess of Kent Children's Hospital, Hong Kong

The aim of this pilot study is two folded. Firstly, it is to explore the Health-Related Quality of Life (HRQOL) of patients with Duchenne Muscular Dystrophy and identify their psychological correlates. Secondly, it is to understand the psychological effects of the illness on the patients' mothers. The study investigated a group of Chinese patients with DMD (N=20, mean age=14.1, S.D.=5.46) seen in the Duchess of Kent Children's Hospital and/or studying in John F. Kennedy Centre and their parents. Instruments used were Campbell's Overall Life Satisfaction Scale, the Piers-Harris Children's Self-Concept Scale, and the Short-Form 36 (SF-36). The 'ambulatory group' and the 'wheelchair-bound group' were compared. Initial analysis by Mann-Whitney U test indicated that there was no significant difference between these two groups. On the 9-item 7-point scale of Campbell's Overall Life Satisfaction Scales, the patient subjects obtained a mean score of 49.2 (S.D.=10). The total self-concept score was well at the mean of the norm reference. Domains of the quality of life profiles of the patient group on SF-36 were within one standard deviation from the norm reference, notwithstanding the clinically significant difference in the Scales of 'Physical Functioning' and 'Role-Physical'. A significant relationship between the total Self-Concept and Mental Scale of SF-36 was found ($r=0.64$, $p<0.05$). Implications on future direction of service to improve the patients' and carers' quality of life were discussed.

Management of Skin Lesions in Neurocutaneous Syndromes

Dr. TUNG Man Kwong
Consultant, Plastic Surgery Division, Department of Surgery,
Princess Margaret Hospital, Hong Kong

The Neurocutaneous Syndromes are also known as the Phakomatosis or the Ectodermal Dysplasia. These include lesions of the skin and of the central nervous system. Several clearly distinct syndromes are recognized. They are Tuberous sclerosis, Neurofibromatosis & Sturge-Weber Syndrome. More syndromes can be found and described in bigger text e.g. Smith's....

From a Plastic Surgeon's point of view it is the skin lesions that are causing the concern. Are they malignant? Pre-malignant? Any potential for malignant change? Is it causing disfigurement? Is it enlarging? Is it infected? Has it got any discharge?

Gadgets available for the Plastic Surgeons are scalpel, diathermy and cauterization machines, laser machines, dermatone, liposuction machine, cryosurgery machine and dermabrator.

Most of these patients when presented are either too young to stand regional anaesthesia or have certain degree of MR. The surgical procedures are mostly done under general anaesthesia.

The Skin Lesions of concern are alopecia, haemangiomata, lymphangioma, naevi, pigmented spots, skin nodules, subcutaneous nodules and ulcers. Majority of these lesions are benign during the paediatric and adolescence period. However they are causes for concern and worry for patients and their parents. They do lead to disfigurement and social embarrassment.

A lot can be done by Plastic Surgeons for certain skin lesions even with only scalpel, diathermy and cauterization machines. These instruments are always available in all hospitals in the territory. For example the adenoma sebaceum in tuberous sclerosis can be shaved/excised, even under local anaesthesia if the patient is cooperative. The use of subcutaneous injection of Adrenaline diluted with saline and positioning of the operation table will very much reduce the blood loss. Similar conditions like haemangioma, naevi, pigmented sports nodules and ulcers may be treated by excision, simple closure, local flaps or full thickness skin graft.

Certain haemangioma can be treated by cryosurgery if it is available. However with the introduction of laser machine in the public service, cryosurgery is becoming much less common. The uneven surface of the skin lesions, e.g. shagreen patches in tuberous sclerosis, can be leveled by dermabrator. In this aspect the CO_2 laser machine can give better result in depth control.

The Ultrapulse CO_2 Laser machine in PMH is available in the Plastic Surgery & Burn Wards, and the Operation Theatre. The working principle is, a strong laser pulse is absorbed by water in the tissue. Heat is formed as a result and the tissue is vaporized. Because of the short duration of the laser pulse, the surrounding tissue destruction is kept to the minimal. CO_2 laser machine is used for cutting, vaporization of tissue and dermabrasion. The computer pattern generator that is coupled with the CO_2 laser machine gives even better results.

The Versapulse Laser System in PMH is available in the Plastic Surgery & Burn Wards. The machine has three different Q-switch wavelengths for pigmented lesions and a variable pulse width green wavelengths for vascular lesions. The working principle is the laser energy of a specific wavelength is absorbed by the coloured pigment in the pigmented lesion. Temperature of the pigment is raised and resulted in selective thermolysis without damage to other tissue components. Similarly, the haemoglobin of RBC in vascular lesions is damaged and lead to shrinkage and coagulation of blood vessels. Not all pigmented or vascular lesions are responsive. If they are responsive, the number of treatment sessions are different, as the thickness of these lesions are different.

There are five recognized Plastic Surgery Centres in the territories and they are KWH, PMH, PWH, QMH and TMH. Some have laser machines installed. Patients with Neurocutaneous Syndromes are welcomed to be referred to their SOPD. In many instance their ugly outlook can be improved eventually to reach their demand and satisfaction.

Neurofibromatosis Type I - The Molecular Perspective

Dr. Ivan F. M. LO
Clinical Genetic Service, Department of Health, Hong Kong

Introduction

Neurofibromatosis type I (NF1) is the most well-known neurocutaneous syndrome. There are major advances of the syndrome, with respect to the understanding of the molecular genetics involved, over the past decade. This talk will address the issues of the molecular genetic mechanisms behind NF1, genotype-phenotype correlation, and the current status of mutational analysis.

The *NF1* Gene

NF1 is a well-documented autosomal dominant disorder with a high prevalence of about 1/4000. It is of full penetrance, but highly variable expressivity. The gene is mapped to the long arm of chromosome 17 at q11.2. The *NF1* gene was identified in 1990 by positional cloning techniques. It is a large gene, spanning about 350 kb of genomic DNA and comprising 60 exons. The *NF1* gene is highly pleiotropic, which means that it can cause a wide spectrum of clinical abnormalities.

Neurofibromin

The *NF1* gene encodes a protein product known as neurofibromin. It consists of 2818 amino acids, with a molecular weight of 250 kD. Neurofibromin is widely expressed in all tissues, but it is most abundant in the central nervous system. There are four alternatively spliced isoforms with differential expression in various tissues. The functions of these isoforms remain to be elucidated, but it is possible that they contribute to the NF1 phenotype.

One important function of neurofibromin is its involvement in the Ras pathway. The Ras pathway is a signal transduction pathway that transduces signals from the plasma membrane to the nucleus via a series of effector mechanisms. The ultimate effects of Ras pathway activation are cellular growth, cellular differentiation, and promotion of cellular survival. The RAS gene itself is a proto-oncogene, i.e. somatic heterozygous mutations of which can trigger tumour formation. The normal Ras proteins play an important role in controlling cellular growth and differentiation. Ras is active only when it

is bound to guanine triphosphate (GTP). However, the Ras protein has an intrinsic GTPase activity that can hydrolyse the bound GTP to GDP (guanine diphosphate), rendering itself inactive. This GTPase activity is in turn activated by other proteins known as GTPase activating proteins (GAP). Neurofibromin has an important catalytic domain called the GAP-related domain, which is able of activating the intrinsic GTPase activity of Ras and thus down-regulating the Ras pathway. Therefore, if there is a malfunction or deficiency of neurofibromin like in NF1, the Ras pathway will be in a hyperactive state. As neurofibromin is a negative regulator of the product of a proto-oncogene, it can be regarded as a tumour suppressor. This role as a tumour suppressor has been confirmed by the demonstration of loss of heterozygosity (LOH), a hallmark of tumour suppressor gene, at the NF1 locus in neurofibromas and other NF1 associated malignant tumours. Nevertheless, the role of neurofibromin as a tumour suppressor does not suffice to account for other features of NF1, for instance, bony dysplasias, vascular dysplasias and cognitive defects. It is likely that there are other important cellular functions of neurofibromin yet to be discovered. Recently, neurofibromin was found to interact with tubulin, the building block of microtubules. This may be a clue to the other functions of neurofibromin.

NF1 Mutations

The *NF1* gene has a high mutation rate. About 50% of all germline mutations were found to be de novo, corresponding to the proportion of patients without affected parents. More than 80% of the new mutations found are of paternal origin, but paradoxically only a small paternal age effect was found. More than 80% of the germline mutations are non-sense or frameshift mutations that are expected to produce truncated, non-functional protein product. Mutational analysis of NF1 is a technically difficult and labour intensive task. The major reasons are the large size of the gene, lack of mutational hotspots, and the presence of numerous homologous loci in our genome that can interfere with the analysis. The most sensitive method thus far is the protein truncation assay, with which the mutation detection rate can be up to 70%.

Genotype-phenotype Correlation

NF1 is characterized by a remarkable degree of interfamilial and intrafamilial variability, because genotype-phenotype correlation is complicated by multiple variables. The first variable is the first

genetic hit, for example, the nature and the location of the mutation. Generally speaking, there is a lack of correlation between the first genetic hit and the NF1 phenotype. However, the fact that unique mutations were identified in some cases of spinal form NF and NF-Noonan syndrome, and that some families with familial cafe au lait spots and Watson syndrome were linked to the NF1 locus, do indicate that there is allelic heterogeneity. Further research is required to resolve this controversy. At present, the only conclusion we can draw is that there is no correlation between the first genetic hit and the NF1 phenotype except in the case of a whole gene deletion, which occurs in 4-7% of patients and causes a more severe phenotype. Somatic mosaicism of the first genetic hit further complicates the issue. The small percentage of patients with this phenomenon may have a milder manifestation, later onset of disease, and the segmental form of NF. The second variable is the second genetic hit, which, according to Knudson's two-hit theory of tumour suppressor genes, is necessary for tumour formation. Obviously, where the second hit occurs, when it occurs and how frequently it occurs will have effect on the distribution, onset and number of benign or malignant tumours. The third variable is NF1 mRNA editing, which may influence the potential of malignant transformation. The fourth variable is modifying genes. The existence of modifying genes is suggested by several observations. First, there is familial clustering of some NF1 features like seizures, optic gliomas, learning disabilities, and scoliosis. Second, male NF1 patients are more prone to develop pseudoarthrosis and myelodysplastic syndrome compared to females. Third, American black patients tend to have less optic gliomas. The fifth variable is environmental factors, such as trauma, pregnancy and oral contraceptives, which are believed to be able of triggering tumour formation in NF1 patients. In conclusion, with so many variables working together to influence the final phenotype, it is conceivable how difficult it is to predict the severity and the natural course of disease in any affected individual.

Treatment

The understanding of the molecular mechanisms of NF1 has shed light on the development of therapeutics. The finding of hyperactive Ras pathway as an important pathogenic mechanism in the development of NF1 associated tumours led researchers to consider using drugs to lower the Ras activity. An example is the farnesyltransferase inhibitors (FTI), which can inhibit the post-translational processing of the Ras proteins. FTIs are undergoing clinical trials in some cancer patients. Hopefully, they can also be tested in NF1 patients in the not too distant future.

Journal Watch

Impact of a Diagnostic Cerebrospinal Fluid Enterovirus Polymerase Chain Reaction Test on Patient Management

Ramers C, Billman G, Hartin M, et al.
JAMA 2000;283:2680-5

Introduction: More than 80% of identified cases of aseptic meningitis are caused by enterovirus, and supportive treatment is all that is required. For patients presenting with symptoms of meningitis, empiric antibiotics are often started due to difficulties in differentiating aseptic meningitis from bacterial meningitis. In this paper, the clinical implications for a rapid diagnostic test for enterovirus -- the enterovirus specific reverse transcriptase polymerase chain reaction (EV-PCR) test were examined. The sensitivity and specificity of the EV-PCR test is almost 100%. Results are available within 5-24 hours.

Objective: To assess how the EV-PCR test will affect the diagnosis and management of meningitis.

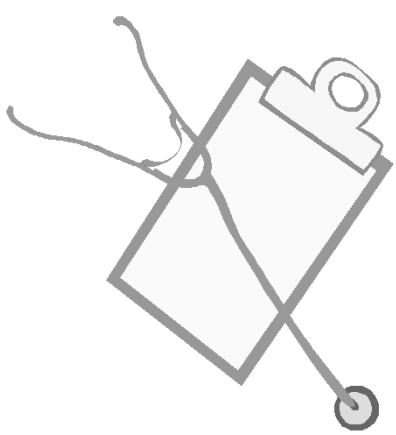
Methods: A retrospective chart review of all 276 hospitalized patients at the Children's Hospital, San Diego, California, in 1998 who received the EV-PCR test.

Results:

- 95 EV-PCR positive patients (prior to discharge) had significantly decreased usage of antibiotics, length of hospitalization, time from test result to discharge, and number of ancillary tests performed relative to the 92 EV-PCR-negative patients (prior to discharge). These differences remained statistically significant between the EV-PCR-positive patients and the EV-PCR-negative patients (prior to discharge) in the subset of patients with a discharge diagnosis of viral meningitis.
- Readmission within 14 days was found to be lower for the EV-PCR-positive patients.
- 89 patients were discharged before their EV-PCR test results were available. Fifty-two percent had a discharge diagnosis of viral meningitis.

Conclusion: The authors suggest that the EV-PCR test should be performed early in the hospitalization with fast turnaround time in order to minimize unnecessary medical tests and intervention.

(Reviewed by Dr. Philomena Tse)



Treatment of Sialorrhea with Glycopyrrolate: A Double-Blinded, Dose-Ranging Study

Mier RJ, Bachrach SJ, Lakin RC, et al.
Arch Pediatr Adolesc Med (Accepted for publication)

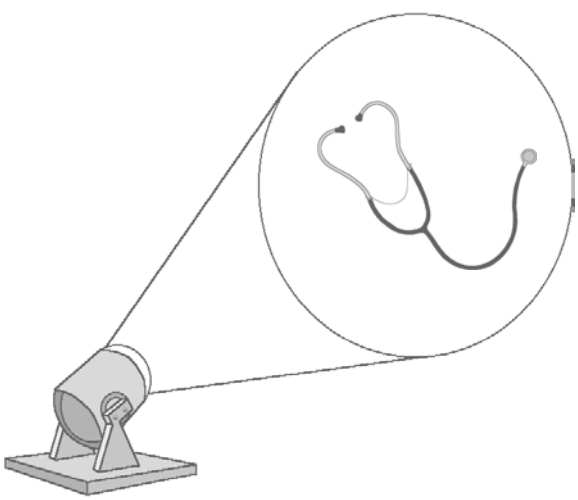
Sialorrhoea (Drooling) is a common problem in children with cerebral palsy and/or developmental disabilities. It has been estimated that about 10 percent of children with cerebral palsy have clinically-significant drooling affecting social and practical functioning. Some of these children with posterior drooling may be 'drowned' in their own secretions and have recurrent chest infection requiring repeated hospitalization and even ventilator therapy. Many treatment modalities have been used to control or diminish drooling, including medications, surgery or irradiation to eliminate gland function, and behavioral or oral motor therapies. None of these has been consistently successful. Current pharmacological intervention with anti-cholinergic agents (e.g. atropine, scopolamine etc.) have been unsatifactory because of undesirable side effects due to muscarinic stimulation. Preliminary study with Glycopyrollate (Robinol), an anti-muscuranic compound, has shown encouraging results. [1, 2] It does not cross the blood-brain barrier and has less central side effects. This study will give us new information on the effect of Glycopyrrolate on sialorrhoea.

References

1. Stern, LM. Preliminary study of glycopyrrolate in the management of drooling. J Paediatric Child Health 1997;33:52-4.
2. Bachrach SJ, Walter RS, Trzcinski K. Use of glycopyrrolate and other anticholinergic medications for Sialorrhea in children with cerebral palsy. Clin Pediatr 1998;37:485-90.

(For more information on the Management of Sialorrhoea, please visit this website: ***ihs.airweb.net****)*

(Prepared by Dr. Philomena Tse)



Negative Emotions in Children with Newly Diagnosed Epilepsy

Oostrom KJ, Schouten A, Olthof T, et al.
Epilepsia 2000;41(3):326-31

When diagnosing and treating children with epilepsy, one cannot help but wonder how the lives of these children will change. Studies in adults found that depression is common in adults who suffered from epilepsy since childhood. This recent study from the Netherlands has addressed the negative emotions associated with epilepsy in children, namely guilt and shame, which were believed to be precursors of depression in adults.

This study, being part of the Dutch Study of Epilepsy in Childhood, enrolled 36 children of age 7 to 15 years with epilepsy diagnosed not long before. Their age-matched schoolmates served as controls. It utilized structured projection, where study objects were administered a series of hypothetical situations involving a protagonist of the same sex as the objects. They were then asked to imagine how much shame and guilt the protagonist felt, and represent it by a visual analogue. The situations in the study were divided into non-illness related, illness related and epilepsy related.

This study found that *both the epileptic children and the controls* ascribed more guilt and shame to situations related to epilepsy. There was no difference between genders. The older children tend to ascribe less shame and guilt. Shame was also more predominantly reported than guilt in all situations. When the test was repeated 3 months after the first one, less shame and guilt were reported. The epileptic children did not report more shame or guilt than their healthy counterparts.

The authors argued that, despite a small sample size, this study found children, epileptic or otherwise, ascribed more shame to incompetence caused by epilepsy than other diseases. It would seem simplistic nevertheless to imply that the root of psychological impact of epilepsy was found. Being a heterogeneous disease, epilepsy will certainly have different impact on different individuals. However this study did point out the negative view about epilepsy of the public in general. Paediatric neurologists, being at the forefront of patient care, should be sensitive and compassionate to the difficulties of the child and always evoke positive attitude toward this chronic and potentially disabling disease.

(Reviewed by Dr. Wu Shun Ping)

Position Statement of HKCNDP:
Myoblast Transplant Therapy in Duchenne Muscular Dystrophy

(This Position Statement was issued at the time of
9th Asian Congress of Paediatrics in Hong Kong in March 1997)

Myoblast Transplant Therapy in Duchenne Muscular Dystrophy – Experiment, Research, but Not Cure

Duchenne Muscular Dystrophy (DMD) is one of the best known inherited muscle diseases, affecting 1 in 3,300 live male births. The gene responsible for DMD is localized in the X Chromosome at position Xp21, with its protein DYSTROPHIN being absent in patients with DMD. It is a severe disease with a progressive course, typically being fatal before the age of 20 as a result of respiratory or cardiac failure. At present there is NO established cure for the disease.

Myoblast Transplant Therapy (MTT) is a new experimental procedure which has been tried on patients with DMD. The technique involves repeated injections of donor myoblasts into various groups of muscles with the hope that they might fuse with host myoblasts, thereby inducing dystrophin production and improving muscle strength. However, MTT is still at a stage of research with much uncertainty and controversy regarding treatment effect. At this point, MTT is not and should not be offered as a miracle cure. Degenerated cardiac and respiratory muscles cannot be replaced and patients still succumb to cardiac and respiratory failure.

At present, there is no clear objective evidence supporting the efficacy of this very expensive procedure, and this therapy is not generally recommended by child neurologists. Meanwhile, patients and their families would have to bear with the significant physical, financial and psychosocial trauma that accompany such therapy should they opt for it without serious consideration.

Scientists over the world are striving to devise new techniques and therapies for managing patients with DMD. Hopefully, we will be able to discover a cure for the disease one day.

News of Society

Welcome New Members

The Society would like to warmly welcome the following new members:

Associate Members:

Dr. Estella Woo	Dr. Kan Yiu Ting

Affiliate Members:

Linda Yau	Lung Ka Wah
Yuen Yuk Wa	Charmian Mok
Law Hung Kai	Kwan Yee Man
Lee Tai Lun	

Invitation of Journal Reviews

One of the visions of the Society is to broaden the horizons of continuing medical education. While you are reading interesting and important articles in various journals and periodicals, you might have some insights and thoughts that you would like to share with your fellow colleagues.

The Society invites members to submit their reviews on journal articles to the new official publication of the Society. Your contributions to the knowledge of other members would be most highly appreciated.

Please send your reviews to Dr. Wu Shun Ping by fax at 2384 5204 or by e-mail at hkcndp@hongkong.com

Activity Announcement

8-11th December, 2000 (Friday-Monday)
Annual Scientific Meeting on Specific Learning Disorders by Professor Galaburda and Professor Camposana.

M Ground, Seminar Room, Queen Elizabeth Hospital.

12th January, 2001 (Friday)
Bimonthly Scientific Meeting on Cerebral palsy.

9th Februray, 2001 (Friday)
Case Management Study Group Meeting. Bring your cases along and discuss among colleagues.

9th March, 2001 (Friday)
Bimonthly Scientific Meeting.

20-25th September, 2002, Beijing, China
The 9th International Child Neurology Congress. This will be the first international paediatric neurology congress held in Mainland China. A major theme of the Congress will be epilepsy. For further information please refer to the Congress website www.ciccst.org.cn/icne2002, or contact the Congress Secretary, Dr. Jiang Yu-Wu, Department of Paediatrics, First Hospital, Beijing Medical University, Beijing, 100034. E-mail: icnc@public3.bta.net.cn

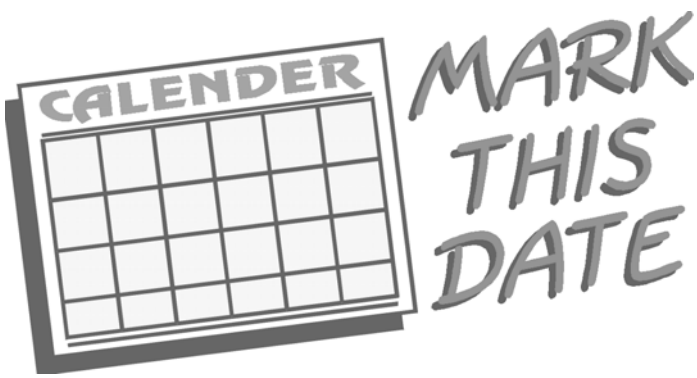
Special Announcement

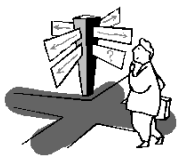
Specific Learning Disability and brain development will be in the limelight this Annual Scientific Meeting on 8-11th December, 2000. Professor Albert Galaburda and Professor Susana Camposano will be our course directors, while at the same time deliver a number of lectures on brain development and specific learning disabilities. ***Please note that*** the venue has been changed to the newly renovated **M Ground Lecture Theatre of Queen Elizabeth Hospital**.

During the Annual Scientific Meeting there are also local free paper and poster presentations. Prizes will be awarded to the best entry.

Please let your colleagues know and encourage them to come to this interesting meeting on cognitive development in children. The Society is also holding a membership drive at the same time. By joining the Society as Associate or Affiliate member the registration fee for the ASM will be waived.

Do not miss this opportunity!





Correspondence Column

The Brainchild has set a special space for fellow members to exchange ideas and express their views. Opinions and feedback regarding Society matters are most welcome. Please write to the editors at hkcndp@hongkong.com or by mail to the Society at The Federation of Medical Societies, 4/F Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, Hong Kong.

We are looking forward to hearing from you!

The Editorial Board

Wishes Everyone a Merry X'mas

and a Happy New Year 2001

Members News

Membership Drive

Both the entrance fee and the 2000 annual subscription will be waived if you register for our 2000 Annual Scientific Meeting before 1st October, 2000 and elect to join our Society at the same time, either as affiliate or associate member.

The normal entrance fee is $200 and the current annual subscription fee is $100 for affiliate member and $200 for associate member. So by registering for our 2000 ASM (registration fee: $200), you will become our member and have the opportunities to hear from Professor A. M. Galaburda, a world-renowed expert in 'Behavioural Neurology'.

Please let your colleagues know and encourage them to join this big family. A lot of education activities are waiting for you.

Membership application form can be downloaded from our website:
www.fmshk.com.hk/hkcndp

The Hong Kong Society of Child Neurology and Developmental Paediatrics
Annual Scientific Meeting - List of Course Directors

1995	*Neuro-Metabolic Diseases*	Professor Kenneth Swaiman University of Minnesota USA
1996	*Paediatric Neuro-Rehabilitation*	Dr Joe Watt University of Alberta Canada
1997	*Neonatal Neurology*	Professor Alan Hill University of British Columbia Canada
1998	*Paediatric Epilepsy*	Professor Brian Neville Institute of Child Health London, UK
1999	*Paediatric Neuro-Epidemiology*	Dr C M Verity Addenbrooke's Hospital Cambridge, UK
2000	*Behavioural Neurology*	Professor Albert M Galaburda Harvard Medical School USA

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