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Invited Lectures

Autism Spectrum Disorder – Local Perspective

SKY LIU

Senior Medical Officer, Child Assessment Service, Department of Health,
Hong Kong

The incidence of Autism Spectrum Disorder (ASD) is on a rising trend both internationally and locally. The reason is multi-factorial, which includes better public awareness, better early identification and diagnostic procedure, and other unknown reason. In this lecture, I will highlight on the following points:

1. Local epidemiology
2. Public awareness and public education
3. Local services on screening, assessment and treatment
4. Local educational services
5. Local use of Complementary and Alternative Medicine
6. Advocacy and parent work

Accommodating Children with Special Needs in Hong Kong

CCC LAM

Consultant Paediatrician, Child Assessment Service, Department of Health,
Hong Kong

A number of milestones mark Hong Kong's development in supporting children with special needs in the community.

During the early and mid 1900s, residential institutions, hospitals and special schools were opened for children with disabilities. In 1972, the then Director of Medical & Health Services invited Dr. KS Holt of the Institute of Child Health, University of London to advise of the planning of assessment services for children with disabilities who have been placed in institutions and or confined to their homes without adequate care. The Report called for medical, educational and social diagnosis, integration, interpretation, formulation and implementation by respective government departments and voluntary organizations.

In 1997, the first White Paper on Rehabilitation, Integrating the Disabled into the Community – A United Effort, was produced by the Health, Welfare and Food Bureau. This Paper drew a course for education, medical treatment and social rehabilitation services. In 1978, the Medical and Health Department introduced a comprehensive observation scheme that provided routine

observation for all infants from birth to the age of five, particularly those considered to have high risk of developing disabling conditions, such that early remedial action may be taken. From the same year, multi-disciplinary assessment centres were rolled out in stages. Children with special needs were given diagnostic formulations with follow on recommendations to meet their specific needs.

Official recognition of the need for equal opportunities and community participation for the disabled formed the next important phase in Hong Kong's history on children with special needs. In 1995, a second White Paper on Rehabilitation: Equal Opportunities and Full Participation – A Better Tomorrow for All as produced, and the Hong Kong's Disability Discrimination Ordinance (Cap. 487) (DDO) enacted. These had vast impact on how parents, advocates, service providers and the public view their rights and roles in accommodating for these children.

Until the 1990s, "children with disabilities" were largely restricted to those with serious and visible handicaps. Autism with significant core features was rising to the fore of public awareness. At that time, "learning difficulties" referred to a mixed basket of students with educational failure, and the term's specific deletion from the 1995 White Paper list of disabilities was justified by a belief that "these pupils do not typically have an impairment". Meanwhile in the late 1990s, a policy of inclusive education was set where children with significant disabilities studying in special schools were relocated to mainstream schools, championed by notions of social justice in education. There was, however, an enormous gap between such a notion and the practice in schools. Many students were indeed excluded from learning in this "inclusive" education setting. Most floundered and dropped out after the compulsory education years.

Advances in neuroscience and developmental medicine during the 1990s – the Decade of the Brain, allowed improved understanding of brain development and its disorders. Attention Deficit Hyperactivity Disorders, Specific Learning Disabilities & Dyslexia, high functioning children with Autistic Spectrum Disorder (ASD) and Specific Language Impairment in otherwise normally developing children, began to be recognized and identified for support. These students emerged from the heterogeneous group of students with "learning difficulties and maladjustment", and demanded specific and effective remediation and accommodations. In 2005, the government carried out a round of review of its rehabilitation programmes. Strong self help groups and advocates pushed successfully for ADHD and SLD to be included as

categories of disabilities in Hong Kong's rehabilitation policy. Much needed to be done during those years to deal with the situation. As example, schools and the Hong Kong Examination and Assessment Authority (HKEAA) had to face the need of providing appropriate accommodations for these students in classroom learning and open examinations. Special arrangements for admission into vocational training institutes and universities had to be developed. Specific accommodations had to be drawn up by these institutions to support the students throughout post-secondary and tertiary education.

The World Health Organization's International Classification of Functioning, Disability and Health (ICF) and its 2007 Child and Youth version (ICF-CY), provided tremendous momentum and guidance on how children with special needs could be accommodated in the community. In the last analysis, positive participation in society is highlighted. Access to appropriate educational settings, information and effective learning, technological measures for enhancing personal function, transport and housing etc., are increasingly provided. Text readers and speech to text software for students with dyslexia, visual and physical impairment, are slowly being applied in schools and examinations. Augmentative and alternative communication devices for children with severe communication disorders still require much research and developmental work.

Much work lies ahead to support our children with special needs in the community. Improvement in cross-sectoral understanding and collaboration have contributed to the advance. Growing public awareness and acceptance of individuals with special needs, along with empowerment of parents and the young persons themselves, have led to ever increasing demand on timely intervention and opportunities. Paediatricians will continue to contribute to the range of missions – providing evidence based evaluations, treatment and expert input for community services, to enable our children with special needs become part of Hong Kong's future social capital.

Aetiology of ASD – Can Genetic Studies Help?

BHY CHUNG

Associate Professor, Department of Paediatrics and Adolescent Medicine;
Department of Obstetrics & Gynaecology; Centre of Reproduction, Growth
& Development ; The University of Hong Kong, Hong Kong

Autism is a debilitating neuro-developmental disorder that is typically apparent by age 3 years. It is characterized by impaired communication, impaired reciprocal social interaction skills, and by restricted repetitive behaviors and interests. Autism spectrum disorder (ASD) is a broader phenotype which includes autism as well as less severe conditions such as Asperger syndrome and Pervasive Developmental disorder- not otherwise specified (PDD-NOS). The most recent estimate of the prevalence of autism is 3 per 1000 and rises to 6 per 1000 when all forms of ASDs are included. It occurs 4 times more commonly in males than in females.

The ASDs are etiologically heterogeneous. About 10% are associated with a Mendelian syndrome e.g. Fragile X syndrome and tuberous sclerosis complex. Another 5-7% are associated with a cytogenetically visible chromosome abnormality. Until a few years ago, the remaining is presumed to be multi-factorial and linkage scans have mapped potential candidate risk loci. More recently, array CGH studies have shown that de novo copy number variations (CNVs) occur in 7-10% of idiopathic ASD patients and that some of them have a syndromic appearance. Genes affected by CNVs are good candidates for research into ASD susceptibility. However, the complexity of autism genetics mandates that the biomedical relevance of these CNVs and the affected genes be approached in an integrated context.

Leadership in Promoting the Health of All Children: A New Mandate for Centre of Excellence on Paediatrics

CB CHOW

Hon Clinical Professor, Community Child Health Unit, Department of Paediatrics and Adolescent Medicine, The University of Hong Kong; Hon Fellow, Hong Kong Jockey Club Centre on Suicide Research and Prevention, The University of Hong Kong; Hon Consultant, Department of Paediatrics and Adolescent Medicine, Princess Margaret Hospital, Hong Kong

The challenges facing our children and families have changed enormously from that in the past. As pointed out by Lester Breslow in 2000, there are three eras of modern health care. The first era is infections (1900-1960 and beyond – the past) which focus on acute and infectious diseases centering around hospitals and doctors. Acute illnesses have been adequately prevented and controlled from improved medical care, provision of universal immunization, better public health measures and improved living standards. The second era (from 1960 and ongoing-the present) focuses on chronic diseases and disability. Major disease burdens nowadays are chronic illnesses and disabilities, injuries and psycho-social disorders. Advances in medicine have enabled us in saving more children with cancer, acute neurological insults, congenital anomalies, metabolic disorders, complex diseases contributing to increase in the number of children with chronic illnesses and disabilities. This has led to subspecialization, increased technology and multidisciplinary approach. The demographic, socio-economic status, traditional values and attitudes changes rapidly and stresses of modern technological life and economic inequality soars. We are witnessing more children born unwanted or to a teenage mother, grow up in broken families or families with income under the poverty line, influenced to smoke, and experiment with drugs and sex due to intense peer pressure and commercialization of nearly every aspect of a young person's life. Child abuse and neglect; learning, behavioural and conduct disorders, aggression, anxiety, depression, suicide etc now affect a very substantial proportion of our children with profound impact on health which can be lifelong. Recent advances in genetics and neuroscience, the better understanding on the social determinants of health as well as adverse and protective factors on trajectory of childhood development have led to the third era (2000 and beyond- the future) which focus on achieving optimal status for all children requiring investment in population-based prevention and early intervention extending well beyond the health care system.

JS PALFREY

T. Berry Brazelton Professor of Paediatrics, Harvard Medical School; Professor of Society and Human Development, Harvard School of Public Health; Director, International Pediatric Centre, Children's Hospital Primary Care Centre; Past President, American Academy of Pediatrics (AAP)

The Current Health Concerns of Children in the 21st Century – USA, Hong Kong and Global Perspective

This talk will focus on the millennial morbidities pointing out that what is causing much of the current illness in children is not traditional infectious disease but rather social determinants and life style issues. We will discuss ways in which pediatricians can help to address such problems as obesity, interpersonal violence, bullying, drug use, gun use and tobacco use.

Early Steps to Assure Positive Mental Health in Children & Adolescents

Using the Brookline Early Education Project as model, this presentation will focus on the success of empowering parents early in children's lives to serve as their first and best teachers. Early intervention of this type has impacts on mental health outcomes in young adults.

How Do We Include Children with Special Needs into Our Society?

With the history of the US special health care law as a backdrop, this presentation will review the least restrictive environment and the ways that children with disabilities can best be services in inclusive environments.

MI REIFF

Professor of Pediatrics; Director, Autism Spectrum and Neurodevelopmental Disorders Program; Director, Minnesota LEND Program; Department of Pediatrics, University of Minnesota

Is the Prevalence of Autism Spectrum Disorder Increasing?

This talk will discuss whether the prevalence of autism is increasing. What are the controversies and what are the data? The talk will discuss the epidemiology behind this, the arguments and explanations pro and con, as well as some of the possible health care and social consequences.

Update on Treatments for Autism Spectrum Disorders (ASD)

There are a wide array of treatments proposed for ASD. They are available to various extents in different communities. This talk will explore some of the most prominent behavioral, medical and proposed complementary treatments for this heterogeneous group of disorders, as well as the presence or lack of evidence behind these treatments.

Oral Presentation (Doctor's Session)

Second Malignant Neoplasms in Childhood Cancer Survivors in a Tertiary Paediatric Oncology Centre in Hong Kong

LWF SUN, FWT CHENG, V LEE, WK LEUNG, MK SHING, CK LI

Lady Pao Children's Cancer Centre, Department of Paediatrics, The Chinese University of Hong Kong, Hong Kong

Introduction: The aim of this study is to evaluate the incidence, risk factors and outcome of second malignant neoplasms in childhood cancer survivors in a tertiary paediatric oncology centre in Hong Kong.

Methods: Retrospective review of patients treated in Children's Cancer Centre in Prince of Wales Hospital between May 1984 and March 2011. Case records of patients developed second malignant neoplasms were reviewed.

Results: Total 1471 new cases aged less than 21 years old were treated in our centre in this 26-year study period. Median follow up time was 5.3 years (range 0-26.1 years). Median age of primary diagnosis was 6.3 years (range 0-20.1 years). Thirteen cases developed second malignant neoplasms with 10-year and 20-year cumulative incidence of 1.3% and 2.9% respectively. Another 4 cases were referred to us from other centres for the management of second malignant neoplasms. The median age of second malignancies was 12.9 years (range 5.5-21 years). The most common primary diagnosis was acute lymphoblastic leukemia (n=6). The most frequent second malignant neoplasms were acute leukemia or myelodysplastic syndrome (n=6) and central nervous system tumor (n=4). Median time interval between diagnosis of primary and second malignant neoplasms was 7.4 years (range 2.1-13.3 years). Median interval was shorter for second leukemia or myelodysplastic syndrome of 4.2 years compared to second solid tumor of 9.1 years. Nine patients died of progression of second malignant neoplasms, mainly resulted from second central nervous system tumor and osteosarcoma. Radiotherapy significantly increased the risk of development of second solid tumor in patients with acute lymphoblastic leukemia ($p=0.027$). Eight patients developed second solid tumor within the previous irradiated field. All patients who developed acute leukemia or myelodysplastic syndrome as second malignant neoplasms had prior use of chemotherapy with alkylating agents, topoisomerase II inhibitors or platinum compounds. Seven out of 17 patients who developed second malignant

neoplasms had a family history of cancer among the first or second-degree relatives.

Conclusion: Childhood cancer survivors were at risk of development of second malignant neoplasms. Radiotherapy was associated with second solid tumour among patients with acute lymphoblastic leukemia up to 12.3 years after completion of treatment. Patients developed second brain tumor and osteosarcoma had poor outcome.

Validation of the Paediatric Appendicitis Score in Chinese Population

CSW LIU, LY CHUNG, HY CHUNG, KY PANG, RL YIU, NSY CHAO, BPY WONG, MWY LEUNG, KKW LIU

Division of Paediatric Surgery, Department of Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Objective: Paediatric Appendicitis Score (PAS) was a scoring system created specifically for children to improve diagnostic accuracy of paediatric appendicitis. The objective is to validate the score in Chinese population.

Methods: Over an 18-month period, hospital notes of children aged 3-18 years admitted with suspected appendicitis were retrospectively reviewed. PAS upon admission was calculated with components include migration of pain (1), anorexia (1), nausea/vomiting (1), fever $>38^{\circ}\text{C}$ (1), right lower quadrant tenderness (2), guarding (2), leukocytosis $\text{WBC} \geq 10000/\text{mm}^3$ (1) and polymorphonuclear neutrophilia $\geq 7500/\text{mm}^3$ (1), comprising a total score of 10. For patients received appendectomy, the operative findings and histological diagnoses were reviewed.

Results: 214 patients were recruited, of which 82 (38.3%) underwent laparoscopic appendectomy.

The mean (median, range) PAS score for children with and without appendicitis were 6.26 (6, 3-10) and 2.69 (3, 0-9) respectively. A cutoff PAS ≤ 2 excludes appendicitis. The prevalence of appendicitis is significantly higher in patients with PAS 7-10 (94.6%) than in patients with PAS 3-6 (37.2%) ($p<0.05$). Using a cutoff PAS ≥ 7 to diagnose appendicitis, the specificity and sensitivity would be 0.98 and 0.45 respectively.

Of the 82 appendectomy done, 77 had histologically confirmed appendicitis. There was more complicated appendicitis in patients with PAS 7-10 (40.54%) than PAS 3-6 (13.3%) ($p=0.01$). The conversion rate was also higher in patients with PAS 7-10 (18.9%) than PAS 3-6 (4.4%) ($p=0.04$).

Conclusion: In Chinese locality, PAS ≥ 7 has high validity for predicting appendicitis with higher likelihood of complicated appendicitis and conversion. PAS ≤ 2 has high validity for ruling out appendicitis. Children with PAS 3-6 warrant further evaluation.

Neurocognitive Dysfunction and Grey Matter Density Deficit in Children with Obstructive Sleep Apnoea

KCC CHAN,¹ AM LI,¹ WCW CHU,² AC LIEW,³ DD RASALKAR,² L SHI,² D WANG,² YK WING⁴

¹Department of Paediatrics, ²Department of Imaging and Interventional Radiology, ⁴Department of Psychiatry, The Chinese University of Hong Kong, Hong Kong, ³School of Information and Communication Technology, Griffith University, Australia

Background: Neurocognitive dysfunction is increasingly reported in children with obstructive sleep apnoea (OSA), local data on this important issue is lacking. Cerebral structural changes related to OSA and grey matter loss has been reported in adult OSA patients; however data is scanty in childhood OSA and its association with neurocognitive dysfunction.

Objective: To compare the neurocognitive function and to investigate the difference in regional grey matter density and cerebral volume in children with and without OSA.

Materials and Methods: Fifty OSA and 27 non-OSA children underwent the neurocognitive function tests and the results were compared. High resolution 3D MR images of the brain were obtained from 23 paediatric patients with OSA (mean age \pm SD = 10.51 \pm 1.88) and 22 gender- and age- matched control subjects (mean age \pm SD = 9.93 \pm 1.70). OSA patients were further categorised into mild (N=15) and moderate-to-severe (N=8) groups based on obstructive apnea index (OAI). Total cerebral volume and regional grey matter density were analyzed using voxel-based morphometry technique and compared among groups.

Results: With analysis of variance, it was revealed that, children in the OSA group showed reduced attention and fine motor coordination scores. Trail Making Test (Part A, p-value = 0.036) and Grooved Pedboard Test (Dominant 5 rows, p-value = 0.015 and non-dominant 5 rows, p-value = 0.002) had statistically significant differences in OSA children compared with non-OSA children. Grey matter volume deficits were observed only in moderate-to-severe OSA group when comparing with controls (p<0.001, uncorrected). Significant negative correlations were found

between the Grooved Pedboard Tests and the ratio of grey matter volume over total brain volume (Pearson correlations range -0.463 to -0.520, p<0.05), i.e. the lower the grey matter volume, the longer the time to finish the test and hence the poorer the performance. However, no significant difference in the mean scores was detected between the OSA groups and the control group (ANOVA, p>0.05).

Conclusion: Children with OSA had impaired attention and fine motor coordination. Regional grey matter reduction was demonstrated in children with moderate-to-severe OSA. The grey matter deficits involved frontal and temporal regions, which are part of the limbic system and might be related to neurocognitive deficit reported in children with OSA.

One Stage Laparoscopic Assisted Endorectal Pullthrough for Hirschsprung Disease: Long Term Clinical Outcome

PMY TANG, NSY CHAO, MWY LEUNG, CSW LIU, KKW LIU

Division of Paediatric Surgery, Department of Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Background: In the recent years, one stage laparoscopic assisted endorectal pullthrough (LERP) has become the standard operation for rectosigmoid Hirschsprung disease (HD). However, few reports had addressed the long term bowel function objectively. We aim to evaluate the clinical outcomes of the one stage LERP in our centre.

Methods: Twenty-five patients (20 males, 5 females) aged 36 months or above with history of one stage LERP performed for HD were evaluated. Their clinical records were retrieved and telephone interviews were conducted.

Results: Median age of patient was of patients was 81.5 months. One patient had Down's syndrome and 1 had spinal bifida were excluded from the study. Seventeen patients (74.9%) had the aganglionic segment confined to the rectosigmoid region and 25.1% had HD affecting descending and distal transverse colon.

The median age of the time of index operation was 38 days of life. The mean length of aganglionic bowel segment was 11.6 cm and the mean resected bowel length was 15.0 cm. The median duration of regular post op anal dilatation was 10 months. Median age of regular bowel training was 24 months. 3 (13.0%) patients suffered from post-operative enterocolitis requiring hospital admission and rectal washout. 29.4% of patients fit the Rome III criteria for

constipation and 22.2% had Bristol stool consistency score less than 3. The median Rintala score was 17.5. The body weight and height percentile, age at normal bowel training, Rintala score and Bristol score shows no significant different between the patients with HD involving rectosigmoid and proximal colon. One patient had Botox injection for post-LEPT enterocolitis with improved outcome. None required re-do pullthrough or anal sphincter myomectomy.

Conclusions: The LERP is a safe and effective procedure for children with HD affecting rectosigmoid and more proximal colon. The long term clinical outcome is satisfactory.

Glucose Tolerance in Chinese Children with Obstructive Sleep Apnoea

Y ZHU,¹ AM LI,¹ CT AU,¹ J YIN,¹ AP KONG,² YK WING³

Departments of ¹Paediatrics, ²Medicine and Therapeutics, ³Psychiatry, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Objective: To examine whether childhood obstructive sleep apnoea (OSA) is associated with abnormalities in glucose tolerance.

Methods: Children with habitual snoring and symptoms suggestive of OSA were consecutively recruited. Non-snoring controls were randomly selected from participants of a community growth survey. All subjects underwent physical examination and an overnight polysomnography (PSG). OSA was diagnosed using the International Criteria of Sleep Disorders version II (ICSD-II) criteria. Normal controls denoted non-snorers whose obstructive apnea hypopnea index (OAHI) <1. Fasting serum glucose and oral glucose tolerance test (OGTT) were performed after overnight PSG.

Results: A total of 88 children were studied (median age 13.1 years; 48.9% boys; 77.3% pubertal; median body mass index (BMI) z-score 0.64), of whom 28 cases were diagnosed as OSA and 60 children were classified as normal controls. Subjects with OSA had higher glucose at 120 minutes of OGTT (6.6 (5.6-7.7) mmol/L vs 6.2 (5.4-6.8) mmol/L, P=0.045), area under the curve (AUC) glucose (814(738-948) mmol/L × min vs 779(674-842) mmol/L × min, P=0.027) and prevalence of impaired glucose tolerance (IGT) (21.4% vs 3.3%, P=0.012) than normal controls, but the two groups did not differ in fasting serum glucose. After controlling for age, gender, puberty and BMI z-score, stepwise multiple regression showed that the oxygen

saturation nadir and the percentage of total sleep time where oxygen saturation < 90% were significantly associated with both glucose levels at 120 minutes and in AUC glucose respectively, and oxygen desaturation index was significantly associated with in AUC glucose. Multiple logistic regression revealed an independent effect of the oxygen saturation nadir on the prevalence of IGT (odds ratio 0.790, P=0.024).

Conclusions: OSA has an independent impact on glucose tolerance abnormalities in Chinese children.

Predictors for Intravenous Immunoglobulin Resistance in Hong Kong Children with Kawasaki Disease

HM YOUNG, LTW CHAN, KF HUEN

Department of Paediatrics, Tseung Kwan O Hospital, Hong Kong

Kawasaki disease (KD) is not uncommon among Hong Kong children. Local descriptive study on its prevalence, clinical characteristics and outcome had been reported. Despite the fact that intravenous immunoglobulin (IVIG) treatment being a well recognized effective treatment, 10% to 20% of children will have resistance to IVIG treatment. This topic has been reported by several Asian countries but not Hong Kong in the past few years. The aim of this study was to describe characteristics of IVIG resistant patients and determine the predictive factors for the initial IVIG treatment failure.

Methods: Children who met KD diagnosis criteria and were admitted to Tseung Kwan O Hospital from 2000 to 2010 for IVIG treatment were retrospectively enrolled for analysis. Patients were divided into IVIG responsive and IVIG resistant groups. Initial response was defined as defervescence by 48 hours after start of IVIG treatment and no recur of fever (>38°C) for at least 7 days, with marked improvement or normalization of conjunctivitis, mucosal changes, rash, and extremity change. IVIG resistance was defined by persistence or recurrence of fever 48 hours after start of IVIG infusion and one or more of the initial symptoms within 2 to 7 days of treatment of IVIG. Clinical and laboratory data before IVIG treatment, as well as echocardiographic results were collected for analysis.

Results: A total of 82 children, 46 boys (56.1%) and 36 girls (43.9%) with KD were enrolled in our study. 66 (80.5%) of them were IVIG responsive, making up a 19.5% prevalence of IVIG resistance. Comparison of the principal

clinical features of KD showed no significant differences between the two groups (Table 1). Table 2 illustrated the laboratory data before treatment initiation which showed statistical significant differences in median age, sex ratio, percentage of infant KD, and illness days of initial IVIG between the two groups. Univariate analysis identified that the IVIG resistance group had initial findings of higher neutrophil percentage, lower lymphocyte percentage, higher serum albumin level and higher serum total bilirubin level than the IVIG responsive group (Table 3). We also found that incidence of coronary abnormalities at fourth week but not at one year were significantly higher among the IVIG resistant groups. However no difference was noted in gastrointestinal manifestation occurrence among the two patient groups (Table 4).

Table 1 Clinical features of the two patient groups

Clinical features	Total Number	No. of patients (%)		p-values*
		IVIG responsive	IVIG resistance	
Conjunctival injection	77	63 (95.5)	14 (87.5)	0.250
Mucosal change	76	62 (93.9)	14 (87.5)	0.332
Extremity change	68	56 (84.8)	12 (75)	0.458
Skin rash	74	58 (87.9)	16 (100)	0.344
Lymphadenopathy	54	41 (62.1)	13 (81.3)	0.148

*Fisher's Exact Test, exact sig. (2-sided)

Table 2 Demographic characteristics of the two patient groups

	IVIG responsive	IVIG resistance	p-values
Age (months)	24 (12-45)	39 (8.25-56.75)	0.730 ¹
Age <12 months	17/66 (25.8%)	5/16 (31.3%)	0.755 ²
Male sex	56.1%	56.3%	0.989 ³
Illness days of initial IVIG	5 (5-7)	5 (4-5.75)	0.127 ¹

¹Expressed as median (interquartile range), Mann-Whitney U test was used; ²Fisher's Exact test was used, exact sig. (2-sided); ³Chi square test was used

Table 3 Laboratory data of the two patient groups

	IVIG responsive	IVIG resistance	p-values
Total white cell (10 ⁹ /L)	15.2 (12.17-17.63)	13.95 (11.4-17.08)	0.377 ¹
Neutrophil (%)	66.6 (59.75-72.38)	79 (68.1-87.55)	0.015 ^{1*}
Lymphocyte (%)	24.2 (17.58-29.98)	14 (8.8-23.35)	0.024 ^{1*}
Haemoglobin (g/dL)	11.45 (+/-0.941)	11.26 (+/-1.224)	0.598 ¹
Haematocrit(%)	0.34 (0.32-0.36)	0.33 (0.30-0.36)	0.535 ¹
Platelet (10 ⁹ /L)	391 (317.5-483.75)	356.5 (394.5-518)	0.413 ¹
ALT (U/L)	43 (16.75-149)	77.5 (30.25-154.25)	0.311 ¹
Total bilirubin (umol/L)	6 (4-11)	10 (6.25-31.5)	0.033 ^{1*}
Albumin (g/L)	38.05 (+/- 4.259)	35.38 (+/- 4.097)	0.026 ^{1*}
CRP (mg/L)	100 (47.8-138.5)	117.6 (77.5-164)	0.163 ¹
ESR (mm/h)	98 (71.5-108.5)	100 (73-110)	0.592 ¹

¹Expressed as median (interquartile range), Mann-Whitney U test was used; *p<0.05.

Table 4 Comparison of coronary abnormality and GI manifestation incidences between two patient groups

	IVIG responsive	IVIG resistance	p-values
Coronary abnormality at 4 weeks	5/66	5/11	0.021 ^{1*}
Coronary abnormality at 1 year	3/66	2/11	0.229 ¹
Gastrointestinal presentation	15/66	7/16	0.117 ¹

¹Fisher's Exact Test was used, exact sig. (2-sided); *p<0.05

Discussion: 19.5% of our patients did not respond to a single dose of IVIG. This figure was comparable to the reported figures from different Asian and Caucasian countries. Recent studies have identified demographic and laboratory characteristics, including age, illness day, platelet count, erythrocyte sedimentation rate, haemoglobin concentration, C-reactive protein, lactate dehydrogenase, and alanine aminotransferase as predictors of IVIG resistance. We have identified neutrophil and lymphocyte percentages, as well as serum albumin and bilirubin levels as significant predictors. Pathophysiologically the low serum albumin could be explained by increased microvascular permeability while a high serum bilirubin might be the result of inflammation of bile duct or gallbladder. Differences in neutrophil and lymphocyte percentages were explained by changes in immune adaptation to KD. Nevertheless, the relatively small sample size of our study could have compromised the power such that other significant predictors could have been missed out. Different scoring systems had been designed to predict IVIG resistance in KD patients. We attempted to incorporate our own data into one of the scoring systems – Egami score, and found a relatively low sensitivity and specificity of 40% and 62% respectively. Racial difference might be a factor in explaining the poor applicability of this scoring system in our patients. A territory-wide multi-centered local study was published in 2005, reporting incidence, clinical characteristics, and coronary outcome on Hong Kong Kawasaki Disease children from 1994 to 2000. However, data analysis on IVIG resistance predictors was not mentioned. We hope that this study can add-in more information on our local data and a future prospective study is deemed necessary to create our own prediction scoring system for IVIG resistance or even coronary aneurysm occurrence.

Conclusion: There are more coronary abnormalities among IVIG resistant KD patients. Serum albumin and bilirubin levels, as well as neutrophil and lymphocyte percentages are independent risk factors associated with the need for IVIG re-treatment. A prospective, multi-centered local study is necessary to identify more significant predictors so as to generate a locally applicable scoring system to predict IVIG resistance in Hong Kong KD children.

Short Sleep Duration is Associated with Elevated Ambulatory Blood Pressure Independent of the Effect of Obstructive Sleep Apnoea in Normal Weight Adolescents

AM LI,¹ CT Au,¹ C NG,¹ C Ho,² YK WING²

¹Department of Paediatrics and ²Department of Psychiatry, Prince of Wales and Shatin Hospital, The Chinese University of Hong Kong, Hong Kong

Objectives: The association between sleep duration and blood pressure in adolescents has not been well studied. Existing data were inconsistent and conflicting. This is the first study to determine the association between sleep duration and 24-hour ambulatory blood pressure in adolescents.

Design: Cross-sectional case-control study

Setting: Local population-based study

Participants: Subjects aged 10-17.9 years were recruited from a school-based cohort established in 2003-2005. Those who were overweight (Body mass index >85th percentile of the local norm) and those who had an obstructive apnoea hypopnoea index (OAH) ≥ 5 were excluded from the analysis.

Main outcome measures: Subjects were invited to undergo nocturnal polysomnography (PSG) and 24-hour ambulatory blood pressure monitoring (ABPM) on the same day. Daytime and nocturnal systolic and diastolic blood pressure (SBP and DBP) readings were analysed separately. A 7-day sleep diary was mailed to subjects 1 week before the day of PSG and ABPM to measure their sleep duration. Daily sleep duration was defined as the average of nocturnal sleep duration plus the average of daytime nap duration over 1 week.

Results: Totally 143 subjects, of whom 83 were boys, with a mean (SD) age of 14.3 (1.8) years were recruited. They were divided into 3 groups according to their daily sleep duration. Subjects with shorter sleep duration had higher daytime SBP ($p < 0.001$), daytime DBP ($p = 0.005$) and nocturnal SBP ($p = 0.002$) than those with longer sleep. Similar results were found after converting the BP data into z score ($p = 0.002$, 0.004 and 0.01 respectively). Multiple linear regression analyses showed that sleep duration was negatively associated with daytime SBP [β (95%CI) = -1.15 (-2.06 to -0.24), $p = 0.014$] and nocturnal SBP [β (95%CI) = -1.95 (-2.93 to -0.96), $p < 0.001$] after adjusting for confounding factors including gender, height, waist z score, wake after sleep onset and OAH.

Conclusions: Short sleep duration was inversely associated with elevated daytime and nocturnal blood pressure in adolescents independent of the effect of OSA and obesity.

Growth and Nutritional Status of Infants with Cleft Lip and Palate Using a Multidisciplinary Approach: A Two-Year Prospective Study

GPG FUNG,¹ BPY WONG,² E POON,¹ KM CHAN,¹ CY CHOW,¹ SY WONG,¹ WM WONG,¹ HB CHAN,¹ KW LIU²

¹Department of Paediatrics and Adolescent Medicine; ²Department of Surgery, United Christian Hospital, Hong Kong

Introduction and objective: Infants with cleft lip and/or palate are at increased risk of feeding difficulty during infancy, leading to poor caloric intake and impaired growth. So far, there is little data on nutritional status on infants with cleft. Recent advances in management of these infants have led to a multidisciplinary approach with emphasis on feeding and nutrition. This study aims to investigate the serial growth and nutritional status of infants with cleft lip and palate.

Subjects and method: This is a prospective cohort study. All infants delivered between 1 October 2008 and 1 October 2010 with diagnosis of cleft lip and/or palate and managed in a tertiary referral centre in Hong Kong are included. Infants with lethal congenital anomalies and stillbirths are excluded. Anthropometric measurements including weight, height, head circumference (HC), midarm circumference (MAC); triceps and subscapular skinfold thickness were performed on all subjects at birth (or at first consultation), and repeated at regular intervals at 2 weeks, 4 weeks, then monthly during clinic visits till 6 months of age. All measurements were carried out using standardized methods. Growth and skinfold thickness curves for these infants were plotted and compared against standardized charts. Failure to thrive (FTT) is defined as body weight <3rd centile; and poor nutritional status is defined as skinfold thickness <3rd centile on more than one occasion within the six-month period. The time to catch up growth is also documented.

Results: 64 infants were recruited in the study with male to female ratio 0.88:1. 4.6% (3/64) were delivered preterm. 23% (15/64) had diagnosis of cleft lip, 50% (32/64) had cleft palate, and 27% (17/63) had cleft lip and palate. 14% (9/64) had syndromal diagnosis (66% Pierre Robin Sequence) Baseline demographics were similar for infants with different types of cleft. In infants with FTT, there is good correlation between body weight and skinfold thickness. There was no significant difference in the incidence of FTT and poor nutrition between infants with non syndromal clefts and the general population (3.6% vs 3% RR 1.17, p=0.86). Infants with syndromal clefts have a significantly higher risk of developing FTT compared to

those with non-syndromal clefts (66% vs 3.7%, RR 19, p<0.0005). Regression analysis shows that syndromal diagnoses, feeding difficulties and presence of comorbidities are significantly associated with growth and nutritional abnormalities.

Discussion and conclusion: This is one of the first studies using an objective measure (skinfold thickness) as an indicator of nutritional status for infants with cleft, and showed that with a multidisciplinary approach, most of these infants with cleft attained satisfactory growth and nutritional parameters up to six months of age. Infants with syndromal clefts, poor feeding or comorbidities should be monitored carefully with early nutritional intervention to avoid growth and nutritional abnormalities.

Poster Presentation (Doctor's Session)

Night Sweats in Children: Prevalence and Associated Factors

HK SO,¹ AM LI,¹ CT AU,¹ JH ZHANG,² J LAU,³ TF FOK,¹ YK WING²

¹Department of Paediatrics, ²Department of Psychiatry, ³Centre for Epidemiology & Biostatistics, Prince of Wales Hospital and Shatin Hospital, The Chinese University of Hong Kong, Hong Kong

Background: Night sweating (NS) is a common presenting symptom in primary care practice, but its prevalence and correlates among children has never been examined.

Objective: In this community based survey study, we aimed to examine the prevalence and associated risk factors of NS in primary school children.

Methods: Parents of children aged 6 to 12 years of age randomly chosen from 13 primary schools completed a validated questionnaire. The following information was obtained for analysis; (1) age, gender, weight, height, sleep duration (average of 7 days) and presence of any chronic medical conditions; (2) history of respiratory diseases in the past 12 months; (3) daytime behaviour, nocturnal and daytime obstructive sleep apnoea (OSA) related symptoms and (4) family information. We defined NS as night sweating occurring ≥ 1 night per week.

Results: Among 6381 children (median age 9.2 (7.7-10.7) years) with complete information on NS, 3225 were boys (50.5%). Seven hundred and forty-seven children (11.7%) were reported to have NS in the past 12 months.

Boys (67.9%) were more likely than girls to have NS ($p < 0.0001$). Children with NS were more likely to suffer from respiratory, atopic diseases and sleep disorders. In addition, they were more likely to be hyperactive and have frequent temper outbursts. By ordinal regression model, NS was significantly associated with boys, younger age, allergic rhinitis, tonsillitis and symptoms suggestive of OSA, insomnia and parasomnia.

Conclusion: NS is prevalent among school aged children and is associated with the presence of sleep related symptoms, respiratory and atopic diseases.

Nocturnal Enuresis in Children: Prevalence, Correlates and Relationship with Obstructive Sleep Apnea

MS SU,¹ AM LI,² HK SO,² CT AU,² C HO,³ YK WING³

¹Department of Respiratory Medicine, Wenzhou Medical College Affiliated Second Hospital / Yuying Children's Hospital, China; ²Department of Paediatrics, Prince of Wales Hospital; ³Department of Psychiatry, Shatin Hospital, The Chinese University of Hong Kong, Hong Kong

Objectives: To examine the prevalence and correlates of nocturnal enuresis (NE) in primary school children, and to compare prevalence of NE in children with and without obstructive sleep apnoea (OSA).

Study design: Parents of children aged 6 to 11 years completed a questionnaire which sought information on sleep related symptoms, demography, family and past medical history. Children screened to be at high risk for OSA and a randomly chosen low risk group underwent overnight polysomnography (PSG).

Results: A total of 6147 children (3032 girls) were studied. The prevalence of NE (≥ 1 wet-night/month) was 4.6% (6.7% of boys and 2.5% of girls). Boys had significantly greater prevalence across all age groups. Five hundred ninety-seven children (215 girls) underwent PSG, the prevalence of NE was not greater in children with OSA, but increased with increasing severity of OSA in girls only. Boys with NE had longer deep sleep duration. Gender and sleep related symptoms were found to be associated with NE.

Conclusions: Differential gender prevalence of NE in relation to increasing OSA severity was demonstrated in this community based study.

Brain Stem Glioma of Chinese Children in Hong Kong

MMK SHING,¹ GCF CHAN,² RCH LI,³ CK LI,¹ HL YUEN,⁴ CK LI,⁵ SC LING,⁵ CW LUK,⁴ SY HA²

Department of Paediatrics, ¹Prince of Wales Hospital; ²Queen Mary Hospital; ³Tuen Mun Hospital; ⁴Queen Elizabeth Hospital; ⁵Princess Margaret Hospital, The Chinese University of Hong Kong, The University of Hong Kong, Hong Kong Paediatric Haematology and Oncology Study Group, Hong Kong

Objective: To assess the outcome of the children with brain stem glioma.

Method: The clinical features, pathology, treatment and outcome of patients with brain stem glioma were reviewed.

Results: There were 55 patients from 1995-2009. The median age was 6.9 years of age (2.1-17.8). The male to female ratio was 1.5:1. Twenty-nine patients did not receive any biopsy or surgery. Twelve of them received radiotherapy (RT) and chemotherapy (CT); 8 patients received RT alone; 9 patients were observed without any specific treatment. Eleven patients received debulking surgery. Three of them were then observed, four patients received RT and CT, while four patients only received RT. Fourteen patients had biopsy. Three patients were then observed without any specific treatment; nine patients received RT and CT; two patients received RT only. One patient received total resection without RT or CT. The pathology of the twenty-six patients (biopsy or surgery) with the brain stem glioma were grade I: 4, II: 5, III: 4, IV: 6; not specified: 7 (WHO classification).

Outcome: Nine patients are still alive (complete response: 3, stable disease: 3, partial remission: 3). Forty-six patients died because of progressive disease. The overall survival was 36.3% at 1 year, 15.3% at two years and 13.4% at five years. Patients who had surgery (n=12) appeared to perform better than those who did not have surgery (n=43) (33.3% vs 7.5% at 5 years; $p=0.080$). Five patients who received radiotherapy (n=39) were alive at 5 years while 2 of the patients (n=16) who did not received radiotherapy was alive (13.7% vs 12.5%; $p=0.184$). Chemotherapy, in addition to RT, did not improve the outcome (chemotherapy and RT: 13.4%; RT alone 13.3%; $p=0.440$).

Conclusion: The outcome of the brain stem glioma is poor. Innovative therapy is necessary for improvement.

The First Reported Primary Haemophagocytic Lymphohistiocytosis (HLH) of Chinese Children in Hong Kong

MMK SHING, GPK CHIANG, V LEE, AWK LEUNG, FWT CHENG, CK LI

Lady Pao Children's Cancer Centre, Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Purpose of study: To assess the presenting clinical features, treatment and outcome of HLH in Hong Kong.

Methods: Retrospective chart review of the presenting clinical features, laboratory findings, treatment and outcome.

Summary of results: From 1991 to 2010, there were 10 patients with HLH in our centre. The median age was 5.5 years of age (0.1 to 14). Male to female ratio was 1:4. The presenting clinical features were persistent fever (n=10), skin rash (n=8), jaundice (n=5), lymphadenopathy (n=5), hepatosplenomegaly (n=10), convulsion (n=2), somnolence (n=1), elevated bilirubin (n=7) and liver enzymes (n=9). Haemophagocytosis was found in bone marrow (n=10) and lymph nodes (n=2). Five patients were investigated for perforin gene mutation. In one patient (age=0.1), the result showed that there was heterozygous one base pair deletion 65 deletion C in exon 2 of both our patient and her father. There was another mutation 853-855 deletion AAG in exon3 of patient and her mother. Collectively, the patient had compound heterozygous mutations in perforin gene, 853-855 deletion AAG and 65deletion C. One patient had spontaneous remission without treatment and one patient died before treatment. Two patients who were treated with VP16, ARA-C, and Danunorubicin, or VP16 and methylprednisolone died. One patient received VP16 and dexamethasone and had complete remission without recurrence. Four patients received HLH-2004 protocol (VP16, dexamethasone and cyclosporine A), achieved complete remission but two relapsed and died after further treatment. The infant with perforin gene mutation had CNS involvement of HLH (MRI and CSF cytology showed evidence of HLH). This patient received HLH-2004 treatment. There was transient improvement and then disease progression again. She received double unit cord blood transplantation. She died of severe veno-occlusive disease. The overall survival of this group of patients was 40%.

Conclusion: HLH is rare and life-threatening disease. We report the first patient in Chinese with primary HLH disease (perforin gene mutation).

Treatment Outcome of High Dose ¹³¹I-MIBG Treatment in Stage 4 Neuroblastoma in Hong Kong

WK LEUNG,¹ V LEE,¹ M KAM,² FWT CHENG,¹ MK SHING,¹ CK LI¹

¹Departments of Paediatrics, ²Department of Clinical Oncology, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Background: High dose radioactive ¹³¹I-MIBG treatment is used as targeted therapy for high risk neuroblastoma patients. We summarized the outcome of MIBG treatment in our center.

Methods: We performed a retrospective chart review of ¹³¹I-MIBG treatment in a tertiary hospital in Hong Kong. Outcome including overall and event free survival, treatment related liver and thyroid impairment were reviewed.

Results: From Aug-2003 to Mar-2011, 15 patients received ¹³¹I-MIBG therapy. All patient received one infusion except one patient received two infusions. All patients were having stage 4 disease with ¹³¹I-MIBG-avid lesions at initial diagnosis. The median age at first ¹³¹I-MIBG therapy was 3.6 years (range 2.1 to 12.2 years). Fourteen ¹³¹I-MIBG-therapies were administered as part of conditioning regimen for haematopoietic stem cell transplantation (HSCT) and 2 were given as palliative treatment. Lugol's solution was given for thyroid protection.

For ¹³¹I-MIBG therapy administered as curative treatment (n=14), the dose ranged from 9 to 12.9mCi/kg, with median dose of 12mCi/kg. Carboplatin, etoposide and melphalan was given 7 to 10 days after MIBG treatment as conditioning for HSCT. At a median follow up of 1.2 years (range 0 to 7 years), 5 out of 14 patients receiving therapeutic ¹³¹I-MIBG treatment relapsed and 4 of them died. The estimated 2-year overall and event free survival was 74.5±12.8% and 63.5±15.3% respectively.

For patients received palliative treatment, one patient succumbed 2.6 years after treatment while the other patient is alive at 3 months after therapy.

Six patients (40%) developed primary hypothyroidism with elevation of TSH at 5 months to 1 year after treatment and 3 required thyroxine replacement. No patient experienced liver derangement immediately after ¹³¹I-MIBG treatment. One patient developed veno-occlusive disease after HSCT with good recovery.

Conclusion: ¹³¹I-MIBG therapy is a safe treatment modality for high risk neuroblastoma though hypothyroidism is commonly encountered despite prophylactic iodide.

Pilot Study of Efficacy and Safety of Chinese Herbal Medicine Gualou (*Fructus Trichosanthis*) in Treating Beta-thalassaemia Intermedia Patients

WK LEUNG,¹ PC LEUNG,² CH KO,² CK LI¹

¹Department of Paediatrics, Prince of Wales Hospital, ²Institute of Chinese Medicine, The Chinese University of Hong Kong, Hong Kong

Objective: Induction of Haemoglobin F (HbF) production has been shown to be useful to improve anaemia in beta-thalassaemia intermedia (BTI) patients. Chinese herb *Fructus trichosanthis* (Gualou) was found to contain an active ingredient which was demonstrated to be effective to induce HbF production in in-vitro study. We carried out a prospective pilot study to evaluate the clinical efficacy of oral Gualou in HbF, F cell production and Hb level in BTI patients and to assess the side effect profile and safety of Gualou.

Methods: Single-centre, single-arm prospective study in a tertiary center Hong Kong. Subjects aged 5 to 12 years old received oral Gualou extract capsules 100 mg daily and subjects over 12-year old received 200 mg daily. Dose was doubled at 6 months after interim analysis and total study duration was 1 year. HbF, F-cell production, Hb level and γ -chain mRNA expression were monitored during and till 3 months after stopping of Gualou.

Results: From Nov 2008 to May 2009, 10 patients with BTI were recruited. Median age was 15.5 years (range 9.5 to 23.5 years). 4 patients were below 12-year old. There were 6 male and 4 female. Median weight was 40 kg (range 23 to 60 kg).

Baseline mean HbF, Hb and F cell count were 6.7 ± 2.5 g/dL, 8.3 ± 1.0 g/dL and $2938 \pm 993 \times 10^9/L$, respectively. At 6 and 12 months, HbF was 6.9 ± 2.5 g/dL and 6.8 ± 2.6 g/dL, Hb was 8.4 ± 1.1 g/dL and 8.3 ± 1.4 g/dL and F cell count was $3093 \pm 1042 \times 10^9/L$ and $2992 \pm 1073 \times 10^9/L$, respectively. All three parameters were not statistically different from baseline. However, for individual subject analysis, we found that five patients were responsive to Gualou treatment. Two of them belonged to the same genotype with HPFH-SEA and TTCT 41/42 deletion. Their HbF, Hb were increased by 5-7% upon treatment. The increase in HbF production were further supported by increase of γ -chain mRNA expression by about 50 fold. There was no severe adverse event during the study period in all subjects.

Conclusions: Our pilot study demonstrated oral Gualou extract was well tolerated. It was found to induce γ -chain mRNA expression in 5 patients though there was no

significant change in HbF and Hb in the whole group. Further trials of the active ingredient of Gualou and possibly at higher dosage would be needed to determine the effectiveness of this herbal medication in beta-thalassaemia intermedia patients.

Long Term Follow Up of Change of Bone Mineral Density in Transfusion Dependent Patients in Hong Kong

WK LEUNG, TF LEUNG, CK LI

Department of Paediatrics, The Chinese University of Hong Kong, Prince of Wales Hospital, Hong Kong

Objective: Low bone mineral density (BMD) is a common problem in thalassaemia major patients. Early identification and treatment can reduce symptoms and possible fractures. We report the serial change of BMD of our transfusion dependent patients over a period of 9 years.

Methods: We reviewed results of Dual energy absorptiometry (DEXA) scans of transfusion dependent patients being treated in a tertiary center in Hong Kong. Patients were grouped according to age at first scan: 10-15 years (group 1), 15-20 years (group 2) and over 20 years old (group 3). Baseline BMD of lumbar spine and hip were compared with follow up scans, which were done at 5 to 6 years (time point 1) and 8 to 9 years (time point 2) after initial scan.

Results: Since 2001, 25 patients were recruited and BMD measured. 24 and 11 patients underwent follow up scans at time point 1 and 2, respectively. There were 15 male and 10 female. At time of first scan, the median age was 17.6 years (range 10.8 to 27.8 years). There were 8, 10 and 7 patients in group 1, 2 and 3, respectively. All except 1 patient were on calcium and vitamin D supplement and 11 patients were on pamidronate (3 in age group 1 and 8 in age group 2).

At baseline, median Z-score at lumbar spine were -2.96, -2.92 and -2.59 for age group 1, 2 and 3, respectively. At time point 1 and 2, lumbar spine Z-score were -2.60 and -2.4 for group 1, -2.76 and -2.1 for group 2 and -2.14 and -1.0 for group 3, respectively. All the three age groups showed a trend of improvement over the study period though statistical significance was not reached (Time point 2 versus baseline: $p=0.120$, 0.063 and 0.075 for group 1, 2 and 3, respectively). Z-score of hip was only available for group 1 and the median value at baseline, time point 1 and

2 were -1.99, -2.51 and -1.5, respectively.

It was observed that for the subgroup of patients not receiving pamidronate, baseline lumbar spine Z-score was also significantly better at time point 2 when compared with baseline (Z-score: -1.36 versus -2.49, $p=0.025$).

Conclusions: BMD of transfusion dependent patients showed gradual improvement over long period of time. Importance of early maintenance of bone health should not be underestimated as adolescent patients in group 3 showed a trend of BMD improvement before reaching adulthood.

Exploring CCL18, Eczema Severity and Atopy

KL HON, S WANG, TF LEUNG

Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Introduction: Childhood atopic dermatitis (AD) is a distressing disease associated with pruritus, sleep disturbance and impaired quality of life. The pathophysiology of AD is complex and the chemokine CCL18/PARC (pulmonary and activation-regulated chemokine) may be involved. CCL18 binds to CLA⁺ T cells in peripheral blood of AD patients and healthy individuals and induces migration of AD-derived memory T cells in vitro. In a previous study of 79 proteins using antibody array, CCL18 was found to be a significant marker of atopy in children with AD.

Objective: To evaluate if CCL18 was associated with AD severity, quality of life, nocturnal scratching, serum eosinophil and IgE levels.

Methods: AD patients ≤ 20 years were recruited. Disease severity was assessed with the SCORing Atopic Dermatitis (SCORAD) index and Nottingham Eczema severity Score (NESS), quality of life with the Children's Dermatology Life Quality Index (CDLQI), and nocturnal scratching with a wrist motion monitor. Concentrations of plasma CCL18/PARC, serum total IgE, and eosinophil counts were measured in these patients.

Results: 108 AD patients, with mean (SD) age being 10.5 (4.4) years, were recruited. Their mean (SD) SCORAD and CDLQI were 48.7 (20.8) and CDLQI 8.7 (5.4), respectively. The mean (SD) plasma CCL18 concentration was 162.2 (129.0) pg mL^{-1} . Plasma CCL18 showed significant correlation with objective SCORAD ($r=0.440$, $p<0.001$; Figure 1), the symptoms of pruritus ($r=0.201$, $p=0.040$) and sleep loss ($r=0.193$, $p=0.049$) but not with

CDLQI or average nocturnal wrist scratching activity (Table 1). Plasma CCL18 levels were also correlated with eosinophil percentage ($r=0.373$, $p<0.001$) and IgE ($r=0.267$, $p=0.005$). When AD patients were classified by their objective SCORAD, plasma CCL18 concentrations showed a positive trend with increasing disease severity ($p<0.001$; Figure 2).

Results of methacholine bronchial challenges were available in 59 of our patients. The correlation between CCL18 and objective SCORAD was not significant in patients with ($n=22$) or without ($n=37$) bronchial hyper-reactivity (BHR).

Conclusions: Serum levels of CCL18 correlate with the clinical severity score, serum eosinophil and IgE levels. CCL18 is associated with AD and atopy.

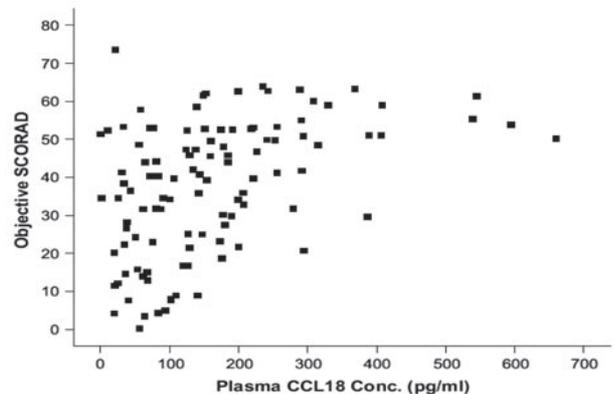


Figure 1 Scatter plot of objective SCORing Atopic Dermatitis against plasma CCL18 concentrations in our patients ($r = 0.424$, $p < 0.001$).

Table 1 Pearson correlation coefficients between clinical parameters and laboratory markers in patients with atopic dermatitis ($n = 108$).

Clinical parameters	Plasma CCL18	p
Age	-0.040	0.680
Total SCORing Atopic Dermatitis (SCORAD) index	0.424	<0.001
Objective SCORAD	0.440	<0.001
SCORAD components		
Extent	0.449	<0.001
Intensity	0.432	<0.001
Pruritus	0.201	0.040
Sleep loss	0.193	0.049
CDLQI	0.153	0.127
NESS	0.582	<0.001
IgE _{log}	0.267	0.005
Eosinophil percentage	0.373	<0.001
Average wrist activity \bar{A}	0.058	0.645

IgE_{log}, log-transformed serum total IgE level
 Pearson correlation coefficients between SCORAD and CDLQI = 0.534 ($p < 0.001$); average wrist activity and CDLQI = 0.212 ($p = 0.085$).

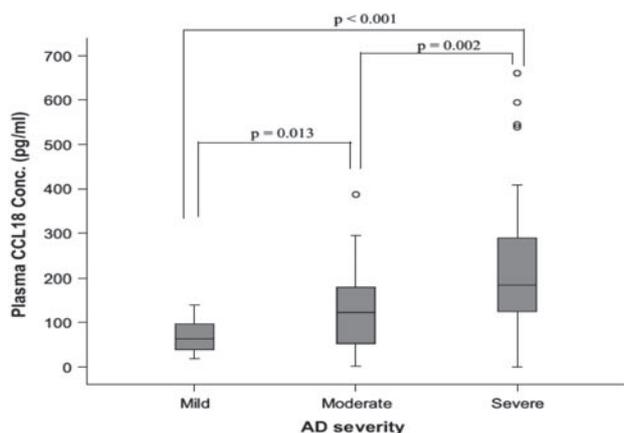


Figure 2 Distribution of plasma CCL18 concentrations in patients with different degree of atopic dermatitis severity ($p < 0.001$ for trend; data are in parametric distribution).

Influenza and Parainfluenza Associated Pediatric ICU Morbidity

KL HON, TF LEUNG

Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Introduction: Respiratory viral infections cause significant morbidity and misery. Although most infections are self-limiting and managed by the general practitioners, some children are seriously affected and require hospitalization. Among these hospital admissions, a small percentage of children would require pediatric intensive care unit (PICU) support.

Objective: To investigate if morbidity in young children admitted to a PICU with a laboratory-proven diagnosis of influenza or parainfluenza infection had increased, and if the pandemic H1N1 in 2009 had resulted in a higher morbidity.

Methods: Retrospective study from January 2003 through December 2009 was carried out. Every child in the PICU with a laboratory-confirmed influenza or parainfluenza infection was included.

Results: 18 influenza (influenza A=13 and influenza B=5) and 17 parainfluenza admissions were identified over the 7-year period (Tables 1 and 2). Parainfluenza type 3 (n=9) was the commonest subtype of parainfluenza infection. The median age of children admitted with influenza was higher than parainfluenza (4.5 versus 1.7 years, $p=0.044$). There was no significant gender predilection in the two categories. Admissions associated

with proven influenza and parainfluenza infections accounted for 2% of PICU annual admissions. There was only one death associated with influenza A in 2003. The duration of ICU stay was generally brief (median 3 days). Influenza affected older children than parainfluenza but there was no difference between the two groups of viruses in terms of morbidity. These respiratory viruses caused both upper (croup) and lower respiratory tract diseases (bronchiolitis, pneumonia). Extrapulmonary presentations such as seizures and encephalitis were less prevalent. 51% these patients required ventilatory support, 45% received systemic corticosteroids for airway obstruction such as croup, and 91% received initial broad spectrum antibiotic coverage pending cultures. Bacterial co-infections were identified in 25% of these patients. The incidence of influenza PICU admissions had not increased significantly in 2009 (H1N1 pandemic) when compared with 2003 (SARS epidemic) (Table 2, Figure 1; $p=0.3$). Using the catchment population of over 1.1 million (25% were children <12 years of age), the annual incidence of severe PICU cases of influenza and parainfluenza were 0.94 and 0.88 per 100,000 children per annum, respectively.

From April to December 2009, 2466 children under the age of 12 years with laboratory-confirmed pandemic influenza A H1N1 infection have been admitted to the Prince of Wales Hospital. Of these only two cases required PICU admission and both survived.

Conclusions: Pandemic H1N1, influenza and parainfluenza viruses may be associated with significant childhood morbidity and PICU admissions.

Table 1

PICU admissions	Total	Influenza A + B (%)	Parainfluenza (%)	Mortality
2003	122	3+0 (2.5)	0 (0)	1
2004	155	3+1 (2.6)	3 (1.9)	0
2005	111	2+1 (2.7)	5 (4.5)	0
2006	127	2+0 (1.6)	3 (2.4)	0
2007	144	0+2 (1.0)	2 (1.0)	0
2008	138	0+0 (0)	3 (2.2)	0
2009	140	3+1 (2.9)	1 (0.7)	0
Total	937	18 (1.9)	17 (1.8)	1

Table 2

Case	Influenza (n=18)	Parainfluenza (n=17)	p
Median age (yr)	4.5	1.7	0.044
Male (%)	10	8	0.44
Bacterial Co-infection	6	3	0.25
Ventilation	10	8	0.37
Systemic antibiotics	16	16	0.40
Systemic corticosteroids	7	9	0.19
Median (IQR) PICU stay (days)	3.0	3.0	0.35
Died in PICU	1	0	0.51

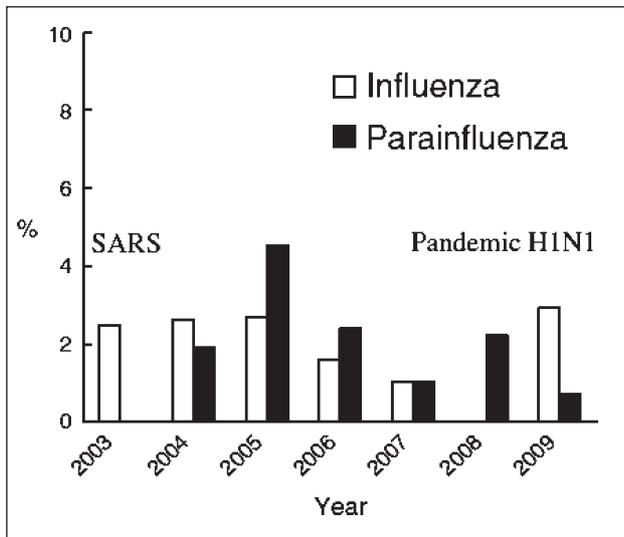


Figure 1 Influenza and parainfluenza PICU admissions

Serum Levels of Heavy Metals in Childhood Eczema and Skin Diseases: Friends or Foes?

KL HON, S WANG, TF LEUNG

Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Background: The incidence of eczema has been increasing in developed countries. Environmental and hygiene factors have been incriminated. Although air and food pollution with heavy metals have been considered as possible culprits, these factors have never been investigated in Hong Kong.

Aim: To evaluate if quality of life and eczema severity are associated with abnormal serum levels of 6 common heavy metals: cadmium, lead, mercury, selenium, copper and zinc.

Methods: Serum or whole blood was taken for measurement of 6 heavy metals from patients referred to the paediatric dermatology clinic. Eczema severity (SCORAD and NESS) and quality of life (CDLQI) were recorded.

Results: 110 patients with eczema and 41 patients with miscellaneous skin conditions were recruited. Serum levels of the 6 heavy metals were generally within the upper limits of local reference ranges (Table 1). Zinc levels were below the lower reference limit of 9.4 nmol/l in 66 eczema patients (60%) and 22 non-eczema patients (53%). Forty-four eczema patients (40%) and 24 (58%) in non-eczema group had low copper levels. In eczema patients, lead levels were generally within normal limits but their levels were

positively correlated with poor quality of life (CDLQI: $r=0.22$ and $p<0.05$), disease severity (objective SCORAD: $r=0.33$ and $p<0.005$; NESS: 0.20 , $p<0.05$), eosinophil count and log-transformed IgE (Table 2). Copper/zinc ratio also correlated with NESS and CDLQI and was higher than non-eczema skin diseases.

Conclusions: Our findings help reassure parents that levels of heavy metals generally do not exceed the local reference ranges for toxicity. However, lead levels have significant correlations with disease severity, quality of life and atopy.

This study throws light on the matter of heavy metals in children with skin diseases. It is important to evaluate whether some of the heavy metals are deficient, whilst others are genuinely harmful for patients with AD. Management is suboptimal if children with high levels of heavy metals and severe disease continue to intake the culprit element. On the other hand, parents should be discouraged from subjecting their children blindly to receive various chelation therapies or even empirical consumption of trace elements.

Table 1. Serum trace heavy metal levels in patients with and without atopic dermatitis

	Eczema (n = 110)	No eczema (n = 41)	p
Age, yr	9.9 (4.6)	11.5 (5.0)	0.078
Male, n (%)	57 (51.8)	27 (65.9)	0.123
CDLQI score*	8.6 (5.3)	4.4 (4.9)	0.001
Log-transformed total IgE	3.11 (0.80)	2.17 (0.57)	<0.001
Eosinophil percentage, %	9.1 (5.7)	4.3 (3.0)	<0.001†
<i>Serum heavy metals</i>			
Copper, μM	12.06 (3.42)	10.71 (3.89)	0.055
Zinc, μM	8.58 (2.78)	9.10 (2.55)	0.285
Copper-to-zinc ratio	1.54 (0.80)	1.22 (0.38)	0.001‡
Mercury, nm	15.07 (11.58)	13.24 (10.13)	0.294‡
Cadmium, nm§	2 (1–3)	2 (2–3)	0.250
Selenium, μM	0.91 (0.25)	0.88 (0.26)	0.454
Lead, μM	0.09 (0.04)	0.08 (0.03)	0.160‡

Results expressed in mean (standard deviation) unless stated otherwise. Significance was calculated with Student *t*-test unless stated otherwise.

Local reference ranges: copper 10.7–25.2 μM ; zinc 9.4–18.4 μM ; mercury <77 nm; cadmium <27 nm; selenium 0.44–1.43 μM for children and 0.89–1.65 μM for adults; lead <1.5 μM .

*Quality of life was measured with CDLQI for children 5–16 years of age.

†Analyzed following square-root transformation.

‡Following logarithmic transformation.

§Expressed in median (interquartile range) and analyzed by Mann–Whitney *U* test.

Table 2. Correlation between clinical parameters and laboratory tests in patients with eczema

	Log(IgE)	SCORAD	Obj. SCORAD	NESS	CDLQI
Log(IgE)	–	0.564*	0.570*	0.494*	0.300†
Eosinophil %	0.582*	0.569*	0.551*	0.394*	0.364*
Zinc	–0.063	–0.136	–0.133	–0.239§	–0.094
Copper	–0.147	–0.017	–0.023	0.005	0.118
Cu/Zn	–0.059	0.075	0.054	0.250§	0.212§
Lead	0.285†	0.329*	0.329†	0.203§	0.217§
Mercury	0.024	–0.080	–0.110	0.027	0.185
Selenium	0.107	–0.036	–0.050	0.013	0.019
Cadmium	0.216§	–0.045	–0.025	–0.192	–0.094

* $p < 0.001$; † $p < 0.005$; ‡ $p < 0.01$; § $p < 0.05$.

Analyzed by Pearson (all variables except cadmium) or Spearman correlations.

Outcome of Children with Life-Threatening Asthma Necessitating Pediatric Intensive Care

KL HON, TF LEUNG

Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Introduction: Asthma is a very common childhood condition. Acute asthmatic attacks cause significant morbidity and account for a significant number of emergency department consultations and hospital admissions. Most children admitted to the hospital because of acute asthma do not require intensive care treatment. Nevertheless, a small percentage of children with life-threatening asthma (LTA) would develop progressive respiratory failure refractory to treatment and require admission to the paediatric intensive care unit (PICU).

Objective: To report the outcome of children with LTA admitted to a university PICU.

Methods: Retrospective study between October 2002 and May 2010 was carried out. Every child with LTA and bronchospasm was included.

Results: 30 admissions of 28 patients (13 M, 17 F) were identified which accounted for 3% of total PICU admissions (n=1033) over the study period (Tables 1 and 2). The majority of patients were toddlers (median age 3.1 years). Few had past history of prematurity, lung diseases, or neurodevelopmental conditions. Approximately half had previous admissions for asthma and one-fourth with history of non-compliance to recommended treatment for asthma. One patient had parainfluenza virus and one had rhinovirus isolated. None of these factors were associated with need for mechanical ventilation (n=6 admissions). Comparing with patients who did not receive mechanical ventilation, ventilated children had significantly higher PIM2 score (1.65 versus 0.4, p<0.001), higher PCO2 levels (9.3 kPa versus 5.1 kPa, p=0.01) and longer PICU stay (median 2.5 days versus 2 days, p=0.03) The majority of patients received systemic corticosteroids, intravenous or inhaled bronchodilators. There was one pneumothorax but no death in this series.

Conclusions: LTA accounted for a small percentage of PICU admissions. Previous hospital admissions for asthma and history of non-compliance were common. Approximately one quarters required ventilatory supports. Regardless of the need for mechanical ventilation, all patients survived with prompt treatment.

Table 1: Clinical data of children with status asthmaticus admitted to PICU

Case	Total (n = 30)	Ventilated (n = 6)	Non-ventilated (n = 24)	P*
Male (%)	13 (40)	5 (75)	8 (40)	0.06
Median age (IQR), yr	3.1 (2.0-5.4)	3.3 (2.0-5.5)	3.1 (1.9-6.8)	0.84
Median (IQR) PIM2, %	0.50 (0.30-1.00)	1.65 (1.45-1.95)	0.4 (0.3-0.5)	< 0.001
Relevant risk factors				
Family history of atopy	12	1	11	0.36
Prematurity < 36 weeks (%)	4 (14)	0 (12)	4 (15)	0.57
History of chronic lung disease, bronchiolitis, pneumonia, or pneumothorax (%)	7 (17)	0 (0)	7 (25)	0.29
Neurodevelopmental delay (mental retardation, cerebral palsy, neuromuscular disease) (%)	2 (14)	0 (25)	2 (10)	1.00
Previous asthma admission (%)	16 (50)	4 (62)	12 (45)	0.66
Maintenance inhaled CS (%)	12 (35)	3 (37)	9 (35)	0.66
Non-compliance (%)	7 (25)	2 (25)	5 (25)	0.60
1 st PaCO ₂ in PICU, kPa	5.5 (4.3-7.9)	9.3 (6.4-10.9)	5.1 (4.3-5.9)	0.01
1 st PaO ₂ in PICU, kPa	9.9 (7.8-13.2)	10.1 (9.4-13.5)	9.3 (7.5-13.3)	0.40
Viral isolation (%)	2 (3)	0 (0)	2 (5)	1.00
Treatment at PICU				
Systemic CS (%)	30 (100)	6 (100)	24 (100)	N/A
Inhaled salbutamol (%)	22 (73)	5 (83)	17 (71)	1.00
Inhaled ipratropium (%)	7 (23)	0 (0)	7 (29)	0.29
Inhaled adrenaline (%)	2 (7)	1 (17)	1 (4)	0.37
Intravenous salbutamol (%)	22 (73)	5 (83)	17 (71)	1.00
Intravenous magnesium sulphate (%)	4 (13)	2 (33)	2 (8)	0.17
Systemic antibiotics (%)	19 (63)	5 (83)	14 (58)	0.37
Outcome				
Median (IQR) PICU stay, day	2.0 (1.0-2.5)	2.5 (2.0-4.5)	2.0 (1.0-2.0)	0.03
Median (IQR) hospital stay, day	5.0 (3.0-7.5)	6.5 (3.0-20.0)	5.0 (3.0-6.8)	0.27
Pneumothorax (%)	1 (3)	0 (0)	1 (4)	1.00
Died in PICU	0	0	0	N/A

CS: corticosteroid; IQR, interquartile range; N/A, not applicable. * Analyzed between ventilated and non-ventilated patients by Fisher exact test or Pearson χ^2 for categorical variables and Mann-Whitney U test for numerical variables.

Table 2: The incidence of PICU admissions for LTA during the study period

Year	PICU admissions	PICU for LTA (%)
2002 (partial data)	33	1 (3)
2003	122	7 (6)
2004	155	3 (2)
2005	111	2 (2)
2006	127	3 (2)
2007	144	5 (3)
2008	138	3 (2)
2009	140	3 (2)
2010 (partial data)	63	3 (5)

Median of 2% of annual PICU admissions were due to LTA (average absolute deviation from Median = 1.0%)

Diagnosing Late Onset Sepsis in Very Low Birth Weight Infants with Neutrophil Triggering Receptor Expressed on Myeloid Cells-1

HS LAM, RPO WONG, K LI, HM CHEUNG, PC NG

Department of Paediatrics, The Chinese University of Hong Kong, Hong Kong

Background: Despite continuing medical advancements, late-onset sepsis (LOS) remains an important cause of morbidity and mortality in very low birth weight (VLBW) infants. Current infection markers that are already in clinical use have poor sensitivities during the early phase of the infection. Triggering Receptor Expressed on Myeloid Cells-1 (TREM-1) has recently been investigated as a potential infection marker.

Objective: To evaluate whether plasma soluble TREM-1 concentration (sTREM-1) and neutrophil and monocyte surface expression of TREM-1 (nTREM-1 and mTREM-1) are useful biomarkers during the early phase of infection in VLBW infants.

Design/Methods: VLBW infants admitted to the Neonatal Intensive Care Unit at the Prince of Wales Hospital, Hong Kong with clinical features of LOS were eligible for enrolment. EDTA blood was obtained within 12 hours of onset of symptoms for sTREM-1, nTREM-1 and mTREM-1 measurement. Infants were divided into non-sepsis and sepsis groups. Receiving operating characteristics (ROC) curves were constructed for the tested biomarkers and area under the ROC curves (AUROC) calculated.

Results: 78 suspected LOS episodes were identified between September 2008 and July 2009. There were differences in surface marker values between the two groups (nTREM-1 non-sepsis = 18.8 (14.6, 31.4) vs sepsis = 7.2 (4.1, 11.7) units, $P < 0.001$). ROC curves were constructed for the biomarkers. AUROC of nTREM-1 and mTREM-1 were 0.89 and 0.66, respectively. The diagnostic utilities of neutrophil TREM-1 were the best with sensitivity of 83% and specificity of 89% (corresponding values all $< 60\%$ for the other markers).

Conclusions: nTREM-1 was a good marker in VLBW infants within the first 12 hours of onset of symptoms. nTREM-1 expression is relatively sensitive and very specific and can potentially help the clinician decide whether or not to initiate treatment in relatively low risk infants suspected of LOS where antibiotics have not already been started. Further studies are warranted.

A Rare Cause of Pleural Effusion – Idiopathic Hypereosinophilic Syndrome

AYF YIP, CW LUK, TY MIU, HL YUEN, GLH CHAN

Department of Paediatrics, Queen Elizabeth Hospital, Hong Kong

Introduction: Idiopathic hypereosinophilic syndrome (HES) is defined as sustained eosinophilia with an absolute eosinophil count $\geq 1.5 \times 10^9/L$ for more than 6 consecutive months with target organ damage but without an identifiable cause. It is a rare disorder which carries significant mortality. Skin rash and fever were the most commonly presented symptoms, followed by pulmonary & gastrointestinal presentation. Steroid is the mainstay of treatment but no standard treatment regime is available yet.

We report a case of idiopathic HES presented with vasculitis, describe its clinical course and highlight the importance of early diagnosis and aggressive treatment.

Case report: A 16-year-old previously healthy Chinese female with no contact or travel history was admitted via the Accident & Emergency (A&E) Department with a 3-week-history of abdominal pain, vomiting, diarrhoea and newly onset skin rash on both soles. There were splinter haemorrhage and tender vasculitic rash on both soles and physical examination was otherwise normal. Initial workup was unremarkable. She was given oral Imodium as symptomatic treatment but her symptoms and signs persisted. Shortly after admission she started to cough with increasing severity. On Day 8 of admission, her condition rapidly deteriorated with the development of desaturation secondary to bilateral pleural effusion and ascites requiring transferral to the Paediatrics Intensive Case Unit (PICU) for further management including oxygen supplement and thoracentesis.

Further workup showed elevated white cell count up to $28.5 \times 10^9/L$ with eosinophilia (markedly elevated up to $20 \times 10^9/L$). Other cell lines showed no abnormalities ESR and autoimmune markers were normal. Sepsis workup was negative. Bronchoscopic examination was unremarkable. Pleural fluid showed high white cell count with 80% being eosinophils. Cultures were negative for bacterial, viral, mycobacterial and fungal growth. Cytology examination showed no malignant cell. CT Abdomen showed gross ascites and diffuse bowel wall thickening. Oesophagogastroduodenoscopy showed mucosal edema over the antral area, while gastric and duodenal biopsy showed evidence of chronic inflammatory infiltrate with sprinkle eosinophils in lamina propria. Skin biopsy showed features compatible with small-vessels vasculitis. Perivascular infiltration by neutrophils, eosinophils and

lymphocytes was noted. Echocardiogram showed increased endocardium & myocardium echogenicity suggestive of underlying inflammation. The troponin I level was elevated up to 1.74 ng/mL (reference range <0.03 ng/mL), indicating myocardial injury. Liver and renal function tests were normal. Bone marrow aspiration & trephine biopsy showed features compatible with bone marrow eosinophilia. Cytogenetic study showed normal karyotype and there was no evidence of FIP1L1-PDGFR α rearrangement (rearrangement if positive represents a subset of chronic eosinophilic leukemia). Extensive investigations were done thereby ruling out reactive eosinophilia or clonal eosinophilia as the underlying cause. A diagnosis of Idiopathic hypereosinophilic syndrome (HES) was substantiated.

Intravenous Methylprednisolone 500 mg Q24H for 3 days was given with good response. Gastrointestinal symptoms and pleural effusion fully subsided. Vasculitic rash gradually faded out. No more eosinophil detected after on D3 of Methylprednisolone administration. Oral Prednisolone was given as maintenance, starting at 15 mg tds and weaned down gradually. Regular out-patient follow up with cell count monitoring showed that the patient was dependent on a low dose of oral Prednisolone 5 mg on alternate day without side effect.

Conclusion: Our case illustrated that manifestations of idiopathic hypereosinophilic syndrome may be non-specific, rapidly progressive & life threatening. Early recognition and initiation of treatment is important. Steroid is the mainstay of treatment but no standard regime is available yet. Many reports had suggested the use of oral steroid to bring about remission but we believe that intravenous Methylprednisolone therapy is the preferable option in achieving prompt control of the disease, which could be life-saving as demonstrated in our patient.

Restrictive Cardiomyopathy Secondary to Primary Carnitine Deficiency – A Case Report

AYF YIP, KL SIU, DML WONG, WH LEE

Department of Paediatrics, Queen Elizabeth Hospital, Hong Kong

Introduction: Primary carnitine deficiency is a rare metabolic disorder, affecting 1 in 40,000 to 100,000 newborn. Heart, brain & skeletal muscle are the most commonly affected organs with cardiomyopathy being the leading presenting problem. While carnitine deficiency is well known to be a cause of dilated cardiomyopathy, its

association with restrictive cardiomyopathy had not been reported.

We reported a newborn suffering from restrictive cardiomyopathy secondary to primary carnitine deficiency.

Case report: A parity 5 baby girl was born prematurely at 36 weeks of gestation by Caesarean Section due to sub-optimal continuous tomography with a body weight of 1.3 kg, which was symmetrically small for gestational age. Her Agar score was 5 at 1 minute & 9 at 5 minutes of life.

Antenatal check-up revealed that her parents were second-degree Pakistanian relatives & their third child was born prematurely at the gestational age of 28 weeks with ruptured duodenal duplication cyst. Timely operation had been performed but the child passed away on Day 68 due to uncontrolled sepsis. Parents refused post-mortem examination because of religious reason. There was no family history of cardiac or metabolic problem.

Our patient was found to have dilated bowel loops & dilated heart since 25 & 33 weeks of gestation respectively. No resuscitation was required at delivery but elective mechanical ventilation support was required soon after birth due to marked anaemia and poor cardiac function. Subtle dysmorphism (namely short crowded face, deep seated eyes, pointed chin & short neck), marked anaemia (secondary to placental haemorrhage) and distended abdomen (later confirmed to be double duodenal atresia with a segment of cystic lesion filled with bile in between) were noted at birth. Renal failure presented as oligouria & edema were noted soon after delivery. The patient was treated accordingly and she was also referred to Paediatric Surgeon & Clinical Geneticist for assessment.

Cardiac assessment done on Day 2 confirmed restrictive cardiomyopathy with the lowest fraction shortening being 9%. Intravenous Dobutamin infusion was given both to augment the cardiac function & to decrease the afterload. Extensive investigation, including sepsis & metabolic workup, confirmed the presence of primary carnitine deficiency. Molecular study showed that our patient was heterozygous for p.Arg83Leu, with codon 83 changed from CGC to CTC. Parents & sibling had been invited for screening but the offer was declined. Post-mortem examination revealed the presence of fine lipid droplets by oil red O stain. There was no viral inclusion, amyloid deposit or fibroelastosis. Electron microscopy confirmed frequent lipid droplets within the myocardial fibers & hepatocytes. These findings were commented by the histopathologist to be compatible with carnitine deficiency.

Conclusion: Restrictive cardiomyopathy accounts for 2-5% of all the paediatric cardiomyopathy, its occurrence

in neonatal population is even rarer. Extensive literature search found no report documenting casual relation between restrictive cardiomyopathy & primary carnitine deficiency. This is the first case report with clinical, biochemical & histological proof to demonstrate that primary carnitine deficiency could be result in restrictive cardiomyopathy. Empirical administration of Carnitine supplement should also be considered in restrictive cardiomyopathy until carnitine deficiency has been ruled out.

Kawasaki Disease Presenting as Intestinal Pseudo-obstruction in a Three-year-old Boy

HM YOUNG, LTW CHAN, KF HUEN

Department of Paediatrics, Tseung Kwan O Hospital, Hong Kong

Case history: A three-year-old boy presented with two-day history of generalized colicky abdominal pain and repeated vomiting of undigested food. On examination, he had high fever, his abdomen was mildly distended with sluggish bowel sounds, believed to be attributed by the drugs, including rectal dimenhydrinate and oral hyoscine given by his own doctors. On day 2, he developed skin rash, conjunctivitis, erythema over palms and cracked lips. The next day, he started to have bilious vomiting, progressive abdominal distension with abdominal pain. Abdominal X-rays showed bowel wall thickening, dilated bowels and air-fluid levels which were consistent with bowel obstruction (Figure 1). Haemoglobin level was 10.6 g/dL; white blood cell count was $15.7 \times 10^9/L$; serum albumin was 27 g/L; liver transaminase level was normal. Erythrocyte sedimentation rate was 71 mm/hr and C-reactive protein was 143 mg/L. Computed tomography of the abdomen showed generalized dilatation of both small and large bowels including the rectum, without free gas or abscess collection in the abdomen or pelvis. Echocardiogram showed no signs of coronary artery dilation. The clinical diagnosis was Kawasaki disease, complicated with intestinal pseudo-obstruction syndrome. The boy was treated with one dose of IVIG (2 g/kg body weight) and high dose aspirin (80 mg/kg body weight/day), together with a histamine-2 receptor antagonist. The gastrointestinal symptoms and signs gradually resolved. He was discharged home with low dose aspirin on day 11. Inflammatory markers normalized and serial echocardiograms showed no coronary artery dilation. Follow-up consultation on week four showed no evidence of intestinal obstruction.

Discussion: Gastrointestinal symptoms and signs comprise about 2.3% in the clinical presentations of KD. Our patient initially presented with only gastrointestinal symptoms and signs, high swinging fever along with other typical features of KD occurred two days after the initial gastrointestinal presentations. Miyake et al described seven of 310 (4.4%) of KD patients over a period of 10 years suffered from symptoms of intestinal paralysis during the acute stage. It is possible that in KD patients with acute surgical presentations, bacteria colonizing the small intestinal mucosa may produce exotoxins which act as superantigens with subsequent V[β]₂⁺ T cells expansion, which further explains why some patients with KD have gastrointestinal complaints at the onset of disease. For these reasons, our patient was covered with systemic antibiotics for possible bacterial translocation.

Concomitant use of both dimenhydrinate and hyoscine in our patient prior to admission could be a contributing factor to ileus. We should be cautious in giving antiemetics or antispasmodics. Prompt diagnosis and treatment is crucial in reducing the risk of coronary artery complications and mortality in KD. Early treatment of IVIG can reduce



Figure 1

the incidence rate of coronary artery aneurysm from 25% to ~3%. However, atypical presentation such as gastrointestinal manifestation may delay the diagnosis of KD as well as the initiation of appropriate medical treatment. High incidence rate of coronary artery involvement was shown in KD patients suffering from intestinal pseudo-obstruction. In Zulian's report, the coronary artery aneurysm rate was 50%. Surgery for KD patients with gastrointestinal symptoms and signs carries significant morbidity and mortality. Mercer et al reported four of 10 KD patients suffering from serious surgical complications were operated for various gastrointestinal presentations. The operative mortality was 25%. Thus prompt diagnosis and treatment of KD with surgical presentations may prevent unnecessary invasive laparotomy. Another area of concern is the possibility of development of bowel stricture after the acute insult. A case report by Beiler et al described a nine-month-old girl with KD developed acquired proximal jejunal ischaemic stricture two weeks after KD presentation, later requiring bowel resection. This possible late complication caused by mesenteric ischaemia may present two to four weeks after the initial onset of the disease.

Genotype and Phenotype Findings of Childhood Onset Hypophosphatasia in a Chinese Family

MK KWOK,¹ CK MA,² TS SIU,² PT CHEUNG,¹ WK POON,¹
YL TUNG,¹ S TAM,² LCK LOW¹

¹Department of Paediatrics and Adolescent Medicine; ²Division of Clinical Biochemistry, Department of Pathology, Queen Mary Hospital, The University of Hong Kong, Hong Kong

Hypophosphatasia is a metabolic bone disease caused by a loss-of-function mutation in ALPL gene, affecting both skeletal and dental mineralization. We report the phenotype and genotype findings of a family with childhood onset hypophosphatasia.

Case report: Patient 1 was the index patient. He presented with exfoliation of deciduous teeth at 12 months of age. In retrospect, his 7-year-old elder brother (patient 2) also shed his deciduous teeth early since 4 years of age. Patient 3, his younger sister, was diagnosed subsequently by screening at 18 months of age. She was totally asymptomatic at the time of diagnosis. All 3 sibs enjoyed normal growth and development. Both their height and weight were at 75th percentile. They did not have any bone pain, deformity, or fracture. Father was asymptomatic. But

paternal grandfather reported shedding of all his permanent teeth in his 30-40s. He was previously diagnosed to have low bone mineral density and was advised to do weight-bearing exercise and increase calcium intake. All patients had low serum alkaline phosphatase (ALP), high plasma pyridoxal 5'-phosphate (P5P) and urine phosphoethanolamine (PEL), and ALPL gene mutations (Table 1). Their father and paternal grandfather also carry the same c.346 G>A heterozygous mutation.

Conclusions: Premature exfoliation of deciduous teeth and low serum ALP should alert clinicians to the possibility of hypophosphatasia. Genetic diagnosis and family screening is important to identify undiagnosed affected family members.

Table 1

Patient	ALP	P5P	PEL : Creatinine ratio	Direct sequencing of ALPL gene on peripheral blood leukocyte- derived DNA
1	90	256	63.3	heterozygous mutation for c.346 G>A, p.A99T in exon 5
2	94	166	38	heterozygous mutation for c.346 G>A, p.A99T in exon 5
3	94	292	72.6	heterozygous mutation for c.346 G>A, p.A99T in exon 5

ALP=normal: 145-420U/L; P5P=normal: <109 nmol/L; PEL:Cr=normal: 2-17 umol/mmol Cr

Central Precocious Puberty in Girls Aged 6 to 8 Years and Magnetic Resonance Imaging (MRI) of the Pituitary: 11-year Experience in a Single Centre

JYL TUNG, GWK POON, AMK KWOK, PT CHEUNG, LCK
LOW

Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital,
The University of Hong Kong, Hong Kong

Introduction: Central precocious puberty (CPP) could be a phenotype of pathology in the central nervous system. While it is generally accepted that all boys with CPP and girls with CPP at less than 6 years of age should undergo brain imaging as part of the workup, there have been controversies as to the use of brain imaging in girls who develop CPP between 6 to 8 years. Existing guidelines or recommendations on this issue were mainly based on overseas experience and might not be applicable to our current practice in Hong Kong.

Objectives: To evaluate the prevalence and clinical characteristics of intracranial lesions in patients with central precocious puberty aged 6 to 8 years in a single centre in the past 11 years.

Methods: Retrospective chart review of girls with CPP and their MRI findings between year 1999 to 2009 in a single centre.

Results: One hundred and eighty-eight girls had central precocious puberty in the study period and 157 of them (83.5%) had MRI of the pituitary done as part of the workup. The reported or documented age of onset puberty ranged from 3 to 8 years and 79.6% of the girls were aged between 6 to 8 years. The prevalence of intracranial pathology among girls with CPP aged 6 to 8 years was 20.0% while among all girls with CPP aged less than 8 years, 34 girls (21.7%) were found to have intracranial pathology. These pathologies included: pituitary adenoma (n=16), pineal cyst (n=8), Rathke's cleft cysts (n=4), arachnoid cyst (n=1), intraventricular cyst (n=1), venous angioma over the left frontal lobe (n=1), hydrocephalus (n=2) and an old infarct over the frontal lobe (n=1). The two cases of hydrocephalus and the case with an old infarct were known before the onset of CPP. None of the lesions detected required further interventions with surgical removal, chemotherapy or radiotherapy within the follow-up period of 7.2 ± 3.0 years.

Conclusions: Brain imaging the girls with CPP in our centre mainly detected benign lesions not requiring any intervention during our follow-up period. Though the current data do not justify a practice of performing routine MRI for girls diagnosed to have CPP at 6 to 8 years, longer follow-up assessment of such lesions detected in childhood may be necessary before concluding on their benign outcome.

A Full-blown Case of Wolfram Syndrome in a Young Chinese Girl

GWK POON,¹ IFM LO,² TMF TONG,² PT CHEUNG,¹

AMK KWOK,¹ JYL TUNG,¹ LCK LOW¹

¹Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, The University of Hong Kong; ²Clinical Genetic Service, Department of Health, Hong Kong

Introduction: Wolfram syndrome is a rare autosomal recessive neurodegenerative disease, characterized by diabetes insipidus, diabetes mellitus, optic atrophy and deafness (DIDMOAD). Affected individuals may also have other clinical manifestations including neurological

symptoms, renal tract abnormalities and psychiatric manifestations. Diabetes mellitus and optic atrophy tend to present early at a median age of 6 and 11 years respectively. Cranial diabetes insipidus and sensorineural deafness often present in the second decade, whilst renal tract abnormalities in the third decade and neurological complications in the fourth decade. Affected individuals often die before 50 years of age.

Case report: A Chinese girl was diagnosed to have insulin-deficient diabetes mellitus at 5 years. She complained of poor vision since 3 years and was diagnosed to have bilateral optic atrophy at 9 years. MRI showed no lesion along the visual pathway. She was further diagnosed to have central diabetes insipidus and later with additional detrusor sphincter dyssynergia around her 10th birthday. MRI brain and pituitary gland were normal. Ultrasound showed bilateral distended pelvicalyceal systems and ureters despite treatment with DDAVP, but her symptoms improved with Detrusitol and Imipramine. She was also found to have mild hearing impairment and was diagnosed to have agoraphobia without panic attack in the same year requiring psychotherapy and psychiatric follow-up.

Genetic analysis: Genetic analysis of the WFS1 gene revealed two missense mutations R456H and G736S, and one nonsense mutation Q667X. Both G736S and Q667X mutations are well reported in Wolfram syndrome. The R456H mutation is known to be associated with type 1 diabetes mellitus and may have a modifying effect. The father was found to be heterozygous for the G736S missense mutation, whilst the mother was found to be heterozygous for both the Q667X and the R456H mutations.

Conclusion: It is becoming apparent that not all diabetes presenting in childhood is type 1 and rare syndromic forms, such as Wolfram syndrome, have been identified in Hong Kong Chinese. Paediatricians should consider Wolfram syndrome when a child presents with diabetes mellitus and optic atrophy.

Universal Newborn Hearing Screening Programme (UNHS) Using 2-stage AABR-AABR TPY MA, S LAM, W WONG

Department of Paediatrics, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Background: Universal newborn hearing screening programme (UNHS) using 2-stage AABR-AABR was introduced to all Hospital Authority (HA) Hospitals since

2007. The aim of this study is to report the screening programme results in a regional research hospital in the year 2009.

Methods: All neonates will underwent AABR as the initial screening in the first day of life. For newborns who did not meet the "pass" criteria, second AABR was repeated on the same day or preferably the next day before the baby was discharged. Neonates who did not pass the second screening test were referred for further evaluation.

Results: A total of 6538 newborns and 6758 screening tests were performed. The screening coverage rate for inborn infants was 99.8%; In total, 196 (3%) of the neonates did not pass the second screening test and were referred for further evaluation. Fifteen cases were found to have hearing impairment.

Conclusions: The new universal hearing screening programme is an effective method to identify congenital hearing loss with a good coverage rate and low false positive rate (2.1%). Among this cohort, the overall hearing impairment rate was 2.3 in 1000 newborns.

Epstein-Barr Virus Related Post-transplant Lymphoproliferative Disorders in Paediatric Allogeneic Haematopoietic Stem Cell Transplant Settings

FWT CHENG,¹ V LEE,¹ KF TO,² PKS CHAN,³ WK LEUNG,¹ MK SHING,¹ CK LI¹

¹Department of Paediatrics, ²Department of Anatomical and Cellular Pathology, ³Department of Microbiology, Prince of Wales Hospital, The Chinese University of Hong Kong, Hong Kong

Background: Post-transplant lymphoproliferative disease (PTLD) after haematopoietic stem cell transplantation (HSCT) is predominately derived from donor B cells and is typically occurs within the first 6 months after transplantation. PTLD is a heterogenous group of disease which is classified into 4 categories: (i) early lesions; (ii) polymorphic type; (iii) monomorphic type and (iv) classic Hodgkin lymphoma-type PTLD.

Methods: We performed a 6-year retrospective review of EBV-related PTLD in paediatric haematopoietic stem cell transplant setting. The clinical profile and outcome of this disease were reviewed.

Results: From January 2005-February 2011, one hundred and three allogeneic HSCT was performed in our centre. Six cases of EBV-PTLD were diagnosed. The incidence was 5.8%. The age ranged from 25 months to 18 years old. The

median age was 8 years old. Three patients (7.5%) received unrelated cord blood transplant, two patients (20%) received matched unrelated donor bone marrow or peripheral blood stem cell (PBSC) transplant, one patient (14.3%) received haploidentical transplant. All of them received anti-thymocyte globulin (ATG) as in-vivo T-cell depletion. The median onset time was 3.5 months post-transplant (2 months to 14 months). In five out of six patients, either donor and/or recipients were EBV seropositive and one patient was diagnosed to have primary EBV infection at 8.5 months post-transplant and had EBV reactivation at 10 months post-transplant. The most common presentations were cervical lymphadenopathies (83.3%) and fever (66.7%). Two patients (33.3%) presented with septic-like pictures with hypotension and one of them developed multiple organs failure. All of them showed EBER positive in biopsy materials. Three (50%) responded to withdrawal of immunosuppressants alone. Two of them (33.3%) responded to addition of rituximab. Majority of them (83.3%) showed remission of PTLD after treatment. One patient died of multiple organs failure before treatment started.

Conclusion: PTLD is not uncommonly seen in paediatric allogeneic HSCT setting especially in matched unrelated donor and haploidentical transplant settings. Early recognition of clinical features of PTLD and timely reduction of immunosuppressants are the key to manage this condition. Further study needs to focus on regular monitoring of plasma EBV viral load and its impact on EBV-related PTLD.

Non-invasive Image-guided Intervention for Vascular Anomalies: An Early Experience

CSW LIU, MWY LEUNG, HS FUNG, TW FAN, PCH KWOK, NSY CHAO, HY CHAN, PMY TANG, BPY WONG, KKW LIU
Division of Paediatric Surgery, Department of Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Aim of the study: Despite better understanding about the nature of vascular anomalies (VA), the management is still challenging. Image-guided intervention has become an essential component of treatment strategy in a variety of VA. We report our early experience of this non-invasive treatment approach for VA in children.

Methods: From January 2010 to January 2011, children with VA requiring image-guided interventions were recruited. According to International Society for the Study

of Vascular Anomalies (ISSVA) classification, patients with low-flow vascular malformation, including venous malformation (VM) and lymphatic malformation (LM) underwent sclerotherapy by sodium tetradecyl sulphate foam and doxycycline respectively. Perioperative ultrasonography (USG) and fluoroscopy was performed to locate the extent of lesion, facilitate outflow circulation control and prevent sclerosant extravasation. Patients with arteriovenous malformation (AVM), a high-flow vascular malformation, underwent super-selective trans-arterial embolization. Patients with haemangioma nonresponsive to propranolol were treated by USG-guided local injection of triamcinolone. Post-treatment patients were followed up clinically and radiologically.

Main results: Thirteen patients were recruited including 5 VM, 5 LM, 1 AVM and 2 haemangiomas. 53.8% were head-and-neck lesions. In addition to cosmetic concern, six patients experienced symptoms including bleeding, pain, and upper airway obstruction. Mean age at intervention was 79.3 months (range 15-215 months). Three patients underwent serial interventions owing to the size and site of lesions. Airway protection with mechanical ventilation was necessary in 5 patients with head-and-neck lesions. There were no early post-treatment complications. Mean follow-up time was 6.5 months (range 0.5-26 months). Decreased lesion size was observed in 80% of cases. All patients had symptoms improvement after intervention.

Conclusion: Image-guided intervention for vascular anomalies is safe and effective. More patients with longer follow-up are necessary to further validate these results.

Bilateral Cystic Adrenal Masses Associated with Beckwith-Wiedemann Syndrome: Explore or Not to Explore?

PMY TANG, NSY CHAO, MWY LEUNG, CSW LIU, KKW LIU

Division of Paediatric Surgery, Department of Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Aim of study: To review and discuss the diagnosis and management of neonatal presentation of Beckwith-Wiedemann Syndrome (BWS) with bilateral cystic adrenal masses.

Method: We present a case-report and review the literature on BWS presenting with antenatally diagnosed bilateral multiloculated adrenal cyst with literature review.

Main results: A 2200 g baby girl born at 33 weeks

gestation was referred to our department because of history of antenatal ultrasound finding of bilateral suprarenal mass. At presentation, she had macroglossia but no other dysmorphic feature or metabolic disturbance. Physical examination showed non-tender ballotable left-sided loin mass, and normal female genitalia with no hyperpigmentation. Ultrasound and CT abdomen during the first week of life revealed 3x3 cm multiloculated septated left adrenal cyst and 0.5x0.7 cm right adrenal cyst without calcification, with normal liver and kidneys. Her serum cortisol, ACTH, 17-OHP level and urine catecholamine were all normal. Suspicion of bilateral adrenal cystic tumours mandated consideration for laparotomy and tumour biopsy. Serial follow-up ultrasound after one month, two months and three months showed an initial enlargement of left adrenal cyst (4 cm) followed by gradual shrinkage (0.9 cm), while the right adrenal cyst had completely resolved by three months of age. The baby remained well and thriving with non-operative management.

Conclusion: This is the first reported case in literature of a neonate with BWS presenting with antenatal diagnosis of bilateral adrenal mass. Two other neonates with similar postnatal presentations had been described, one of whom underwent exploratory laparotomy for histological confirmation of benign adrenal cysts secondary to resolving neonatal adrenal haemorrhage. This report demonstrated that appropriate expectant management with close follow-up imaging may avoid unnecessary laparotomy. With the rarity of this clinical entity, however, the precise timing for the resolution of adrenal cysts in BWS remains unclear, and further study is required to formulate the ideal management plan.

Obstructive Voiding is Associated with Physiological Phimosis: Fact or Myth?

PMY TANG, NSY CHAO, MWY LEUNG, CSW LIU, KKW LIU

Division of Paediatric Surgery, Department of Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Aim of the study: For boys referred for phimosis, voiding obstruction is one of the most concerned problems. In this study, we aim to study the voiding parameters in boys with physiological phimosis.

Method: 33 boys (median age 7, range 5 to 12) with physiological phimosis (Kikiro's grade >2) were recruited

and their voiding history documented. Uroflowmetry and bladder ultrasonography were performed for post-void residual volume (PVR).

Main results: Two boys had history of daytime or diurnal incontinence. Three boys complained of daytime urinary frequency. Eleven boys (33%) had preputial ballooning during micturition. On physical examination, Kikiro's grading 3, 4 and 5 occurred in 9%, 39% and 52% of boys respectively. The maximum flow rate in voiding (Vmax) is abnormal (<10 ml/second) in 4 boys (12%), three of whom had history of daytime urinary frequency or incontinence. PVR was normal in all boys except the one with history of diurnal incontinence. The mean voiding capacity was 247 ± 119 ml, representing $87\% \pm 35\%$ of the age-corrected expected bladder capacity. Boys with preputial ballooning or Kikiro-5 grading showed no significant differences in Vmax, PVR and voiding capacity compared with other boys.

Conclusions: Obstructive voiding with abnormal uroflowmetry parameters are rarely seen in boys with asymptomatic physiological phimosis, and is not related to preputial ballooning or Kikiro's grading. However, symptoms of urinary frequency and incontinence may be associated with abnormal uroflowmetry parameters and should be further investigated.

A Modern Presentation of a Known Rarity: Twin Fetus-in-fetu Diagnosed as Fetal Abdominal Cyst on Antenatal Ultrasound

KKY PANG,¹ NSY CHAO,¹ TK TSANG,² TW FAN,² B LAU,³ SH TING,⁴ KL SIU,⁵ MWY LEUNG,¹ KKW LIU¹

¹Division of Paediatric Surgery, Department of Surgery, ²Department of Radiology and Imaging, ³Department of Obstetrics and Gynaecology, ⁴Department of Pathology, ⁵Department of Paediatrics, Queen Elizabeth Hospital, Hong Kong

Introduction: Fetus-in-fetu is a rare condition with less than 200 reported cases in the literature, with less than 100 twin fetus-in-fetu reported. Majority of this condition presented as painless retroperitoneal mass, either palpable or symptomatic with their mass effect.

We hereby report a case of twin fetus-in-fetu, which was diagnosed antenatally and the patient remained asymptomatic after birth.

Case report: A Chinese baby girl was admitted to our unit on the day of birth for antenatal diagnosis of retroperitoneal mass. This pregnancy was conceived

naturally. Early antenatal checkup was done in China and was told to be normal. However, ultrasound done in Hong Kong at term showed a 32 x 30 x 30 mm mass at left retroperitoneal region of the fetus. The baby was born 2 days later with birth weight 4.07 kg.

Apart from the abnormal antenatal ultrasound, the baby was normal on the first assessment after birth. Physical examination did not reveal abdominal distension nor palpable mass. Abdominal X-ray showed no dilated bowel or calcification. AFP and bHCG levels were normal for age. The baby tolerated full feeding.

In view of the abnormal antenatal scan, an ultrasound was performed on day 4. It showed a complex cystic mass between the spleen and the left kidney, with maximal diameter of 42 mm. There were two heterogenous masses within the cyst, each with a spine and two long bones at the position of fetal femurs. No cardiac or cranial structure was identified. Each of the masses also possessed an umbilical cord like structure but flow was not detected on doppler.

Elective laparotomy was performed on day 14, revealing a retroperitoneal mass between the left kidney and left adrenal. The mass was resected with intact capsule. The baby made good recovery from the operation and was discharged without complication.

Pathological section showed two fetiform masses, crown-rump-length measuring 37 mm and 35 mm respectively. Each mass has an umbilical cord connecting to the same placenta-like mass without chorionic villi. Within each of the masses, upper limbs, lower limbs, spine, rib cage, intestines, an ambiguous genitalia and a non-patent anus were identified. The skin covering was intact except at the cranial end. There was no skull and no heart. Microscopic examination identified striated muscles in the limbs, ganglion cells in the spinal cord, respiratory mucosa, intestinal mucosa and primitive brain tissue.

Discussion: Although regarded as a variant of mature teratoma in the current WHO classification, the etiology of fetus-in-fetu and its relationship with teratoma remained controversial. Another postulated etiology is the inclusion of one of the fetuses in a multiple pregnancy. Our case report demonstrates that despite early diagnosis and small size of the fetu and the host, there can be relatively high level of differentiation and organogenesis.

Surgical Treatment for Primary Spontaneous Pneumothorax in Paediatric Patients: Improved Outcome with Standardized Procedure

KKY PANG, BPY WONG, RL YIU, PMY TANG, NSY CHAO, MWY LEUNG, KKW LIU

Division of Paediatric Surgery, Queen Elizabeth Hospital and United Christian Hospital, Hong Kong

Aim of the study: To compare the outcomes of video assisted thoracoscopic surgery (VATS) for primary spontaneous pneumothorax (PSP) before and after the implementation of standardized surgical procedure.

Methods: From March 2004 to June 2010, a total of 27 adolescents with 31 lungs had primary VATS performed. Standardized operative procedure was implemented since September 2007. The operative procedures included:

- 1) apical resection,
- 2) pleurectomy,
- 3) mechanical abrasion and
- 4) chemical pleurodesis using minocycline at closing of the operation.

A retrospective review was carried out at a mean follow-up time 35.3 months.

Results: Before the implementation of standardized procedure, thirteen lungs were treated with VATS. Mechanical abrasion on the pleura was done to all patients, but variable surgical procedures were done in addition:

- 1) apical resection in 92%,
- 2) pleurectomy in 39%,
- 3) chemical pleurodesis in 8%.

The recurrence rate was 53.9% (n=7, out of 13). Of all the recurrences, 85.7% (n=6) needed reoperations.

Since September 2007, eighteen consecutive lungs were treated with VATS with standardized procedure. Two lungs (11.1%) developed recurrent pneumothorax but both resolved with observation only.

Conclusion: Primary spontaneous pneumothorax is a common emergency in adolescents, a patient group with inherently high recurrence rates even after VATS. The results of VATS can be improved by standardizing the surgical procedure. Routine chemical pleurodesis using minocycline appeared to give a better outcome.

Effects of Prenatal Methylmercury Exposure on Children's Neurodevelopment

KM KWOK,¹ MHM CHAN,² HS LAM,¹ CWK LAM,² HK SO,¹ AM LI,¹ HIS CHAN,² TF FOK¹

Departments of ¹Paediatrics and ²Chemical Pathology, The Chinese University of Hong Kong, Hong Kong

Introduction: In some high fish consumption populations, exposure to methylmercury (MeHg) before birth has been suggested to adversely affect childhood neurodevelopment. Fish consumption is the most common form of exposure, but evidence showing associations between prenatal MeHg and adverse neurocognitive outcomes is conflicting. Since fish consumption is common in Hong Kong, a longitudinal study was carried out to assess for any associations between prenatal MeHg exposure and adverse neurodevelopmental effects in our population.

Method: 1057 children from the original cohort were eligible for recruitment. Children with conditions that may adversely affect neurodevelopment, but was not related to mercury exposure were excluded. Prenatal MeHg exposure was determined from the cord blood at birth. Subjects were assessed by several neuropsychological tests that are related to different neurocognitive functions: Hong Kong Wechsler Intelligence Scale for Children (HK-WISC), Hong Kong List Learning Test (HKLLT), Tests for Everyday Attention for Children (TEACH), Boston Naming Test, and Grooved Pegboard Test. The associations between prenatal MeHg exposure to MeHg and the tests results were analyzed by multiple regression with adjustment of relevant covariates.

Results: 609 subjects were recruited and one subject with Noonan syndrome was excluded. HKWISC subtest Picture Arrangement was negatively associated with prenatal MeHg exposure ($\beta=-0.943$, $p=0.049$). Time per Target of subtest Sky Search in the TEACH was positively associated with prenatal MeHg exposure ($\beta=1.092$, $p=0.029$).

Conclusion: Two of twenty five subtests in five neuropsychological tests were found to have association with prenatal MeHg exposure. The effects of exposure were not consistent across subtests. It is possible that these findings were due to chance. These results do not provide any evidence on the basis of neurocognitive outcomes to suggest that restriction of current levels of fish consumption amongst pregnant women in Hong Kong is warranted.

Adverse Effects of Prenatal Methylmercury Exposure on Childhood Cardiac Autonomic Function

KM KWOK,¹ HS LAM,¹ CWK LAM,² HK SO,¹ AM LI,¹ TF FOK¹

Departments of ¹Paediatrics and ²Chemical Pathology, The Chinese University of Hong Kong, Hong Kong

Introduction: Low dose prenatal methylmercury (MeHg) exposure from fish consumption has been shown to adversely affect cardiac autonomic function in children of populations with high maternal fish consumption behaviour, as reflected by resting blood pressure and heart rate variability (HRV). Since fish consumption behaviour of pregnant women in Hong Kong is substantial, associations between prenatal exposure to MeHg and cardiac function of children should be examined.

Method: 1057 children from the original cohort were eligible for recruitment. Children with conditions that may adversely affect cardiac or neurological functions, but not related to mercury exposure were excluded. Prenatal MeHg exposure was determined from the cord blood at birth. Physical characteristics including blood pressure were measured by standardized technique. HRV was measured by the Polar RS800 Heart Rate Monitor for 5 minutes with subjects in supine position. Time domain measurement (RMSSD, pNN50, and coefficient of RR intervals) and frequency domain components (High Frequency HF, Low Frequency LF) were analysed by the HRV analysis software. Multiple regression analysis with adjustment of related covariates was performed to investigate the association between prenatal MeHg exposure and cardiac autonomic function.

Results: 608 subjects were recruited. Three subjects with heart-related problems and 1 subject with neurological problem were excluded. No association was observed between blood pressure and prenatal MeHg exposure. In boys, increased MeHg exposure was associated with decreased high frequency (HF) component and the coefficient of HF ($\beta=-0.266$, $p=0.022$, $\beta=-0.112$, $p=0.023$). Moreover, the exposure was negatively associated with RMSSD, pNN50 and the coefficient of RR intervals ($\beta=-9.696$, $p=0.048$, $\beta=-0.341$, $p=0.019$, $\beta=-0.012$, $p=0.051$). No association was observed in girls.

Conclusion: Both the time domain parameters and HF component were associated with prenatal MeHg exposure

and this suggested that the low dose prenatal MeHg exposure may be associated with reduced parasympathetic activity. Further studies to investigate the clinical relevance of these findings are warranted.

Inverted Duplication 18q12.1-q22

A LIU,¹ A HO,¹ TC YUNG,¹ T TAM,² K CHAN,² E LAU,² M TANG,² TY TAN,¹ B CHUNG^{1,2}

¹Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, The University of Hong Kong, ²Prenatal Counseling Unit and Prenatal Diagnostic Laboratory, Department of Obstetrics and Gynaecology, Tsan Yuk Hospital, The University of Hong Kong, Hong Kong

Background: Partial duplications over the long arm of Chromosome 18 have been associated with congenital heart disease, multiple dysmorphism, seizure, growth retardation and developmental delay. Genotype-phenotype correlation, however, is still controversial, as is the association with the full Trisomy 18 phenotype. We report a Chinese infant with an inverted duplication involving a novel region within Chromosome 18q.

Case report: The proposita was born to a non-consanguineous healthy Chinese couple. Dysmorphism was noted and ventricular septal defect (VSD), secundum atrial septal defect (ASD), patent ductus arteriosus (PDA) as well as coarctation of aorta were detected. Karyotyping of the patient, followed by multicolour fluorescence in-situ hybridization and multicolour banding suggested an inverted duplication of 18q12.1 to q22 over the long arm of Chromosome 18. Array comparative genomic hybridization demonstrated a copy number gain between 18q12.1 and 18q22.1 (Genomic coordinates: Chr 18: 30273585-62939673) with an estimated size of 32.67-32.74Mb. Maternal karyotype was normal, and that of the father was not available.

Discussion: The inverted duplication carried by our patient is to our knowledge, previously unreported. Features observed in the index patient were comparable to those described in patients with partial duplication involving and distal to 18q21. The absence of a classical Trisomy 18 phenotype in our patient also supported the hypothesis of proximal and distal critical regions in Edwards syndrome, with the proposed distal region lying within 18q22.3-qter.

17p13.3 Class I Microduplication in a Newborn with Microcephaly, Aortic Stenosis and Dysmorphic Facial Features

A HO,¹ A LIU,¹ KS LUN,¹ T TAM,² K CHAN,² E LAU,² M TANG,² T TAN,¹ B CHUNG^{1,2}

¹Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital;

²Prenatal Counseling Unit and Prenatal Diagnostic Laboratory, Department of Obstetrics and Gynaecology, Tsan Yuk Hospital, The University of Hong Kong, Hong Kong

Background: Deletion of chromosome 17p13.3 is known to result in Miller-Dieker Syndrome. Microduplication of the same genomic region results in a distinctive clinical syndrome (Roos et al. 2009). We report a newborn presented with dysmorphic features, microcephaly and congenital heart disease, who was confirmed to have 17p13.3 microduplication syndrome by array comparative genomic hybridization (aCGH).

Clinical information: The proband was the 2nd child of a non-consanguineous Chinese couple. He was noted to be microcephalic from fetal USG in China. He was born full term via LSCS with normal Apgar scores. His head circumference, body weight and height at birth were 31.5 cm (<3rd percentile), 3.33 kg (25-50th percentile) and 48 cm (10th percentile) respectively. Distinct facial features including flat midface, posteriorly rotated and low-set ears, fleshy earlobes and triangular chin. Echocardiogram revealed valvar aortic stenosis and tiny patent ductus arteriosus. USG brain was normal. Karyotype and FISH for Elastin gene were normal. aCGH by NimbleGen CGX-12 array identified a 790 kb copy number gain in the 17p13.3 region involving the YWHAE gene, but not the PAFAH1B1 gene.

Bruno et al (2010) proposed that there are 2 classes of 17p13.3 microduplication. Class I involves YWHAE gene, but not PAFAH1B1 gene while Class II involves PAFAH1B1 gene, with or without CRK and YWHAE genes. Class I 17p13.3 microduplication is associated with autistic features, developmental delay and subtle dysmorphic features such as pointed chin, fleshy ears and cupid bow. Microcephaly has been reported but often there is a tendency to post-natal overgrowth. Only one patient in Bruno's cohort was noted to have cardiovascular manifestations, namely aortic root dilatation and mitral valve prolapse.

Conclusion: To our knowledge, this is the first patient with 17p13.3 microduplication syndrome reported in Chinese. Growth and neuro-development need to be closely monitored. The young age of diagnosis together with the associated tendency to autism spectrum disorder pose a challenge in genetic counseling.

Infantile Systemic Hyalinosis Presenting as Multiple Joint Pain in a Pakistani Infant Girl

BHY CHUNG, E LAUSCH, A SUPERTI-FURGA

Department of Paediatrics and Adolescent Medicine, The University of Hong Kong, Hong Kong; Lausch E, Centre for Pediatrics and Adolescent Medicine, University Hospital Mathildienst, Freiburg, Germany; Superti-Furga A, Centre Hospitalier Universitaire Vaudois, University of Lausanne, Switzerland

Background: Infantile systemic hyalinosis (ISH) is a rare autosomal recessive disorder characterized by abnormal hyaline deposits in the papillary dermis and other tissues. It presents in early infancy with severe pain with movement, progressive joint contractures, thickened skin and hyperpigmented macules over bony prominence. Gingival hypertrophy, skin nodules, perianal masses are common but late findings. We report a female infant with ISH and subsequently confirmed to have known pathogenic mutations in the ANTXR2 gene.

Clinical information: The proband was the 3rd child of a Pakistani couple who were first cousins. Her elder sister passed away at 8 months with unknown cause. She was born full term by NSD with normal growth parameters and Apgar scores. Newborn examination was normal. She presented with decreased limb movement at 3 months. Clinical assessment showed that limb movement was limited by severe pain; her ears were simple but prominent and there were hyperpigmentation over the finger knuckles and ankles. Skeletal survey showed metaphyseal/submetaphyseal widening. Bone marrow examination excluded myeloproliferative disorders. The absence of fever and skin rash, normal ophthalmologic examination and normal levels of serum inflammatory markers made the diagnosis of Chronic infantile neurological, cutaneous and articular (CINCA) syndrome unlikely.

As limb pain and the skin findings could be the only findings in the early stage of ISH, genetic testing was offered to the parents with genetic counseling. A known pathogenic homozygous mutation, c.[652T>C] was found in the ANTXR2 gene causing an amino acid substitution in codon 218, p.C218R, confirming the diagnosis of ISH. The parents were carriers of the same mutation.

Conclusion: Although ISH is more common in ethnic groups with consanguineous marriage, it has been reported in Chinese. This patient developed protein-losing enteropathy at 5 months, perianal masses at 6 months and mild gingival hypertrophy at 8 months. The prognosis of patients with ISH was poor and they usually die at infancy with recurrent infections and malnutrition.

Two Chinese Patients with Loeys-Dietz Syndrome due to TGFBR2 Mutations

BHY CHUNG, SY CHAN, PC CHOW, KT WONG, KS LUN, KT CHAU, YL LAU

Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, The University of Hong Kong, Hong Kong

Background: We previously reported 6 patients with Marfan-like phenotype due to transforming growth factor b-receptor 2 (TGFBR2) mutations (Am J Med Genet Part A 149A:1452-1459). Loeys-Dietz syndrome (LDS) is a recently described autosomal dominant connective tissue disorder characterized by facial dysmorphism, cleft palate, aortic dilatation, blood vessel tortuosity and a high risk of aortic dissection. It is caused by mutations in the TGFBR1 and 2 genes. Two of the 6 patients reported in 2009 were re-assessed and confirmed to have phenotypic features of LDS.

Clinical information: K.T.H. and T.W.S. (Patient 4 and 5 in AJMG 149A:1452-1459) both presented with asymptomatic murmur and marfanoid features in their childhood. In addition they have unique craniofacial features including craniosynostosis, hypertelorism and bifid uvula. They did not fulfill the Ghent or modified Ghent criteria. Both have significant progressive aortic root dilatation requiring surgical replacement in adolescence. K.T.H. had a missense mutation c.1069G>A/p.G357R in TGFBR2 gene while T.W.S. also had a missense mutation c.973A>C/p.T325P in the same gene.

Conclusion: The arterial involvement of LDS is more extensive and the propensity to rupture is higher when compared to Marfan syndrome. A third of LDS patients can presented with aortic dissection or death before 19 years of age. The youngest age of presentation was 6 months and aortic dissection can occur when the aortic root is <4 cm. LDS patients are also prone to cerebral/abdominal arterial dissection and cervical spine instabilities. It is important to recognize LDS as a differential diagnosis of Marfan-like phenotypes so that accurate genetic counseling, lifelong surveillance and timely surgical intervention can be offered.

Astragaloside IV Enhanced Haematopoiesis via the EGFR-MEK-EKR1/2 Signalling Pathway

RX DENG,^{1,2} GCF CHAN,¹ JG SHEN,² JY YE,¹ M MA,¹ JL CHEN,¹ C LIU,² CM DEE,¹ M YANG¹

¹Department of Paediatrics and Adolescent Medicine, ²School of Chinese Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong

Background: *Radix Astragali*, a major component of Danggui Buxue Tang (DBT) decoctions, is extensively used in Chinese medicine and is known to promote haematopoiesis *in vivo* and *in vitro*. Astragaloside IV (AS-IV) is one of the major compounds of *Radix Astragali*. As the major compound involved in DBT, whether it is responsible for the haematopoiesis enhancement of DBT remains unknown. Therefore, we investigated the function of AS-IV by an *in vitro* haematopoiesis model and explored the possible underlying cell signaling mechanism.

Design and methods: Murine colony forming units (CFU) assays were used to determine the effects of AS-IV on haematopoiesis *in vitro*. K562 cells were used as a haematopoietic cell model *in vitro* to explore the possible molecular mechanisms underlying AS-IV's activity. Phosphorylation of EKR1/2 (pEKR1/2) quantification were analyzed by flow cytometry, which included: 1) a control group; 2) AS-IV treatment group; 3) inhibitor groups with PD98059 or Gefinitib; 4) and a combination treatment group of the corresponding inhibitor and AS-IV. Meanwhile, cells proliferation was analyzed using ³H-Thymidine radioactive assay and the examinations of cells' survival percentage were analyzed by Annexin V/PI staining using flow cytometry. The pEKR1/2, tEKR1/2, Bcl-2 and Bax expression were analyzed by western blot.

Results: AS-IV promoted the formation of bone marrow colony forming units (CFUs) in erythrocytes, granulocytes, monocytes *in vitro*. AS-IV also significantly enhanced CFU-megakaryocytes (MK) formation. However, CFU-MK formation was significantly reduced by PD98059, suggesting that ERK1/2 activating process was one of the main pathways of AS-IV action. In K562 cells, AS-IV also promoted cell proliferation, prolonged S-phase (DNA synthesis) and prevented cells from apoptosis. The impact of EGFR inhibitor Gefinitib on the ERK1/2 phosphorylation process was further investigated. We demonstrated that AS-IV acted via the EGFR-ERK1/2 signalling pathway, which up-regulated the anti-apoptotic protein Bcl-2 expression through the ERK1/2 phosphorylation.

Conclusion: We found that astragaloside IV promoted haematopoiesis *in vitro* and this is possibly in part via activation of the EGFR-EKR1/2 signaling pathway.

Epac Regulates Human Mesenchymal Stem Cells Homing

JL YU,¹ SK CHUNG,² GCF CHAN¹

Departments of ¹Paediatrics and Adolescent Medicine & ²Anatomy, The University of Hong Kong, Hong Kong

Background: Currently, one of the challenges confronted by the actual clinical applications of human mesenchymal stem cells (hMSCs) is how to enhance the homing and engraftment of hMSCs to the target tissues with high efficiency. To overcome such barrier, mechanisms responsible for the hMSCs homing and engraftment are emerging as one of the key research foci. As of now, the exact mechanism and soluble factors involved in hMSCs migration and adhesion have not been completely unfolded. Exchange protein directly activated by cAMP (Epac), a novel protein discovered in cAMP signaling pathway, caught our attention due to its potential role in regulating cells adhesion and migration by triggering the downstream Rap family signaling cascades. However, the real biological role of Epac in cells homing process, especially for hMSCs, remains elusive.

Objective: This study was the first one to evaluate the regulatory role of Epac in the homing process of hMSCs.

Methods: Two Epac isoforms expressions in hMSCs were detected at transcriptional and protein level by using RT-PCR, western blotting and flow cytometry respectively. Functional Epac expression was analyzed by Rap1-pull down assay. The adhesion and migration of hMSCs under the influence of Epac activation were assessed by adhesion and transwell migration assays. Immunofluorescence staining was used to visualize protein expressions and the cells morphological change.

Results: We confirmed that hMSCs expressed functional Epac and the adhesion and migration capacities of hMSCs were enhanced significantly by Epac activation. Such homing enhancement effects were associated with corresponding morphological changes induced by Epac stimulation. Furthermore, we also found stromal cell derived factor 1 (SDF-1) which is a potent chemokine in regulating hMSCs homing, could increase Epac expression levels in a time dependent manner. This Epac activation contributed directly to the chemotactic responses induced by SDF-1 suggesting Epac is involved actively in the SDF-1 signaling cascades.

Conclusion: This study revealed that Epac plays a significant role in hMSCs homing by promoting adhesion, migration and also by enhancing chemotactic effect induced by SDF-1. These research findings imply that Epac may serve as a therapeutic target to regulate hMSCs homing in the future.

Serotonin Enhanced Megakaryopoiesis, Proplatelet Formation and Stimulated Thrombopoietin Release from Human Mesenchymal Stem Cells

JY YE, GCF CHAN, M YANG

Department of Paediatrics and Adolescent Medicine, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong

Background: Serotonin (5-HT) is a monoamine that has been identified not only as a neurotransmitter, but also as a novel growth factor. It has a specific affinity for platelets and megakaryocytes in the hematopoietic system. We demonstrated previously that 5-HT enhances murine megakaryopoiesis via 5-HT₂ receptors (Yang M, et al. 1996). However, the molecular mechanism by which 5-HT regulates the growth of megakaryocytes remains unknown. In the terminal stage of mammalian megakaryocyte development, platelets are released from proplatelet protruding from megakaryocytes into bone marrow (BM) sinusoids. Cytoskeleton reorganization is suggested to be an important intracellular process for these morphologic changes. 5-HT affects not only cell proliferation, but also cell migration and remodeling by activating cytoskeleton reorganization (Matsusaka S, et al. 2005). The potential effects of serotonin on cytoskeleton reorganization in proplatelet formation and its underlying mechanisms have not been investigated.

Objectives: In the present study, we explored how 5-HT regulates megakaryopoiesis and proplatelets formation.

Methods and results: Our results showed that 5-HT significantly promoted CFU-MK formation and reduced apoptosis on megakaryocytes through phosphorylation of Akt. These effects were attenuated by addition of ketanserin, a 5-HT₂ receptor inhibitor. In addition, 5-HT was able to stimulate the F-actin reorganization in megakaryocytes through activating the p-Erk1/2 expression. Bone marrow derived mesenchymal stem cells (MSCs) played an important role in regulating megakaryopoiesis and thrombopoiesis through cell-cell interaction and by the release of thrombopoietic growth factor, such as thrombopoietin (TPO). When activated, thousands of membrane-derived microparticles (MPs) were released from MSCs, and RNA/proteins contained within these MPs could be disseminated into circulating blood. In this study, we investigated the inductive effect of 5-HT on stimulating TPO and MPs released from MSCs, and the expression of TPO contained in these MPs. According to our results, 5-HT enhanced bone marrow colony forming unit-fibroblast (CFU-F) formation and TPO production in both RNA and protein level. More interestingly, both the number

of MSC-derived MPs and the expression of MPs-capsulated TPO were considerably increased by 5-HT stimulation.

Conclusions: In summary, our findings suggested that 5-HT played an important role in megakaryopoiesis and thrombopoiesis. This effect was likely mediated via 5HT₂ receptors with subsequent activation of Akt and Erk 1/2 phosphorylation, which led to survival of megakaryocytes and proplatelets formation. 5-HT also stimulated TPO released from MSCs in both dissociative and MP-bounded form, which indirectly promoted megakaryopoiesis and thrombopoiesis.

Mesenchymal Stem Cells Enhanced Metastasis of Neuroblastoma via SDF-1/CXCR4 and SDF-1/CXCR7 Signalling

M MA, JY YE, RX DENG, CM DEE, GCF CHAN

Department of Paediatrics and Adolescent Medicine, Li Ka Shing Faculty of Medicine, The University of Hong Kong, Hong Kong

Background and objective: Bone marrow is a frequent metastatic site for neuroblastoma, a common form of childhood cancer with poor prognosis. The SDF-1/CXCR4 axis has long been proposed as an important pathway during metastasis of various cancers, including neuroblastoma (Zhang L, *et al.* 2007). But the expression and function of CXCR7, the other known receptor for SDF-1, in metastatic neuroblastoma has yet to be defined. In this study, we investigated the chemotactic effects that can induce the migration of neuroblastoma cells towards human bone marrow mesenchymal stem cells (MSCs), with specific emphasis on the above two pathways.

Materials and methods: Three human MSCs cultures from different healthy donors & 5 neuroblastoma cell lines (BE2c, BE2M17, IMR32, SK-N-LP and SH-SY5Y) originally derived from metastatic sites were used. ELISA was used to determine the concentration of SDF-1 released by MSCs in conditioned culture medium. CXCR4 and CXCR7 expression on neuroblastoma cells was determined by flow cytometer. shRNA plasmid transfection was used to establish stable CXCR4 and CXCR7 knockdown IMR32 cell lines and assessed the respective functions of these two receptors on neuroblastoma cells. Transwell migration assay, invasion and adhesion assays were utilized to test the migration, invasion and adhesion efficiency of normal or knockdown neuroblastoma cells towards MSCs under the influence of MSCs conditioned medium, SDF-1 or AMD3100 (CXCR4 inhibitor).

Results: SDF-1 was secreted by all human bone marrow derived MSCs. CXCR4 and CXCR7 were expressed on neuroblastoma cells surface. The migration of IMR32, SK-N-LP and SH-SY5Y was dramatically enhanced under treatments with culture medium from MSCs. The migration ability of CXCR4 knock down (IMR32-sh-CXCR4) but not CXCR7 knock down (IMR32-sh-CXCR7) was partially blocked under culture medium of MSCs and SDF-1. In contrary, all migration could be blocked by AMD3100. The invasion ability of IMR32-sh-CXCR4 but not IMR32-sh-CXCR7 was suppressed when treated with either culture medium of MSCs or SDF-1. All invasion activity could be blocked by AMD3100, suggesting such effect was associated with SDF-1/CXCR4 axis.

Conclusion: Our data suggested that: 1) SDF-1 was secreted by MSCs; 2) metastatic neuroblastoma cell lines expressed both CXCR4 and CXCR7, and the interactions of these receptors with SDF-1 secreted by MSCs may play an important role in the migration, invasion but not adhesion in the metastasis of neuroblastoma to bone marrow.

The Epidemiology and Outcome of Childhood Central Nervous System Germ Cell Tumour: Report from the Hong Kong Paediatric Oncology Study Group

GCF CHAN,¹ MMK SHING,² HL YUEN,³ RCH LI,⁴ CK LI,⁵ CW LUK,³ SY HA,¹ CK LI²

¹Queen Mary Hospital, The University of Hong Kong, ²Prince of Wales Hospital, The Chinese University of Hong Kong, ³Queen Elizabeth Hospital,

⁴Tuen Mun Hospital, ⁵Princess Margaret Hospital, Hong Kong

Objective: Brain tumours are the 2nd commonest group of malignancy in childhood and among them, germ cell tumours (GCT) are particularly common in Chinese children (Chan GCF, *et al.* ASPR 2011). We would like to further explore the epidemiology and outcome of central nervous system (CNS) GCT in our patients' cohort.

Materials and methods: Prospective collection of childhood pediatric brain tumors data from the 5 major Public Hospitals was performed since 1999. These 5 hospitals are the only institutes with pediatric oncology service in our locality. Standard data entry sheet was used and the data collection and entry was performed by designated data managers. The data was further crosschecked with the Hong Kong Cancer Registry database which collected all the pathology reports of cancer in the territory. CNS-GCTs were diagnosed either by histological

proof or imaging with positive serum/CSF tumor markers (i.e. AFP or B-HCG). Except for those with teratoma, most patients were treated with either BEP or SFOP chemotherapy protocols + irradiation.

Results: From Jan 1999 to Dec 2009 (11 yrs), a total of 76 cases of childhood (≤ 18 yrs) CNS-GCT were diagnosed. Their median age was 12.9 yrs and ranged from 0 to 18 yrs. A high male to female ratio was noted (M:F=62:14=4.5:1). Excluding 17 patients age >15 yrs, the incidence of CNS-GCT was 5.3/million ≤ 15 yrs children/year. Compared to the SEER data (1975-95), we have a much higher frequency locally ($<2\%$ vs 23%). The incidence was almost 6 times higher than the Western data. Within the tumour subtypes, teratoma was relatively more in infant and geminoma was more among adolescents. Overall, 55/76 (72%) patients had geminoma, 9/75 (12%, 6/9 malignant) had teratoma and other non-geminomatous GCT accounted for 12/76 (16%). The 5 yrs overall survival was 92% for geminoma and 65% for non-geminoma. Comparing the 2 treatment protocols (BEP n=25 vs SFOP n=41), there was no significant difference both in term of overall or event free survival.

Conclusion: CNS-GCT is common in our Southern Chinese patient cohort as compared to the SEER data. What accounts for such ethnic difference remains to be explored. In general, CNS-GCT, in particularly geminoma, has good outcome if treated properly with current chemotherapy protocol plus irradiation.

The Data Managers were supported by the Children's Cancer Foundation of Hong Kong.

Oral Presentation (Nurse's Session)

Holistic Fever Management in Paediatrics

KM LEE,¹ WY LEE²

¹Department of Paediatrics, St. Teresa's Hospital; ²The Hong Kong Polytechnic University, Hong Kong

Background: Fever is one of the common symptoms of disease in children. However, in the real clinical situation, it shows that many parental misconceptions of fever management may cause them to have unrealistic fear, worry and anxiety of fever because of uncontrollable fever pattern of their children. This parental exaggerated fear of fever was firstly defined as "fever phobia" in 1980 by Dr. B. D. Schmitt. As an extension of our poster presentation of the Hong Kong Paediatric Nurses Association Ltd. (HKPNA) in 2009, an educational programme of fever management was provided which explored the parental concerns and misconceptions of fever management comprehensively as well as educated the parents a holistic fever management in children.

Aims: The aims of this study were: 1) to reduce the parental fever phobia and 2) to educate the parents about the misconceptions on fever management during the hospitalization of the sick children.

Methods: This was an educational programme which involved a non-experimental pre-post comparison study. A total of 6 identical workshops were conducted to parents on every Sunday between Jan and Feb in 2010 at the St. Teresa's Hospital Children Ward. Participants were come on first-come-first-serve basis during their sick child's hospitalization. Each workshop included a leaflet, a talk and a set of semi-structured questionnaires of fever management. There were totally 30 parents (n=30) who had participated in the workshop in the end. The level of parental misconception on fever was evaluated by the questionnaires before and after the workshop. Comparisons of the categorical data were analyzed with McNemar's test and Wilcoxon Signed Ranks test while comparison of the continuous data was analyzed with paired *t*-test. A *p*-value of less than 0.05 was considered to be statistically significance.

Results: The mean age of the admitted children was 26 months \pm 12 and around 63.3% (n=19) parents with their educational levels were under the category of tertiary, university or above. Around 56.7% (n=17) parents were unable to define fever and believed that teeth's growing was the common cause of fever instead of infection prior attending to the workshop. Around 46.7% (n=14) parents

did not recognize that fever could increase heart rate and respiratory rate due to the increase of metabolism. 80% (n=24) parents also concerned about brain damage being the potential harm effect of fever which caused them with severe to mild anxious and worry of fever. Around 46.7% (n=14) parents did not recognize tepid sponge could help relieving fever with nearly 50% (n=15) giving antipyretics incorrectly regardless of medical advice. There were significant improvements of parental knowledge, misconceptions and feelings of fever management ($p<0.05$) after the workshop. Regarding to the feelings, 90% (n=27) claimed that they can manage fever with mild or even without fear and anxiety ($p=0.001$).

Conclusion: Parental misconceptions of fever management still exist nowadays. Continuous educational interventions and reinforcement are needed to dispel the parental misconceptions and fear of fever. Further community-based educational programmes of fever management are suggested to be promoted.

Reduction of Pathological Fracture in Developmental Disabilities Unit

CH Lo

Department of Paediatrics and Adolescent Medicine, Caritas Medical Centre, Hong Kong

Introduction: An increasing number of fractures were noted from 2-6 incidents per year to 9 incidents in 2003 and 2004, we have investigated and worked out some solutions for caring the children who are with extra high risk of pathological fracture.

Methods: A retrospective case control study on "Risk Factors for Pathological Long Bone Fractures in Non-ambulatory Cerebral Palsy Children" had been carried out by the Department of Paediatric and Adolescent Medicine in 2002.

Based on the result of the study, an improvement program was implemented aimed to reduce the incidence of pathological fracture in this group of children.

1. Identification of children with high risk of fracture by obtaining previous history of fracture, assess for presence of spasticity or contracture, and children with low Body Mass Index (BMI).
2. Measures included re-organization of daily care activities.
3. Revision and standardization of manual handling technique and use of assistive devices for transfer.

4. Nutritional supplement: Calcium D3, maintain weight-for-height ratio >3 rd percentile as much as possible.
5. Encourage weight-bearing exercise if possible.
6. Perform bone density scan for children with repeated episodes of fracture and provided a course of Pamidronate therapy.

Results: The result of the study by the Department of Paediatric and Adolescent Medicine in 2002 showed that among the population of 235 children, 47% of them had a weight below 5th percentile which contributed one of the significant risk factors of malnutrition. Over 80% of pathological fracture happened in femur. The improvement program was effective in preventing first fracture of lifetime and significantly reduced the rate of fracture (including first fracture and recurrent fracture) to 2-3 episodes per year in the following years. Intravenous Pamidronate was given to 4 cases with bone density increased 19-42%.

Conclusion: The result of study in 2002 showed that the predictive risk factors of fracture are low BMI, significant contracture of extremities and prolonged immobilization. Pathological fracture of long bones in non-ambulatory children with cerebral palsy is common. The implementation of improvement program successfully reduced the rate of fracture in the following years. History of previous fracture was the major predisposing factor to subsequent fracture, despite minimal handling.

Paediatric Nurse Clinic in the Care of Asthma Children with Their Family

SP LEUNG, SY LAM, GPW LEE, KF LAU, YS LUI

Department of Paediatrics and Adolescent Medicine, Tuen Mun Hospital, Hong Kong

Objectives: To evaluate the effectiveness of the Paediatric Nurse Clinic for the care of asthma children.

Methods: The Paediatric Nurse Clinic (Respiratory Care) commenced in June, 2009 to provide holistic care for asthma children and their family members. It is also to provide continuous assessment and progress supervision of children, so as to reduce re-admission rate of asthma attack and provide knowledge to them.

The services provide as followings:

1. Health assessment with diagnostic test
2. Health education of asthma
3. Nurse consultation/counselling
4. Care planning and intervention
5. Phones follow up

6. Case follow up
7. Refer case to other health care professional if necessary

Results: There are 166 children recruited to follow up at Paediatric Nurse Clinic in June 2009 to March 2011.

The outcome measures have been made in the followings:

1. Satisfaction survey by patients or family members (average score): 9.45 out of 10.
2. Assess asthma knowledge by questionnaires (average score): 11.3 out of 12.
3. Re-admission rate for asthma discharge patient (within 28 days): 0 %
4. Range of overall admission rate of asthma children every 3 month monitoring: 0-10%.
5. Compare the numbers of admission before and after attending the Paediatric Nurse Clinic within 1 year, cohort 88 children in June 2009- March 2010, Pre-clinic numbers of admission: 38, post-clinic numbers of admission: 4.

Conclusion: There was significant improvement in asthma self management among children and their family members who follow up at designated clinic. Asthma children and their family members also showed satisfaction with the service provided by the clinic. It also can provide intermittent monitoring to children's condition on top of follow up at Special Outpatient Clinic.

Taking Care of a Hospitalized Child with Acute Gastroenteritis: The Experience of Mothers

LW Chow

Department of Paediatrics and Adolescent Medicine, United Christian Hospital, Hong Kong

Background: Acute gastroenteritis (GE) was a worrying childhood disease and it was a major cause of paediatric hospitalization over the world and also in Hong Kong. Parents were eager to stay with their hospitalized child but it was a stressful and exhausted event. However, parent's experiences of looking after a hospitalized child with acute GE in Hong Kong were not known yet.

Aims and objectives: The aim of this study was to explore the experience of Hong Kong Chinese parents in taking care of their hospitalized child with acute GE. The objectives were: 1) to identify the reasons why parents choose to stay with their hospitalized child with acute GE, 2) to investigate the types of care parents give to their hospitalized child with acute GE, 3) to identify the difficulties that parents encounter in taking care of their

hospitalized child with acute GE, and 4) to identify the ways that healthcare professionals can support parents in caring their hospitalized child with acute GE.

Methods: The exploratory qualitative design was adopted in this study. It was conducted in a general paediatric ward in an acute hospital under the Hospital Authority in Hong Kong. Convenience sampling was used and twelve mothers were recruited. Data was collected by individual face-to-face semi-structured interview with an interview guide. All interviews were audio-taped for data analysis. Besides, some demographic data regarding the participated parents and their children was collected. Furthermore, field notes were used as supplement to record the non-verbal communication of participants in interviews.

Results: Content analysis was used and four major categories were identified from the interview data. The major categories were 1) Reasons for staying with child, 2) Carer stress, 3) Ambiguous feeling towards disease management, and 4) Expected support.

Conclusion: It was realized that all participants were keen on staying with their hospitalized child in order to provide physical and psychological care throughout the day. Also, parents could fulfill their own emotion needs if they could take care of their child by themselves. On the other hand, there was no doubt that staying with the hospitalized child with acute GE was really a stressful experience. Parents' personal physiological needs were put aside, and they were frightened to witness their child being suffered. Moreover, parents felt frustrated because there were no medications prescribed to relieve their child's discomfort and their child was under isolation care. Parents expected that the hospital could make some improvements on the hospital facilities and the healthcare providers could give reassurance so as to facilitate and support them in caring for their hospitalized child with acute GE.

Implications for practice: Healthcare providers should be more supportive and considerate of those parents who were staying with their child in the hospital throughout the day. Also, they should report the condition and progress of the hospitalized child to their parents in detail with simple words. On the other hand, hospital policy makers should employ more healthcare providers to care for the hospitalized children and their parents individually. Furthermore, the hospital could make some improvements on hospital facilities in order to facilitate the parents in caring for their hospitalized child with GE.

Implications for further research: It was suggested that interviews could be carried out to explore other carers' experiences. Besides, a larger sample size could be recruited

and hospitals from different geographic locations could be involved to gain richer and more holistic understanding of parents' experience in taking care of a hospitalized child with acute GE.

The Impact of Cancer and Its Treatment on Hong Kong Chinese Childhood Cancer Survivors' Psychosocial Well-being and Quality of Life

WHC LI,¹ SY CHUI,² KY HO¹

¹School of Nursing Studies, LKS Faculty of Medicine, The University of Hong Kong; ²Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Hong Kong

Background: Notwithstanding cancer survival rates are higher than before as a result of advances in cancer screening and breakthroughs in cancer treatment, childhood cancer survivors are at risk of adverse physical and psychosocial effects of the cancer treatment, severely affecting their quality of life. Whilst much of the attention has focused on the physiological care of childhood cancer survivors, the consequences of cancer and its treatments on psychosocial well-being remain relatively underexplored.

Aims: The aim of the study was to shed light on the impact of cancer and its treatments on the psychosocial well-being and quality of life of Hong Kong Chinese childhood cancer survivors.

Methods: A cross-sectional study was employed. A total of 137 childhood cancer survivors (9-16-year olds) who had their medical follow-up at the out-patient clinic were invited to participate in the study. Participants were asked to respond to the Chinese version of the Centre for Epidemiologic Studies Depression Scale for Children, short form of the State Anxiety Scale for Children, Rosenberg's Self-Esteem Scale, Coping Behaviour Checklist for Chinese Children and Paediatric Quality of Life Inventory.

Results: This study showed that a significant number of childhood cancer survivors in Hong Kong were potentially at risk of depression, or at least presented some depressive symptoms. The study also indicated that greater symptomatology of depression in childhood cancer survivors was associated with higher state anxiety, lower self-esteem, and poor quality of life.

Conclusion: This study has addressed a gap in the literature by examining the impact of cancer and its treatments on the psychosocial well-being of Hong Kong Chinese childhood cancer survivors. The study reveals that

cancer and its treatments have tremendous impact on the psychosocial well-being, in particular the quality of life of childhood cancer survivors. It is essential for health care professionals to develop and evaluate appropriate interventions with the aims at promoting psychosocial well-being and quality of life for childhood cancer.

Early Experience and Outcome on Home Parenteral Nutrition

AWS CHAN, DYK LEUNG, TSF LEUNG, RMS WONG, NS Tsoi

Department of Paediatrics & Adolescent Medicine, Queen Mary Hospital, Hong Kong

In 2009 the multidisciplinary parenteral nutrition support team was set up including doctors from paediatric & neonatal intensive care unit (PNICU), gastroenterology and paediatric surgery; nurses from PNICU, general paediatrics ward and paediatric surgical ward; allied health members including pharmacist and dietitian. Team members meet weekly to review and set up plans for patients receiving parenteral nutrition (PN) currently, patients with PN related cholestasis, patients with nutritional problems and home PN cases.

It is an unequivocal fact that home parenteral nutrition (HPN) is an expensive therapy. It is not only on the costs spending on pumps, equipment and consumables but also on the beyond measurable aspect of parents' commitment and energy towards taking care of their child at home.

We have 6 cases on HPN since 2003. The details of the cases are listed in the table below:

No	Maturity/ DOB	Birth weight	Diagnosis	HPN Period / outcome
1	32 18/1/01	1330 g	IUGR, Meconium ileus, Short gut syndrome	7/03-3/04 Weaned after 8m HPN
2	30 6/7 9/3/0	1310 g	Prematurity, NEC, 36 cm small bowel + colon	12/05-4/07 (~16m) Weaned after 16m HPN
3	27 3/7 3/2/04	1030 g	Prematurity, NEC, 20 cm small bowel + colon	2/06-8/07 (~16m) Weaned after 16m HPN
4	39 5/7 9/11/04	3240 g	NEC, 30 cm small bowel without colon	6/06-3/07 (~9m) Weaned after 9m HPN
5	32 6/7 17/4/07	3.15 kg	Mitochondrial disease, functional IO, Faconi's syndrome	4/09-12/10 Died
6	30 6/7 25/9/08	1.4 kg	Prematurity, NEC 9.5 cm small bowel + colon	8/10-till now

From the experiences that we learned through the 6 cases, parents' commitments contribute to the success of HPN

significantly. A well planned preparatory HPN program is essential to allay parents' fears of the complicated skills. Early parental participation in child care under nursing supervision aids to build up their confidence. All skills are written in checklist format to ensure parents comply to standard. Nurse taking the role of facilitator is important to identify parents' need physically, psychologically & financially. Multidisciplinary team involvement to ensure the whole family is being supported in the home care. Home PN is a joint decision from the medical team and the family working for the benefit of the child's development both physically and psychologically.

Primary Nurse Home Care Program – A Way to Enhance, Empower and Engage

LF CHEN, LS NG, YK HUI, SW CHENG, YM YEUNG,
GSK LUK

Department of Paediatrics, Queen Elizabeth Hospital, Hong Kong

Introduction: With advanced medicine and improved technology, more infants rely on medical equipment and survive with complex care procedures. As a result of these special care needs, the frequency of long-term hospitalization will increase. Paediatric home care has now become a significant and necessary component of the health care. A practice model of primary nursing program has been implemented in which a dedicated home care coordinator and primary nurse are responsible for coordinating, supporting and enabling parents of chronic cases to provide the complex care in a home setting to facilitate discharge in a timely and efficient manner. It is important to identify each family's coping mechanism, address their expectations, concerns and preferences, and alleviate parents' anxiety through education and communication.

Objectives: This paper aims to examine the impact of this model on the clinical practice. The hypothesis of this model would reduce the average length of hospitalization and increase the family satisfaction.

Methods: The clinical data of the length of hospitalization and the unscheduled readmission rate were reviewed and analyzed. A caregiver satisfactory survey was conducted from July 2010 to April 2011.

Results: The chronic case average mean length of hospitalization is reduced from 425 days (2008) to 262.4 days (2010). Besides, 75% (up to April 2011) chronic case length of hospitalization is less than 150 days. The unscheduled readmission rate is 26% (within 7 days) and

32% (within 14 days). From the caregiver survey, totally 21 chronically ill cases and their families are recruited. 95.2% caregivers agree and support the implementation of primary nurse home care program. 85.7% caregivers express satisfaction upon discharged. 95.2% caregivers express different degree of satisfaction towards the content of special care educated.

Conclusion: A significant reduction in the length of hospitalization is shown. Positive feedback from discharged parents is expressed. Home care program enhances better quality of life of these chronically ill children. It empowers the family to take care of their own child with medical needs at home. An organized, smooth and safe transition to home care is driven by the engagement of health care professionals. The Primary Nurse Home Care Program facilitates quality care for these children in their own homes.

Poster Presentation (Nurse's Session)

Atopic Eczema: Nurse-led Eczema Education Program Improved Patients' Quality of Life

YH CHAN, UI NG, WP MA, MW LEUNG, SH YEUNG,
MC LUK, ML LEE, YY TUNG, PK MA, KYC LAM,
HYT TAN, DCK LUK

Department of Paediatrics and Adolescent Medicine, United Christian Hospital, Hong Kong

Background: In Hong Kong, up to 20% of children have atopic eczema. Atopic eczema is a chronic debilitating skin disease which may have high impact on the quality of life to affected individuals and their families. Paediatric nurses play an important role in the care of children with atopic eczema. The effectiveness of the nursing care program could be evaluated using measurements.

Objectives: To evaluate the effectiveness of nursing care program for children with atopic eczema using the Children's Dermatology Life Quality Index (CDLQI) in Chinese version and the Family Dermatology Life Quality Index (FDLQI) in Chinese version.

Methods: Patients with atopic eczema following up in the Paediatric Skin Clinic were recruited. They received the nursing program for eczema care which included education on eczema skin care and tubular bandage wrapping. The improvement in the patients' quality of life was assessed by CDLQI and FDLQI.

Results: Twelve patients with atopic eczema were recruited from the Paediatric Dermatology Clinic of the

United Christian Hospital from August 2010 to March 2011. Their ages ranged from 6 to 18 years old and included X boys and X girls.

The impact on the quality of life was measured with CDLQI and FDLQI and the results were tabulated as follows:

	CDLQI	FDLQI
Before nursing program (m=)	16.3	15.4
After nursing program (m=)	5.7	9.4

Both the CDLQI and FDLQI were high before the nursing program which reflected a more significant impact on the quality of life by eczema. After the nursing program, lower scores were achieved.

Conclusion: Both Chinese versions were used to Children's Dermatology Life Quality Index and Family Dermatology Life Quality Index from Cardiff Dermatology centre to assess the impact of eczema on quality of life. Both the CDLQI and FDLQI presented after eczema nursing program the scores was decreased. These results suggested that nursing eczema program was beneficial in the management of atopic eczema and resulted in an improvement in the quality of life of patients and their families.

Paediatric Nurse Clinic in the Care of Asthma Children with Their Family

SP LEUNG, SY LAM, GPW LEE, KF LAU, YS LUI

Department of Paediatrics and Adolescent Medicine, Tuen Mun Hospital, Hong Kong, Hong Kong

Objectives: To evaluate the effectiveness of the Paediatric Nurse Clinic for the care of asthma children.

Methods: The Paediatric Nurse Clinic (Respiratory Care) commenced in June, 2009 to provide holistic care for asthma children and their family members. It is also to provide continuous assessment and progress supervision of children, so as to reduce re-admission rate of asthma attack and provide knowledge to them.

The services provide as followings:

1. Health assessment with diagnostic test
2. Health education of asthma
3. Nurse consultation/ counselling
4. Care planning and intervention
5. Phones follow up
6. Case follow up
7. Refer case to other health care professional if necessary

Results: There are 166 children recruited to follow up at Paediatric Nurse Clinic in June 2009 to March 2011. The outcome measures have been made in the followings:

1. Satisfaction survey by patients or family members (average score): 9.45 out of 10.
2. Assess asthma knowledge by questionnaires (average score): 11.3 out of 12.
3. Re-admission rate for asthma discharge patient (within 28 days): 0%.
4. Range of overall admission rate of asthma children every 3 month monitoring: 0-10%.
5. Compare the numbers of admission before and after attending the Paediatric Nurse Clinic within 1 year, cohort 88 children in June 2009-March 2010, Pre-clinic numbers of admission: 38, post-clinic numbers of admission: 4.

Conclusion: There was significant improvement in asthma self management among children and their family members who follow up at designated clinic. Asthma children and their family members also showed satisfaction with the service provided by the clinic. It also can provide intermittent monitoring to children's condition on top of follow up at Special Outpatient Clinic.

Effects of Music Therapy on Oxygen Saturation, Heart Rate and Mean Arterial Pressure of Premature Infants Receiving Endotracheal Suctioning: A Pilot Study

SM CHENG

Department of Paediatrics, Princess of Wales Hospital, Hong Kong

Background: During hospitalization, preterm infants experience many inevitable procedures such as endotracheal (ETT) suctioning that can cause them stress. Music therapy is one of the most practical and economical measures in relieving stress among preterm infants (Kemper & Danhauer, 2005). A pilot study was conducted to investigate the effects of music therapy on oxygen saturation, heart rate, and mean arterial blood pressure in premature infants receiving endotracheal suctioning.

Objectives: To examine the effects of intrauterine music "Transitions" on heart rate, mean arterial blood pressure and oxygen saturation after suctioning in premature infants in the neonatal intensive care unit.

Methods: The convenience sample included 24 infants born at 28 to 36 weeks' gestational age who were hospitalized in a teaching hospital in the Eastern part of New Territories.

A 2-group pretest-posttest experimental design was used whereby infants were randomly assigned to order of music versus no-music groups. Data were collected on 2 occasions (2 episode of suctioning) over a 4-hour period. Those physiological data for infants in both groups were collected at 2 minutes before suctioning and every minute for 15 minutes after suctioning. After collecting the baseline data, the infants were randomly assigned to either the control or the intervention (music) group. Mann-Whitney U-test will be used to test for any significant differences between the two groups before and after ETT suctioning. In addition, each of the physiological outcomes was also be analysed instead through a summary measure – the area under the curve (AUC) to compare each outcome variable between the two groups at baseline and post intervention.

Results: There was no significant difference on the summary measures, the area under the curve (AUC), of outcome measures between the two groups at the baseline. Although the change of the AUC of oxygen saturation (post-pre) between the two groups were also not statistical significant (all $p > 0.05$), it was of note that music group had a better improvement than the control group on oxygen saturation. Moreover, the results showed that the mean oxygen saturation after ETT suctioning of the control group was significantly lower than that of the music at 3rd minutes ($p = 0.004$), 4th minutes ($p = 0.011$). However, there were no significant difference of heart rate and mean arterial blood pressure among and between two groups.

Conclusion: Music therapy significantly improves the oxygen saturation during and after suctioning in this study which can reduce the side effects of suctioning. Based on results of this study, music therapy may serve as effective measures to offset the side effect of ETT suctioning.

Implications for practice and research: Music is a non-invasive and economical measure that can be implemented at the infant's bedside. Findings from this study demonstrated that preterm infants in a NICU did not have adverse reactions to a music intervention.

The Effect of Limb Massage on Infant's Venipuncture Pain

YM CHIK,¹ WY IP²

¹Department of Paediatrics and Adolescent Medicine, United Christian Hospital; ²The Nethersole School of Nursing, Faculty of Medicine, The Chinese University of Hong Kong, Hong Kong

Aims: The purpose of the study was to investigate the effect of upper limb massage on relieving pain among infants undergoing venipuncture in Hong Kong.

Methods: This study was a crossover, double-blind, randomized controlled trial. Sixty-five Chinese infants at the neonatal intensive care unit (NICU) in a regional hospital were randomly assigned to two groups in different order to receive interventions. Massage 1st group (N=32) received 2-minute massage performed by the researcher before venipuncture administration on the first occasion then received usual care (control) on the second occasion; and vice versa in Massage 2nd group (N=33). The infants' behaviour and physiological responses were recorded by a monitor and a video recorder with real time counter on two occasions, 1) right after the intervention, and 2) during the first 30 seconds of venipuncture procedure. The mean pain scores of infants were given by a trained nurse according to the Premature Infant Pain Profile (PIPP) in reviewing the record from the video recorder and the monitor. Ethical approval of this study was obtained from the study hospital.

Results: Mixed between-within subjects analysis of variance (ANOVA) indicated that the mean pain scores were significantly lower in infants who received massage [Massage 1st: 5.84 ($SD=3.51$); Massage 2nd: 7.30 ($SD=4.43$)] versus control [Massage 1st: 12.66 ($SD=3.10$); massage 2nd: 11.33 ($SD=4.37$)] [(Wilks' Lambda=0.39, $F(1,63)=98.32$, $p=0.00$, partial eta squared=0.61]. No significant differences in mean pain score between different orders in receiving massage.

Conclusion: Upper limb massage may be an effective technique for relieving infants' venipuncture pain.

Evaluation of Premie's Parents Support Meetings in Hospital

SY YEUNG

Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Hong Kong

Introduction: Given Birth to a preemie is a scary and overwhelming experience to parents. These parents would face the uncertainty that comes with having a baby in the hospital. Members of the Premie's Parents Support Group know that anxiety and fear as they are volunteers who have had babies in the Neonatal Intensive Care Unit (NICU). They aim to provide support and share experience with parents having preemies in the NICU.

Premie's parents support group was established as early as 1994 in our hospital. Meetings led by a nurse specialist were held to provide parent-to-parent support by enthusiastic volunteer parents from the group. Parents with NICU preemies born at or before 34 weeks were invited to join the sharing.

Evaluation: Evaluation on the preemie's parents support meetings was conducted between April 2010 and March 2011. 8 meetings were held with 87 numbers of invitation letter distributed. 43 families had attended the sharing involving 29 numbers of father and 39 numbers of mother. Meetings were held in Saturday afternoons lasting for 1½ hour.

68 evaluation questionnaires were distributed to both mom and dad. Thirty-six numbers of returned questionnaires were collected in the collection box with a return rate of 53%. Score 6 was regarded as mostly agreeable and score 1 was the least agreeable. 100% of the parents at the returned questionnaire agreed that the meeting met their needs and 81% marked a score of 5 or above. 100% of parents agreed that they were able to ventilate their feelings while 97% of them agreed that they were able to gain psychological support in the sharing at a score of 4 or above. Other comments they benefit from the sharing includes: sharing other preemies parents experience and learning that 'she is not the only one'; understanding more information on preemies and how to look after them; being able to talk and ventilate and feeling less stress; etc....

Conclusion: It is really difficult to understand the parents' feelings of having preemies in hospital when their friends or relatives may not have such experiences. This unique parent-to-parent support meeting surely has its value in NICU to offer supports to the desperate parents.

Quality Improvement Program: Paediatric Diabetic Mellitus Control

SL LAU,¹ M MOK,² KY LEUNG,³ E LAM,³ L HO,¹ KL NG,¹ P CHEUNG¹

¹Department of Paediatrics and Adolescent Medicine; ²Diabetes Nurse Specialist; ³Department of Dietetics, United Christian Hospital, Hong Kong

Introduction: The increasing incidence in childhood and adolescent diabetes mellitus is a world-wide phenomenon (Green & Patterson, 2001). Poor treatment adherence and irregular follow up attendance affect disease control. This is reflected by high HbA1c levels and follow-up non-attendance (DNA) rates. Poor glycaemic control leads to multi-organ complications. A paediatric diabetes mellitus service improvement program was introduced in 2010. It aimed at improving disease control, decreasing DNA rate, strengthening multidisciplinary collaboration and enhancing communication with patients, their families and schools.

Methodology: The program included building a paediatric diabetes mellitus management team, holding regular meetings and defining the roles and responsibilities of each discipline, developing "Gentle Reminders" for multidisciplinary follow-ups, capturing and monitoring attendance by "DMNS/Dietitian/Paediatrician Attendance Record" and "Monthly OPD slots", case discussion after clinics, telephone support, referring patients to support group, liaising with schools and social worker by paediatric and adolescent nurse, and providing health education to patients and their care givers.

Results: The annual evaluation included measuring the pre and post HbA1c levels, DNA rate, and manpower and the team cooperation. There were 30 patients with an average age of 14 years old. There was a significant improvement in HbA1c levels with pre and post intervention HbA1c values of 8.8 mmol/L and 7.8 mmol/L respectively ($p < 0.05$). 101 reminders were sent representing missed opportunities before intervention. The DNA rate was 10%. Patients were seen by paediatricians, DMNS and dietitian for 131, 105, 58 times in the 12 month intervention period. Six patients were successfully transitioned to adult care. There was an improvement by report of team cohesiveness. Early transition care for patients and their families is planned and will be the next target improvement. Important elements contributing to program success is the coordinating role by the paediatric and adolescent nurse, and the composite effect of small steps described in the methodology section.

Relationship Among Therapy-related Symptoms, Depressive Symptoms and Quality of Life in Hong Kong Chinese Children Hospitalized with Cancer

W H C L I,¹ S Y C H U I,² K Y H O¹

¹School of Nursing Studies, LKS Faculty of Medicine, The University of Hong Kong; ²Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Hong Kong

Background: With advances in cancer screening and medical treatment, the cancer survival rates are higher than ever before. Yet, the course of treatment for cancer, in particular the adverse effects of cancer treatment are distressing, which may severely affect the quality of life of children. Whilst much of the attention has focused on medical treatment, the consequences of therapy-related symptoms on the psychological well-being and quality of life in children hospitalized with cancer remain relatively underexplored.

Aims: This study explored the relationships among therapy-related symptoms, depressive symptoms and quality of life in Hong Kong Chinese children hospitalized with cancer.

Methods: A cross-sectional study was employed. A total of 135 Hong Kong Chinese children (8-18-year olds) admitted for cancer treatment in a paediatric oncology unit were invited to participate. Participants were asked to respond to the Therapy-Related Symptom Checklist for Children (TRSC-C), Centre for Epidemiologic Studies Depression Scale for Children and Pediatric Quality of Life Inventory (PedsQLTM 3.0 Cancer Module).

Results: The results of the study indicated that Hong Kong Chinese children hospitalized with cancer generally presented some depressive symptoms and reported poor health-related quality of life. Besides, there was a strong negative correlation between therapy-related symptoms and health-related quality of life. Conversely, there was a strong positive correlation between therapy-related symptoms and depressive symptoms. The findings indicate that children hospitalized with cancer reported greater symptom severity would report greater symptomatology of depression and experience poor quality of life.

Conclusion: The study has addressed a gap in the literature by exploring the relationships among therapy-related symptoms, depressive symptoms and quality of life in Hong Kong Chinese children hospitalized with cancer, an area of research that is under-represented in the existing literature. The findings reveal that treatment of cancer has tremendous impact on children's quality of life. There is an imperative need for nurses to evaluate appropriate nursing

interventions that can help children ease the burden of cancer treatment, enhance their quality of life and provide support for them to fight cancer at every step of their long and difficult journey.

Paediatric Rehabilitation Service: Transition from Acute Care to Home or Community Care

CH Lo

Department of Paediatrics and Adolescent Medicine, Caritas Medical Centre, Hong Kong

Introduction: Children after acute insult or illness affecting their neurological system may require a long period for rehabilitation before going back to the community. Patients and their families encounter lots of stress and problems in accepting and helping the patients to live with the disabilities. The Paediatric Rehabilitation Unit (PRU) of Caritas Medical Centre was established in October 2007 aimed to provide holistic step-down service to patients after major surgeries or required neurological rehabilitation.

During the transition period, we tailored rehabilitation program by our multidisciplinary team to maximize patient's functional level, provide holistic care to empower carers, assist the patients and their families to adapt living with the disabilities, arrange and co-ordinate appropriate resources in the community to facilitate care at home after discharge such as modification of the home environment according their needs.

Aims: To explore the case profile and services provided by the Paediatric Rehabilitation Unit (PRU).

Methods: A retrospective review of data since service commenced in October 2007 to February 2011 was carried out to review the patient profile and outcomes.

Results: During the review period from October 2007 to February 2011, there were 205 admissions received different services including neurological rehabilitation; intensive rehabilitation program after major orthopaedic or neurological surgeries; nutritional rehabilitation program and carer empowerment program. 168 patients admitted for shorter period for different investigations or procedures such as Phenol or Botox injection for spasticity management; oral gastro-endoscopy and 24 hours oesophageal pH monitoring; intravenous Pamidronate infusion therapy; step-down care after gastrostomy surgeries, etc. 199 ambulatory day admissions for multidisciplinary assessment, investigations or some

simple interventions. 1408 day admissions for regular physiotherapy and occupational therapy.

Conclusion: The Paediatric Rehabilitation Unit (PRU) provided holistic care bridging between acute service and community or home care. During admission, we trained up the kids in order to maximize their abilities; liaise with the external community resources; empower the parent/carer to facilitate home/community care; and relief the workload of the acute services. Majority of patients, including long-stay, short-stay and ambulatory cases, improved in their self-care abilities, muscles strength and range of movement, as well as their ambulatory state. We provide ongoing assessment by our multidisciplinary team after discharge for monitoring the progress.

Oral Feeding Performance in Infants

SK NG,¹ IFK LEE²

¹Department of Paediatrics and Adolescent Medicine, United Christian Hospital; ²The Nethersole School of Nursing, The Chinese University of Hong Kong, Hong Kong

Introduction: Oral feeding is challenging for the newborn infants and new mother. It is not only important for the growth and survives of infants but also affects mother-child interaction. Feeding problem frequently occurs in both of preterm and term infants during their first year of life. Healthy newborn infants may need to readmit or prolong stay in hospital by feeding-related problem. However, research on oral feeding in infants who are born at 34 gestational age (GA) or after are limited.

Aims: This study aimed to examine the oral feeding performance in infants (34-43 weeks GA) and the relationships among oral feeding performance, birth weight, gender, number of feeds, Apgar scores at 5 minutes and weight at observed feed.

Methods: This was a cross-sectional study. A convenience sample of 85 infants (41 female, 44 male) was recruited from United Christian Hospital during November, 2010. Oral feeding performance was assessed by three main indexes: proficiency, consumed and efficiency of the infants in the feeding process. Other variables in the study were collected from the infants' medical records.

Results: The participants (N=85) were predominantly infants with GA ranged from 34 to 43.1 weeks (*mean GA*=39.0±1.9 weeks) and mean birth weight=3.12±0.6 kg. Significant association was found between method of delivery and the consumed score. Those infants with

Spontaneous Vaginal Delivery (SVD) were associated with higher consumed score than using vacuum extraction during delivery (SVD-VE) ($p<0.001$). Also, near term infants were significantly associated with lower proficiency score than term infants ($p=0.003$). Bivariate correlation analysis revealed that post-gestational age, gestational age, birth weight, weight at observed feed, number of feeds and days after birth were significantly correlated with feeding performance (all p values <0.005).

Discussion and Conclusion: The infants' birth history and adaptation after birth were found associated with oral feeding performance. However, the feeding technique and the milk flow may also contribute to oral feeding performance in infants.

The Prevalence of Overweight and Obesity among Primary and Secondary School Students in Hong Kong

CM YU,¹ RLT LEE,² WT CHIEN,² L LOBO,² ALM WONG,² Q LAW,² J FUNG,² SKS LEE²

¹Private Practice, Hong Kong; ²School of Nursing, The Hong Kong Polytechnic University, Hong Kong

Background: The prevalence of obesity in children and adolescents is worldwide high with no exception in Hong Kong. Given the impact of obesity on health, this raised public health issues that need to be seriously addressed particularly those in lower social classes and also has created enormous concerns and interests for researchers to examine the obesity related diseases with their impacts on adolescent health and development.

Aims: This study was conducted to assess the prevalence of overweight and obesity among adolescents in low and middle income groups in a sub-urban area in Hong Kong.

Methods: A cross-sectional study and the adolescents were recruited from 15 primary and secondary schools in sub-urban area in Hong Kong. Standard anthropometric measurements including weight, height, body mass index (World Health Organization 2007 cut offs z-score was used), pulse, blood pressure, waist circumference, scapula and triceps skin fold thickness were collected from each adolescent. Data were computed with descriptive statistics and t-test.

Results: A total of 4,714 adolescents, aged 9-16 years ($M=12.11$, $SD=1.76$), with males ($n=2,422$, 51.1%) and females ($n=2,318$, 48.9%) were recruited in the study. Stature (cm) showed significant inter-group variation except

in the 10 year age group ($p<0.001$). BMI ranged from 11.13 to 42.00 kg/m with the mean ($M=19.42$) and the standard deviation ($SD=3.74$). Male adolescents, when compared with the female adolescents, obtained higher mean scores in 'Height' (153.28 vs. 150.27, $t=9.36$, $p<0.001$), 'Weight' (47.10 vs. 43.64, $t=9.60$, $p<0.001$), 'Body Mass Index' (19.73 vs. 19.09, $t=5.90$, $p<0.001$), 'Waist Circumference' (68.53 vs. 64.41, $t=15.17$, $p<0.001$) and 'Systolic Blood Pressure' (109.15 vs. 104.96, $t=12.07$, $p<0.001$). Female adolescents, when compared with the male adolescents, obtained higher mean scores in 'Triceps Skin Fold Thickness' (15.68 vs. 14.69, $t=-4.80$, $p<0.001$) and Scapula Skin Fold Thickness' (15.82 vs. 14.69, $t=-4.80$, $p<0.001$). The results also revealed that the incidence of overweight and obesity was higher in male adolescents as compared to the female adolescents, highlighting the possible role of change in the lifestyles in income levels.

Conclusion: The findings indicate that children and adolescents in Hong Kong have various health education needs related to lifestyle behaviours and their choices. School nurses are needed to promote healthy lifestyles, particularly boys, to reduce the prevalence of overweight and obesity and subsequent diseases. Current data and further prediction analysis will not only serve as a reference standard, but also be of high clinical importance in order to identify or categorize adolescent, particularly males and to take preventative measures to minimize health problems in the later life.

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