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ABSTRACTS

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MALAYSIAN PAEDIATRIC ASSOCIATION
Content

YOUNG INVESTIGATOR AWARD PAPERS

YIA 1
ASSESSMENT OF MORTALITY PREDICTION TOOLS IN PAEDIATRIC INTENSIVE CARE UNIT UMMC AND ESTABLISHING A NOVEL PAEDIATRIC INTENSIVE CARE REGISTRY (PEDICARE2008)
Anis Siham Zainal Abidin\(^1\), LCS Lum\(^2\), N Ismail\(^3\), M Sabri\(^4\)

YIA 2
PREVALENCE OF GLUCOSE-6-PHOSPHATE DEHYDROGENASE (G6PD) INTERMEDIATE IN THE NEWBORNS AND THE ACCURACY OF SEMI-QUANTITATIVE FLUORESCENT SPOT TEST (FST) AS SCREENING TOOL
Azizan Aziz\(^1\), Thong MK\(^2\), Lim CT\(^2\)

YIA 3
JUVENILE IDIOPATHIC ARTHRITIS IN MALAYSIA: THE SELAYANG EXPERIENCE
Cham Weng Tarng, Tang Swee Ping

YIA 4
HEALTH-RELATED QUALITY OF LIFE IN SABAH CHILDREN WITH TRANSFUSION DEPENDENT THALASSAEMIA
Choong Yee Wan, Lee Chee Chan

YIA 5
HYPOTHYROIDISM IN PRETERM INFANTS IN SPECIAL CARE NURSERY (SCN), UNIVERSITY MALAYA MEDICAL CENTRE (UMMC)
Hannah Wardiah Rosland, Chin-Theam Lim, Fatimah Harun

YIA 6
CORONARY ARTERY ABNORMALITY IN COMPLETE KAWASAKI DISEASE CHILDREN TREATED WITH IMMUNOGLOBULIN
Amelia Alias\(^1\), Chan TL\(^2\), Mohd Amin Itam\(^1\), Mat Bah MN\(^1\)

ORAL PRESENTATIONS

ORAL PRESENTATIONS 1

OP1.1
A DESCRIPTIVE STUDY OF APNOEA OF PREMATURITY IN INFANTS LESS THAN 34 WEEKS OF GESTATION AND A COMPARATIVE STUDY OF ORAL CAFFEINE VERSUS ORAL AMINOPHYLLINE IN THE TREATMENT OF APNOEA OF PREMATURITY
Lim Boon Soo, Lim Chin Theam

OP1.2
HUMAN RHINOVIRUSES AMONG CHILDREN HOSPITALISED WITH ACUTE LOWER RESPIRATORY TRACT INFECTIONS
Mohamad Reza Etemadi\(^7\), Farid Azizi Jalilian\(^1\), Norrashidah Abdul Wahab\(^3\), Zamberi Sekawi\(^7\), Putri Yubb\(^2\), Zurina Zainudin\(^7\), Shamarina Shohaimi\(^4\), Norliah Othman\(^2\)
**OP1.3**  
**INTRAHOSPITAL VARIATION OF THE BACTERIOLOGY OF NOSOCOMIAL INFECTIONS BETWEEN THE NICU AND OTHER ICUS**  
Noorzaitun Ariffin¹, Noraida Ramli¹, Choo Keng Ee¹, Fahisham Taib¹, Habsah Hassan², Nor Rosidah Ibrahim¹, Azriani Abd Rahman³, Van Rostenberghe Hans¹

**OP1.4**  
**PREVALENCE OF SLEEP DISORDERED BREATHING (SDB) AND RISK FACTORS IN MALAY, CHINESE AND INDIAN SCHOOL CHILDREN IN MALAYSIA**  
Norzila Mohd Zainudin¹, Jamalludin Ab Rahman², Norrashidah AW³, Asiah Kassim¹, Hasniah Abdul Latif⁴, Rus Anida Awang⁵, Ahmad Fadzil Abdullah⁶, Ramli Zainal⁷, Samsinah Hussain⁸

**OP1.5**  
**EVALUATION OF A WRITTEN ASTHMA ACTION PLAN IN THE MANAGEMENT OF CHILDREN WITH ASTHMA AT UNIVERSITY MALAYA MEDICAL CENTRE**  
Wong Su Sien, Anna M Nathan, Jessie A de Bruyne

**ORAL PRESENTATIONS 2**

**OP2.1**  
**MEDICAL PERSONNEL’S KNOWLEDGE ON PEAK EXPIRATORY FLOWMETRY IN CHILDREN**  
Azizah Musa, Asiah Kassim, Dayang Zuriani Sahadan, Norzila Mohamed Zainudin

**OP2.2**  
**SOCIAL DETERMINANTS (SDH) OF INFANT MORTALITY (IMR) IN MALAYSIA**  
Amaluddin Ahmad

**OP2.3**  
**NUTRITIONAL STATUS OF SCHOOL CHILDREN AGED 6-10 YEARS OLD IN SELANGOR**  
Asiah Kassim¹, Norzila Mohamed Zainuddin¹, Jamalludin Abdul Rahman², Ramli Zainal³, Ahmad Fadhlil Abdullah⁴, Norrashidah Abdul Wahab⁵, Rusanida Awang⁶, Hasniah Abdul Latif⁷, Shamsinah Hussain⁸

**OP2.4**  
**NON-ATTENDANCE TO THE PAEDIATRIC CLINICS IN A MALAYSIAN TERTIARY HOSPITAL: A SIZEABLE PROBLEM AND IDENTIFICATION OF AN EFFICACIOUS INTERVENTION**  
Mohammad Tamim Jamil, Nik Zainal Abidin Nik Ismail, Azman Bin Zulkifli, Noorizan Abdul Majid, Hans Van Rostenberghe

**OP2.5**  
**WEANING PRACTICE AMONG MOTHERS IN HUSM**  
Nor Rosidah Ibrahim, Chiew Yee Soon, Tay Mei Ee, Tay See Hoon, Wee Shi Ning, Noraida R, Noorizan A M, Norsarwany M

**ORAL PRESENTATIONS 3**

**OP3.1**  
**SPECTRUM OF CONGENITAL HEART DISEASE IN HOSPITAL SULTANAH AMINAH, JOHOR BAHRU**  
Amelia Alias¹, Chan TL², Mohd Amin Itam¹, Mat Bah MN¹
**OP3.2**

**IMMUNOGENICITY OF A 3-DOSE PRIMARY SERIES OF THE PNEUMOCOCCAL NON-TYPEABLE HAEMOPHILUS INFELUENZAE PROTEIN-D CONJUGATE VACCINE (PHiD-CV) IN HEALTHY MALAYSIAN AND SINGAPOREAN INFANTS**  
Fong Seng Lim¹, Kah Kee Tan², Poh Chong Chan⁴, Chia Yin Chong⁴, Mia Tuang Koh⁵, Yee Leong Teoh⁶, Aurélie Fanic⁷, Ilse Dieussaert⁷, Kristien Swinnen⁷, Dorota Borys⁷

**OP3.3**

**THE ROLE OF ROUTINE RSV SCREENING IN CHILDREN PRIOR TO SURGERY AND CARDIAC CATHETER PROCEDURE IN INSTITUT JANTUNG NEGARA**  
Leong Ming Chern, Khairul Faizah, Adura Abdul Rauf, Hasri Samion, Mazeni Alwi

**OP3.4**

**SAFETY OF MMR VACCINATION. IS ADMISSION NECESSARY?**  
Muzhirah Aisha Haniffa

**POSTER PRESENTATIONS**

**P1**

**REAL-TIME QUANTITATIVE PCR CHIMERISM ASSAY AFTER BONE MARROW TRANSPLANTATION (BMT) IN SEX MATCHED SIBLING**  
Aliza Mohd Yacob¹, Ida Shahnaz Othman², Hishamshah Ibrahim², Raba’atun Sham¹, Fateha Misnar¹, Erniza Assari¹, Maznah Abu Bakar¹, Ezalia Esa¹, Zubaidah Zakaria¹

**P2**

**PREVALENCE OF LATE DIAGNOSIS OF DUCT DEPENDENT AND CYANOTIC CONGENITAL HEART DISEASE IN HOSPITAL SULTANAH AMINAH JOHOR BAHRU**  
Amelia Alias¹, Chan TL², Mohd Amin Itam¹, Mat Bah MN¹

**P3**

**LIPAEMIC BLOOD: A CASE REPORT**  
Ashikin MN, Teoh SL, Loo HM, Kuan GL

**P4**

**DEMOGRAPHIC PROFILE AND PHYSICAL CHARACTERISTICS OF CHILDHOOD OBESITY IN UNIVERSITY MALAYA MEDICAL CENTER**  
Azriyanti AZ, Rosmini T, Yazid J, Fatimah H

**P5**

**NUTRIENT DRINKS’ AND HEPATIC IMPAIRMENT IN CHILDREN: A CASE REPORT**  
Cham Weng Tamg, Tang Swee Ping, Lim Sern Chin

**P6**

**PAEDIATRIC TRAUMA: PATTERNS OF INJURY PRESENTING TO A PAEDIATRIC TRAUMA AND EMERGENCY UNIT IN A TERTIARY HOSPITAL IN MALAYSIA**  
Cheah Peng Loon, Jessie de Bruyne, Priya Sathish Chandran

**P7**

**A CASE REPORT: HYPERCALCIURIC BARTTER’S SYNDROME WITH NEPHROCALCINOSIS**  
BH Ching¹, PY Tam¹, Janet YH Hong²
<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
<th>Authors</th>
</tr>
</thead>
<tbody>
<tr>
<td>P8</td>
<td>TREATMENT OF SUBCUTANEOUS PANNICULITIS-LIKE T-CELL LYMPHOMA WITH</td>
<td>Chong Lee Ai, Manimalar Naicker, Lalitha Pillay, Hany Ariffin</td>
</tr>
<tr>
<td></td>
<td>CORTICOSTEROID MONOTHERAPY: A CASE REPORT</td>
<td></td>
</tr>
<tr>
<td>P9</td>
<td>PENILE TUMOUR IN A 2 YEAR OLD CHILD AT HUSM: A CASE REPORT</td>
<td>Fahisham T, Norsarwany M, Mohd Ashraf MD, Ariffin N</td>
</tr>
<tr>
<td>P10</td>
<td>ATYPICAL PRESENTATION OF CHILDHOOD LEUKAEMIA</td>
<td>Florence Wong, Grace Chow Kam Yoke, Tang Swee Ping, Cham Weng Tarng</td>
</tr>
<tr>
<td>P11</td>
<td>LANGERHANS CELL HISTIOCYTOSIS – A RARE CASE OF SEVERE PULMONARY</td>
<td>Haiza Hani Hamidon, C-Khai Loh, Hamidah Alias</td>
</tr>
<tr>
<td></td>
<td>INVOLVEMENT</td>
<td></td>
</tr>
<tr>
<td>P12</td>
<td>LI-FRAUMENI SYNDROME IN MALAYSIAN FAMILIES</td>
<td>Hany Ariffin, Chong Lee Ai, Zarina A Latiff, Mahfuzah Mohamed, Eni Juraida</td>
</tr>
<tr>
<td>P13</td>
<td>CASE REPORT: AGROBACTERIUM RADIOBACTER INFECTION IN PAEDIATRIC</td>
<td>Hayati J¹, Norsarwany M¹, Siti Asma’ H², Fahisham T¹, Ariffin N¹</td>
</tr>
<tr>
<td></td>
<td>ONCOLOGY PATIENT, HUSM EXPERIENCE</td>
<td></td>
</tr>
<tr>
<td>P14</td>
<td>MOWAT WILSON SYNDROME CAUSED BY DE NOVO CHROMOSOME 2</td>
<td>YL Heng, WT Keng, Roziana Ariffin, Farah Amalina, Aminah Mekesat</td>
</tr>
<tr>
<td></td>
<td>PERICENTRIC INVERSION</td>
<td></td>
</tr>
<tr>
<td>P15</td>
<td>OCHOA SYNDROME: SMILE TO DIAGNOSE A NEUROGENIC BLADDER</td>
<td>Indra Ganesan</td>
</tr>
<tr>
<td>P16</td>
<td>SHORT STUDY OF ACUTE GASTROENTERITIS IN PUTRAJAYA HOSPITAL,</td>
<td>Janet YH Hong¹, Sofia ANM², Fuziah MZ¹</td>
</tr>
<tr>
<td></td>
<td>FROM APRIL TO JUNE 2010</td>
<td></td>
</tr>
<tr>
<td>P17</td>
<td>TRENDING OF CHILD ABUSE CASES PRESENTED TO PUTRAJAYA HOSPITAL</td>
<td>Janet YH Hong¹, AB Ismail³, Jabbar Al³, JSL Wong¹, Ashwin Kaur JS¹, Fuziah MZ¹, Nora³ MS²</td>
</tr>
<tr>
<td>P18</td>
<td>PARENTS’ PERCEPTION AND SATISFACTION OF QUALITY FAMILY-CENTERED</td>
<td>Khairun Nisa Mohammad, Jayah K Pubalan</td>
</tr>
<tr>
<td></td>
<td>CARE IN PEDIATRIC WARDS AT HOSPITAL UNIVERSITI SAINS MALAYSIA</td>
<td></td>
</tr>
<tr>
<td>P19</td>
<td>PARENTS VIEWS OF LUMBAR PUNCTURE IN CHILDREN</td>
<td>Wong SLJ, Yeoh AAC, Ooi TC, Lye CS</td>
</tr>
</tbody>
</table>
P20
MITOTANE RELATED VAGINAL BLEEDING IN A GIRL WITH RECURRENT ADRENOCORTICAL CARCINOMA
Karmila A, Nurshadia S, Jalaludin MY, Harun F

P21
SEASONAL PATTERN AND CLINICAL FEATURES OF RESPIRATORY SYNCYTIAL VIRUS (RSV) INFECTION IN SABAH, MALAYSIA
Kelvin Chia, EC Pang, CW Koh, SM Fong, TL Soo

P22
INTRAVENOUS IMMUNOGLOBULIN IN INFLUENZA A H1N1-ASSOCIATED ACUTE NECROTIZING ENCEPHALOPATHY: A CASE REPORT
Koh Lee Min, Tang Swee Ping, Lim Sern Chin, Cham Weng Tarng

P23
NON-ACCIDENTAL OR ACCIDENTAL HEAD INJURIES IN MALAYSIAN INFANTS: THE CLINICAL CLUES
Meera Thayalasingam1, Irene Cheah Guat Sim2, Abhi Veerakumarasivam1

P24
MANAGING CEREBRAL PALSY PATIENTS: ARE WE DOING ENOUGH?
Mastura Ibrahim, Aina Mariana A Manaf, SL Wong

P25
IMMATURE BRAIN TERATOMA IN AN INFANT IN HOSPITAL UNIVERSITI SAINS MALAYSIA: REPORT OF A CASE
Norsarwany M1, Ariffin N1, Mohd Iqbal I1, Hasnan J2, Rohaizan Y3, Abdul Rahman G4, Stewart K5

P26
NETHERTON SYNDROME: A CASE REPORT
Suhaila Omar, Muzhirah Aisha Md Haniffa

P27
CHRONIC IMMUNE THROMBOCYTOPENIC PURPURA IN CHILDHOOD: A CLINICAL PROFILE
Abdul-Wahab1, Nargis M1, Aye Aye1, Haymond P2, Naznin M3

P28
VEIN OF GALEN MALFORMATION IN NEWBORN WITH HEART FAILURE
Norazah Zahari, Lim Chin Theam

P29
RIGHT ADRENAL PHAEOCHROMOCYTOMA IN A BOY WITH HYPERTENSIVE CRISIS INITIALLY SUSPECTED TO BE DUE TO AN INTRACRANIAL LESION
Nurshadia S1, Karmila A1, Lucy C2, Ramanujam T3, Lucy LCS1, Fatimah H1, Jalaludin MY1

P30
INFLUENZA TYPE B VIRUS INFECTION ASSOCIATED ACUTE NECROTIZING ENCEPHALOPATHY OF CHILDHOOD
Putri Y1, Nur Hafiza N2, Norashidah AW3, Norlijah O3, Hamid MZA1
<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
<th>Authors</th>
</tr>
</thead>
<tbody>
<tr>
<td>P31</td>
<td>TWO PREGNANCIES RESULTING IN 2 DAUGHTERS WITH ABERRANT CHROMOSOME X IN A MOTHER WITH VARIANT TURNER SYNDROME</td>
<td>Sathyabama R¹, Keng WT¹, Vigneswari G², Roziana A²</td>
</tr>
<tr>
<td>P32</td>
<td>EPIDEMIOLOGIC FEATURES OF CHILD ABUSE IN SABAH WOMEN AND CHILDREN'S HOSPITAL FROM JANUARY 2010 TO JUNE 2010</td>
<td>Suhaila I, Fauziah MZ, TL Soo</td>
</tr>
<tr>
<td>P33</td>
<td>A REPORT OF TWO LIFE-BORN INFANTS, HOMOZYGOUS FOR SOUTHEAST ASIAN OVALOCYTOSIS</td>
<td>Surini Yusoff¹, Narazah Mohd Yusoff², Keng Wee Teik³, Chng Gaik Siew³, Winnie Ong Pei Tee¹, Yeoh Seoh Leng⁴, Nur Hasnah Ma'amor⁵, Masafumi Matsuo⁶, Hans Van Rostenberghe⁷</td>
</tr>
<tr>
<td>P34</td>
<td>CHRONIC PAIN SYNDROMES: A MISSED OR MISDIAGNOSIS?</td>
<td>Tang Swee Ping</td>
</tr>
<tr>
<td>P35</td>
<td>INFLIXIMAB THERAPY IN SEVERE REFRACTORY KAWASAKI DISEASE</td>
<td>Tang Swee Ping¹, Hung Liang Choo²</td>
</tr>
<tr>
<td>P36</td>
<td>CLINICAL FEATURES AND PREDICTORS OF RENAL DISEASE IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS</td>
<td>Lim Sern Chin, Tang Swee Ping, Wan Jazilah Wan Ismail</td>
</tr>
<tr>
<td>P37</td>
<td>GAUCHER'S DISEASE TYPE 2: A CASE REPORT</td>
<td>Tengku Putri Zainab, Soo Min Hong, Ngu Lock Hock</td>
</tr>
<tr>
<td>P38</td>
<td>L-ASPARAGINASE INDUCED SAGITTAL SINUS THROMBOSIS IN CHILDHOOD ACUTE LYMPHOBLASTIC LEUKAEMIA: A CASE REPORT</td>
<td>Teoh SL, Kuan GL</td>
</tr>
<tr>
<td>P39</td>
<td>POLYARTERITIS NODOSA – A RARE PRESENTATION</td>
<td>Tina Sivalal¹, Tang Swee Ping², Cham Weng Tarng², Soo Min Hong¹</td>
</tr>
<tr>
<td>P40</td>
<td>A CASE REPORT OF PENTALOGY OF CANTRELL</td>
<td>Yeoh Chiu Yen, Sunita, Yong Lai Peng, Nachal RMN Nachiappan, Yogeswari Sithamparanathan</td>
</tr>
<tr>
<td>P41</td>
<td>DAPSONE SYNDROME – FIRST PAEDIATRIC CASE REPORT IN MALAYSIA</td>
<td>Z Zurina¹, O Elizawaty², S Thevarajah³, O Norlijah¹</td>
</tr>
<tr>
<td>P42</td>
<td>BRONCHIAL ARTERY EMBOLISATION IN BRONCHIECTASIS WITH NEOVASCULARISATION AND HEMOPTYSIS</td>
<td>Teh Keng Hwang, Choong Pheik Sian</td>
</tr>
</tbody>
</table>
P43
CHILDHOOD OBESITY – PREVALENCE AMONG 7 AND 8 YEAR OLD PRIMARY SCHOOL STUDENTS IN KOTA KINABALU
Chong Hon Loon
ASSESSMENT OF MORTALITY PREDICTION TOOLS IN PAEDIATRIC INTENSIVE CARE UNIT UMMC AND ESTABLISHING A NOVEL PAEDIATRIC INTENSIVE CARE REGISTRY (PEDICARE2008)

Anis Siham Zainal Abidin, LCS Lum, N Ismail, M Sabri

1. University of Technology Mara, Shah Alam, Malaysia
2. University of Malaya, Kuala Lumpur, Malaysia

Introduction: Two generic paediatric mortality scoring systems have been validated in the paediatric intensive care unit (PICU) worldwide. Paediatric Risk of Mortality (PRISM III) requires an observation period of 24 hours, and measures severity at 24 hours after admission, which represents a limitation for clinical trials that require earlier inclusion. The Paediatric Index of Mortality (PIM2) is calculated 1 hour after admission but does not take into account the stabilization period following admission. To avoid these limitations, we chose to conduct assessments at 1-hour, 12-hours and 24-hours after PICU admission. The aim of our study was to validate PRISM III and PIM2 at the time points for which they were developed, and to compare their accuracy in predicting mortality at those times with their accuracy at 12 hours. A novel computerized paediatric registry was developed for audit purposes and further PICU research.

Methods: All children admitted from August 2008 to December 2009 in one tertiary PICU (UMMC) were prospectively included. Data collected generates scores and predictions using PRISM III and PIM2.

Results: There were 563 consecutive admissions with 47 deaths. All four models exhibited good discrimination (area under the receiver operating characteristic curve ≥0.7) but only PRISM III -12hours showed good calibration (p>0.05). The Standardized Mortality Ratio was 1.05 (p<0.001).

Conclusions: Our study showed PRISM III-12 hours model offered the best risk adjusted mortality prediction tool for our PICU in UMMC.
PREVALENCE OF GLUCOSE-6-PHOSPHATE DEHYDROGENASE (G6PD) INTERMEDIATE IN THE NEWBORNS AND THE ACCURACY OF SEMIQUANTITATIVE FLUORESCENT SPOT TEST (FST) AS SCREENING TOOL

Azizan Aziz¹, Thong MK², Lim CT²

1. Hospital Batu Pahat, Johor
2. Department of Paediatrics, Faculty of Medicine, University Malaya Medical Centre, Kuala Lumpur

Aim: To determine the correlation between G-6-P-D Intermediate status and the actual G-6-P-D enzyme levels in infants during neonatal period at UMMC.

Methods: This study was a hospital-based observational cohort study on newborn delivered, admitted, and subsequently has assessment of jaundice at University Malaya Medical Centre (UMMC) Kuala Lumpur from 1st January 2008 to 30th June 2009. The detection of G-6-P-D Intermediate status was performed on cord blood sample with Semiquantitative Fluorescent Spot Test. A subgroup analysis was carried out from April 2008 to September 2009 to study the association between the actual G-6-P-D enzyme levels and G-6-P-D Intermediate status.

Results: The prevalence of G-6-P-D Intermediate was 1.51% with male to female ratio was 1:35. The prevalence was highest among Chinese (2.12%), followed by Malay (1.7%) and Indian (0.58%). From the subgroup analysis the prevalence of enzyme deficient infants was 7.5%. The enzyme analysis in G-6-P-D intermediate infant only 46% were actually enzyme deficient with the FST having 32% sensitivity and 98% specificity and the worrying facts that is FST misses 2.8% of G-6-P-D normal infants which actually enzyme deficient.

Conclusions: G-6-P-D intermediate infants diagnosed by FST need actual confirmatory G-6-P-D enzyme levels.
YOUNG INVESTIGATOR AWARD PAPERS

YIA3

JUVENILE IDIOPATHIC ARTHRITIS IN MALAYSIA:
THE SELAYANG EXPERIENCE

Cham Weng Tarng, Tang Swee Ping

Department of Paediatrics, Hospital Selayang, Selangor

Introduction: Juvenile idiopathic arthritis (JIA) is one of the commonest childhood rheumatological disorders worldwide with varying incidence and prevalence depending on geographical area and ethnicity. Such epidemiological data on JIA is still lacking in Malaysia.

Objective: To determine the demographic characteristics and JIA subtypes of all JIA patients seen at a tertiary referral center.

Methods: A retrospective review was done using the electronic medical records of all patients diagnosed with JIA seen at the Paediatric Rheumatology Unit, Hospital Selayang from October 2003 till August 2010. Data collected included demographic data, age at diagnosis, JIA subtype and original referring hospital.

Results: There were a total of 160 patients with JIA (156 Malaysians, 4 foreigners) with male:female ratio of 1:1.8. Majority were Malays (52.5%) followed by Chinese (28.1%), Indians (16.2%) and other races (5.3%). The frequency of JIA subtypes seen were polyarthritis (total 41.9%, Rheumatoid factor (RF) positive: 8.1%, RF negative: 33.8%), systemic (38.8%), oligoarthritis (13.8%), enthesitis-related arthritis (7.5%) and psoriatic arthritis (1%). Most (94.3%) were external referrals, predominantly from around Klang Valley (42.5%), followed by Penang (9%), Negeri Sembilan (8%), Perak and Johor (5.6% each), Pahang, Kedah and Kelantan (5% each), Malacca (3.1%), Sabah (2%), Sarawak (1%), Terengganu (0.6%) and Singapore (0.6%).

Conclusion: The predominant subtypes of JIA seen were polyarthritis and systemic onset and this is contrary to western literature whereby oligoarthritis is the commonest subtype. This may be due to a referral bias as often the more severe subtypes are referred to Hospital Selayang, being a tertiary referral centre.
HEALTH-RELATED QUALITY OF LIFE IN SABAH CHILDREN WITH TRANSFUSION DEPENDENT THALASSAEMIA

Choong Yee Wan, Lee Chee Chan
Sabah Women And Children Hospital, Sabah

Background: Sabah is the state with the most transfusion dependent Thalassaemia patients. Studies upon the quality of life for children with chronic illness such as Thalassaemia are limited. With the improved healthcare services, we conducted a study to investigate the quality of life for our Thalassaemia patients using PedsQL 4.0 generic Scale to score on physical, emotional, social, and role (school) functioning comparing to the healthy controls.

Methods: Children with Thalassaemia receiving blood transfusions and treatments at Thalassaemia Treatment Centre, Sabah Women and Children Hospital, Malaysia were included in the study. The children quality of life is scored using PedsQL 4.0 generic Scale Score. Accordingly, the questionnaire was also administered to a control group of healthy school going children. Sociodemographic data were also collected from patients and controls using an interview schedule designed for the study.

Results: 180 thalassaemia patients were approached, all given the consent to be interviewed. 200 healthy controls were approached, and all agreed to participate. The age range for the patients and the schoolchildren is between 5 to 16 years and 6 to 16 years respectively. Both study and control groups are comparable in term of demographic background. However, this study reviewed that thalassaemia patients had significant lower quality of life compared to the controlled group.

Conclusion: Quality of life for Thalasseamia patients in terms of physical, emotional, social, and school functioning is not comparable with the healthy counterparts. Medications with less pain can be considered as an option for the thalassaemia patients. Beside medical treatments, medical professionals and society should give more attention to improve other aspects in life for the thalassaemia patients, hence improve their quality of life.
Objective: To determine the incidence of transient hypothyroxinaemia and persistent hypothyroxinaemia (true congenital hypothyroidism) and factors affecting transient hypothyroxinaemia in preterm infants up to 28 days of life.

Methods: This was a prospective longitudinal study from 1st January 2010 to 30th June 2010 of preterm infants born at 25 to 36 weeks of gestation. Blood samples for FT4 and TSH were collected and sent at birth, days 7, 14 and 28 of life. Data were collected using a standard data collection form. SPSS programme was used to analyse the data.

Results: The incidence of transient hypothyroxinaemia was 59.3% and the persistent hypothyroxinaemia was 2.4%. There was one patient with persistent hypothyroxinaemia with high TSH with borderline normal FT4 beyond 28 days of life. This might indicate true congenital hypothyroxinaemia and L-thyroxine treatment was commenced. The incidence of transient hypothyroxinaemia was significantly affected by the presence of patent ductus arteriosus (PDA) (p<0.01), oxygen dependency up to 28 days (p<0.02), clinical sepsis (p<0.02) and those who were exposed to at least 2 occasions of povidone-iodine compound (p<0.001).

Conclusion: The incidence of transient hypothyroxinaemia was high (59.3%) and the persistent hypothyroxinaemia was 2.4%. Transient hypothyroxinaemia was significantly associated with PDA, clinical sepsis, oxygen dependency up to 28 days of life and those who were exposed to more than 2 occasions of povidone-iodine compound. It is recommended to monitor thyroid functions in preterm infants < 36 weeks of gestation receiving intensive care up 4 to 8 weeks of age to avoid missing cases of true congenital hypothyroidism.
CORONARY ARTERY ABNORMALITY IN COMPLETE KAWASAKI DISEASE CHILDREN TREATED WITH IMMUNOGLOBULIN

Amelia Alias¹, Chan TL², Mohd Amin Itam¹, Mat Bah MN¹

1. Paediatric Cardiology Unit, Department of Paediatrics, Hospital Sultanah Aminah, Johor
2. Gleneagles Hospital, Kuala Lumpur

Objective: Intravenous immunoglobulin (IVIG) therapy in Kawasaki disease (KD) has been shown to reduced coronary artery aneurysm to 1-2%. However, we still observed significant number of coronary aneurysm post IVIG in our centre. The objectives are to study the prevalence of coronary artery abnormality (CAA) and associated risk factors.

Methods: A retrospective study performed on all children with KD from 1st January 2005 to 30th July 2010. Japanese Ministry of Health criteria were used to classify coronary arteries abnormality. Children with incomplete or atypical KD were excluded. Data were extracted from Pediatric Cardiology Clinical Information System

Results: A total of 126 KD were diagnosed during the study period with 69% were male and 52.4% were Chinese. The median age of diagnosis was 1.4yr [Q1, 0.6yr Q3, 2.3yr]. Of these 126, 118 (93.7%) received IVIG within 10 days of illness. Ten patients (7.9%) required more than one dose of IVIG. CAA were noted in 28 (22.2%) of patients with 21 ectasia, 4 small fusiform, one small saccular and 2 medium fusiform aneurysm. Of these 28, 22 who had IVIG within 10 days of illness (18 ectasia, 3 small and one medium coronary aneurysm). Significant risk factors for CAA were older children (2.3 v s 1.7yr, p=0.03), presentation after 10days of illness (p=0.066) and required more than 2gm/kg of IVIG (p=0.04).

Conclusions: The prevalence of CAA in complete KD treated with IVIG was 22.2% with 5.5% significant aneurysm. Risk factors for coronary abnormality were older children, late presentation and require more than 2gm/kg of IVIG.
A DESCRIPTIVE STUDY OF APNOEA OF PREMATURITY IN INFANTS LESS THAN 34 WEEKS OF GESTATION AND A COMPARATIVE STUDY OF ORAL CAFFEINE VERSUS ORAL AMINOPHYLLINE IN THE TREATMENT OF APNOEA OF PREMATURITY

Lim Boon Soo, Lim Chin Theam

Department of Paediatrics, Faculty of Medicine, University Malaya Medical Centre, Kuala Lumpur

Objectives: To study the incidence of apnoea of prematurity (AOP) and to compare the efficacy and safety of oral aminophylline with oral caffeine in the treatment of AOP.

Method: This was a prospective study. All preterm infants of Malaysian origin less than 34 weeks of gestation, admitted to the Special Care Nursery (SCN), University of Malaya Medical Centre (UMMC) from 1st May 2008 to 28th February 2010, were included in the descriptive study. Those on intravenous aminophylline, when weaned from ventilator support were allocated to either oral aminophylline (control group) or oral caffeine (study group) to compare the efficacy and safety of these two drugs.

Results: The overall incidence of AOP was 49.0%. Almost all infants ≤ 29 weeks of gestation or birth weight ≤ 1,000 grams had AOP. The incidence of AOP decreased with increasing gestational age and larger birth weight (p<0.05). AOP was significantly associated with pregnancy-induced hypertension, chronic lung disease (CLD) and retinopathy of prematurity (ROP) (p<0.05).

Oral caffeine treatment had decreased the frequency of AOP and the duration of oxygen usage (p<0.05). It was comparable with oral aminophylline in terms of needs for bag-mask ventilation and re-intubation, mean weight gain and total duration of hospitalization. Both oral aminophylline and oral caffeine had minimal side effects and were not associated with CLD, ROP and intaventricular haemorrhage (IVH).

Conclusion: The overall incidence of AOP was high among infants ≤ 29 weeks of gestation or birth weight ≤ 1,000 grams. Oral caffeine is superior to oral aminophylline for treatment of AOP.
HUMAN RHINOVIRUSES AMONG CHILDREN HOSPITALISED WITH ACUTE LOWER RESPIRATORY TRACT INFECTIONS

Mohamad Reza Etemadi¹, Farid Azizi Jalilian¹, Norrashidah Abdul Wahab³, Zamberi Sekawi¹, Putri Yubbu², Zurina Zainudin², Shamarina Shohaimi⁴, Norlijah Othman²

1. Department of Medical Microbiology and Parasitology, Faculty of Medicine and Health Sciences, University Putra Malaysia
2. Department of Paediatric, Faculty of Medicine and Health Sciences, University Putra Malaysia
3. Department of Paediatric, Hospital Serdang
4. Faculty of Science, University Putra Malaysia

Human rhinovirus (HRV), was previously associated with upper respiratory infections is now increasingly recognised as a cause of acute lower respiratory tract infections (ALRTI) in hospitalised children. Newly discovered HRV-C has been detected especially in patients with acute wheezing illnesses. The aim of the study was to assess the incidence of HRV infections and to compare the clinical features of HRV-A and C among children less than five years hospitalised with ALRTIs.

Methodology: Nasopharyngeal aspirates collected from 165 patients, were screened for HRV using RT-PCR of the VP4/2 gene and further sequencing of the amplicons was performed.

Results: Of the 165 enrolled children 54 (33%) were positive for HRVs, of which 20 (37%) were co-infected with other viruses. The median age of children with total HRV infection was 8.7 ranging from 1.1 to 45.1 months. HRV-A and HRV-C were detected respectively in 14 (39%) and 22(61%) of a total of 36 sequenced samples analysed. Children infected with HRV-C were found to be of older age and were more likely to be associated with vomiting and rhonchi as compared with HRV-A. On the other hand, children with HRV-A were more susceptible to a family history of atopy as compared with HRV-C. The study also revealed that HRV-C infected children tend to have a higher neutrophil count while lymphocytosis was associated with HRV-A.

Conclusion: The prominent etiologic role of HRVs suggests that laboratory diagnosis of HRV should be routinely included in the hospitalised children. This is the first report of HRV-C infection in Malaysia.
INTRAHOSPITAL VARIATION OF THE BACTERIOLOGY OF NOSOCOMIAL INFECTIONS BETWEEN THE NICU AND OTHER ICUS

Noorzaitun Ariffin¹, Noraida Ramli¹, Choo Keng Ee¹, Fahisham Taib¹, Habsah Hassan², Nor Rosidah Ibrahim¹, Azriani Abd Rahman³, Van Rostenberghe Hans¹

1. Departments of Paediatric, School of Medical Sciences, Universiti Sains Malaysia, Kelantan
2. Department of Microbiology, School of Medical Sciences, Universiti Sains Malaysia, Kelantan
3. Department of Community Medicine, School of Medical Sciences, Universiti Sains Malaysia, Kelantan

Objectives: To determine the bacteriology of nosocomial infection within the neonatal intensive care unit (NICU) of Hospital Universiti Sains Malaysia (HUSM) and compare it to other intensive care and high dependency units (ICU/HD) in HUSM.

Methodology: All records of cultured organisms from the NICU and ICU/HD in HUSM were retrieved from the microbiology database. Bacteria cultured from patients, more than 48 hours after admission were considered nosocomial. Separate analyses were performed for all cultured nosocomial organisms and for those of blood cultures only.

Results: Gram negative results were more common in the ICU/HD than in the NICU (68.0 vs 53.0 %; p <0.001). Among the commonly isolated Gram negatives, ICU/HD had in comparison to NICU more acinetobacter sp (33.0 vs 1.7 %; p<0.001), Acinetobacter Baumanii (7.6% vs. 5.7 %; p=0.151) and Pseudomonas aeruginosa (20.5 vs 15.6 %; p=0.012). Compared to ICU/HD, the NICU had more E. Coli (3.7 vs 7.9%; p<0.001) and Klebsiella Pneumoniae (13.2 vs. 37.2%; p<0.001). Among the Gram positive organisms Staphylococcus Aureus (39.1 vs 15.6 % p<0.001) was more common in ICU/HD and Coagulase negative staphylocci (30.7 vs 55.3%; p<0.001) were more common in NICU. The results for blood cultures only, were similar but some of the differences were still more pronounced.

Conclusion: The bacteriology of nosocomial infection differs significantly among wards within HUSM. Rational antibiotic policies should be ward-specific within the same hospital. Adequate hand washing upon entry of the NICU is essential to avoid import of a different nosocomial flora.
ORAL PRESENTATIONS 1

OP1.4

PREVALENCE OF SLEEP DISORDERED BREATHING (SDB) AND RISK FACTORS IN MALAY, CHINESE AND INDIAN SCHOOL CHILDREN IN MALAYSIA

Norzila Mohd Zainudin¹, Jamalludin Ab Rahman², Norrashidah AW³, Asiah Kassim⁴, Hasniah Abdul Latif⁵, Rus Anida Awang⁶, Ahmad Fadzil Abdullah⁷, Ramli Zainal⁸, Samsinah Hussain⁹

1. Paediatric Institute, Kuala Lumpur
2. International Islamic University Malaysia
3. Serdang Hospital
4. Universiti Kebangsaan Malaysia Medical Center
5. Penang Hospital
6. Tengku Ampuan Afzan Hospital
7. Institute for Health Systems Research
8. University of Malaya, Kuala Lumpur, Malaysia

Introduction: The prevalence of SDB and contributing risk factors in Malaysian school children is unknown.

Aim: To determine the prevalence of parental report of SDB and risk factors among school children aged 6 to 10 years old in three primary schools using the translated University Michigan Paediatric Sleep Questionnaire (UMPSQ).

Method: All children whose parents responded to the questionnaire and consented undergone a physical examination documenting height, weight, skin-fold thickness, neck and abdominal circumference, tonsillar size, nostril examination and presence of micrognathia or retrognathia.

Results: Data were analysed on a total of 1459 sets of questionnaires. The prevalence of parental reporting of SDB among Sekolah Rendah Jenis Kebangsaan (SRJK) school children was 15%, Sekolah Rendah Jenis Kebangsaan Cina (SJKC) 13.7% and Sekolah Rendah Jenis Kebangsaan Tamil (SJKT) 15.7%. Obesity is a risk factor of SDB in all schools. Gender is a risk factor in SRJK and SJKC school children. Increased neck and abdominal circumference, increased skin fold measurements and enlarged tonsils are significant risk factors only in SRJK school children.

Conclusion: Different schools representing different ethnicity may have different risk factors of SDB.
ORAL PRESENTATIONS 1

OP1.5

EVALUATION OF A WRITTEN ASTHMA ACTION PLAN IN THE MANAGEMENT OF CHILDREN WITH ASTHMA AT UNIVERSITY MALAYA MEDICAL CENTRE

Wong Su Sien, Anna M Nathan, Jessie A de Bruyne

Department of Paediatrics, Faculty of Medicine, University Malaya Medical Centre, Kuala Lumpur

Objectives: To evaluate the impact of a written asthma action plan on a) reducing exacerbations which require emergency department visits/unscheduled clinic visits, b) asthma control and c) quality of life in asthmatic children.

Subjects: Eighty children, between the ages of 6 and 17 years, who had asthma were enrolled in this randomised controlled single blinded study. Forty children in the study group were given a written action plan while the other forty were only given verbal instructions as to how to manage exacerbations. Other tools include Asthma Control Test, Standardized Paediatric Asthma Quality of Life Questionnaire and physician assessment of asthma symptoms were used to assess asthma control at every visit over nine months.

Results: The relative risk (RR) of exacerbation in the study group compared to the control group was 0.59 (95% CI 0.19-1.84). When it was limited to the patients with moderate to severe asthma, the RR was 0.50 (95% CI 0.05-4.94). There was no difference in the Asthma Control Test, quality of life and physician assessment of asthma control at the end of study in between the control and study group.

Conclusion: Although there was a reduction in asthmatic exacerbations in children using the written asthma action plan, this did not reach statistical significance. There was no impact on asthma control or quality of life. Larger studies looking at children with moderate-severe asthma are needed to confirm the benefit of a written asthma action plan.
OBJECTIVE: To assess medical personnel’s knowledge on Peak Expiratory Flowmetry (PEF) in children and their practical technique during PEF Test.

METHOD: Subjects were participants attended 4 Paediatric Asthma Workshops conducted by Paediatric Asthma Committee, Paediatric Department, Hospital Kuala Lumpur in 2010. Participants were given questionnaire regarding PEF test prior their practical technique assessment. The practical technique was assessed by the three doctors and nurses from the committee and they followed the same standardization.

RESULTS: Total of 89 participants were assessed and 84% of them were female. Majority are nurses (62%) followed by doctors (35%) and Medical Assistants (3%). Generally 80.9% of them had done PEF test on patients prior to attending these workshops but only 75% had done the test on paediatric patients. However, only 28% had done the test and interpret the result according the standard. About 51% referred the result to normal predictive value of respective height and gender. Fifty four (61%) participants were not aware of the availability of PEFR standards for Malaysian children.

CONCLUSION: The medical personnel’s knowledge and practical technique of PEF test for children are not adequate and this may affect the management and monitoring of childhood asthma.
ORAL PRESENTATIONS 2

OP2.2

SOCIAL DETERMINANTS (SDH) OF INFANT MORTALITY (IMR) IN MALAYSIA

Amaluddin Ahmad

Cyberjaya University College of Medical Sciences, Selangor

Objective: To examine the relationship between SDH and IMR in Malaysia.

Method: Ecological (area-based) population health survey involving all 135 administrative districts of Malaysia.

Results: Linear regressions demonstrated that 3 variables are significant determinants of infant mortality at p< 0.05 level. This includes socioeconomic status, income distribution and social development. The model was able to explain 23% of the variability observed in the IMR.

Conclusion: It is suggested that the way to further improve IMR differentials is by addressing these factors by making the necessary social and health policy changes.
NUTRITIONAL STATUS OF SCHOOL CHILDREN AGED 6-10 YEARS OLD IN SELANGOR

Asiah Kassim¹, Norzila Mohamed Zainuddin¹, Jamalludin Abdul Rahman², Ramli Zainal³, Ahmad Fadhil Abdullah⁴, Norrashidah Abdul Wahab⁵, Rusanida Awang⁶, Hasniah Abdul Latif⁷, Shamsinah Hussain⁸

1. Paediatric Department, Kuala Lumpur Hospital
2. International Islamic University, Kuantan
3. Institute of Health System Research, Kuala Lumpur
4. Kuantan Hospital
5. Serdang Hospital
6. Penang Hospital
7. University Kebangsaan Malaysia Medical Centre
8. University Malaya Medical Centre

Objective: To assess nutritional status of school children aged 6 to 10 years old in Selangor.

Method: All school children in Standard 1, 2, 3 and 4 from randomly selected seven primary schools in Selangor were given questionnaire forms. Children with parental consent underwent physical examination by the research team. Data collected include demographic, weight, height, biceps skin-fold thickness, neck circumference and waist circumference. Nutritional status was categorized following WHO 2006 growth chart standard.

Results: Total 4434 school children aged 6-10 years old participated. Nutritional status of the school children was categorized into 6 groups i.e. very thin (1.5%), thin (6.6%), normal (67.3%), overweight (12.1 %), obese (8.1%) and very obese (3.7%). About 11% of the rural school children were within thin and very thin compared to urban school children 6.5%. However, the distribution of overweight, obese and very obese school children in rural and urban schools was similar i.e. 24%.

Conclusion: The increasing trend of overweight and obesity among adult was also seen in school children aged 6-10 years old in Selangor.
Objective: To determine the rate, causes and risk factors of non-attendance to the paediatric clinic in a tertiary hospital in Malaysia and to determine the efficacy of one telephone call to confirm a new appointment.

Method: For all non-attending patients during a 2-month period a proforma was filled up based on patients’ records. During a phone call, additional questions were asked and a new appointment was offered.

Results: Of 1563 patients who had an appointment, 497 (31.8%) were non-attendees. Weather conditions, the subspecialty and timing (morning or afternoon) had a significant effect on non-attendance. Forgetfulness was the main cause. Only 160 patients could be successfully contacted. Among the contactable patients, 55 already had and 10 had reasons not to get a new appointment. Of the 95 remaining patients, 73 (76.8%) attended the new appointment.

Conclusion: The non-attendance rate was high. One telephone call had a reasonable efficacy for the contactable patients but because a high number of patients were not contactable overall effectiveness was poor.
ORAL PRESENTATIONS 2

OP2.5

WEANING PRACTICE AMONG MOTHERS IN HUSM

Nor Rosidah Ibrahim, Chiew Yee Soon, Tay Mei Ee, Tay See Hoon, Wee Shi Ning, Noraida R, Noorizan A M, Norsarwany M

Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan

Introduction: Proper weaning is very crucial for infants in achieving normal physical and mental well being. The practices vary depending on the family socio-economic status, educational level, customs, beliefs and taboos.

Objective: To identify and understand the weaning practice among mothers in Hospital Universiti Sains Malaysia (HUSM), their food preferences and food taboos.

Method: This is a cross sectional study among mothers in HUSM. All the mothers were interviewed using standard questionnaires by the researcher.

Results: Two hundreds mothers were included in the study, majority of them being Malay. More than 50% of the respondents received secondary educational level and more than 30% of them with the family income of RM 2000/month. Eighty one percent (81%) started weaning between the ages of 4-6 months of age. Seven percent started earlier and 12% started later. Three percent started between 9 -12 months. The majority of them introduced one type of food only. Majority of the mothers started weaning with porridge. 93% of them did not practice any food taboo. The minority (7%) that practiced food taboo claimed that they will not give beans (fear of bloatedness), cabbage (cold food), and pumpkin (cold food) to their babies before the age of one year.

Conclusion: Weaning during the recommended period was observed in 81% of a Kelantane population. Food taboos were rarely practiced.
ORAL PRESENTATIONS 3

OP3.1

SPECTRUM OF CONGENITAL HEART DISEASE IN HOSPITAL SULTANAH AMINAH, JOHOR BAHRU

Amelia Alias¹, Chan TL², Mohd Amin Itam¹, Mat Bah MN¹

¹. Paediatric Cardiology Unit, Paediatric Department, Hospital Sultanah Aminah Johor Bahru
². Gleneagles Hospital, Kuala Lumpur

Objective: Congenital heart disease (CHD) is the commonest form of congenital malformation. Studies have reported right sided heart lesion is more common than left heart lesion in Asian population. None of these studies, however, came from Malaysia. Our study is looking into the spectrum of CHD in our center.

Methods: This is an observational cross sectional study of all newly diagnoses CHD from 1st January 2006 to 30th July 2010. All data were entered into Pediatric Cardiology Clinical Information System. The diagnosis of CHD was made with 2D-Echocardiogram and coded with International Pediatric Congenital Cardiac Code (IPCCC).

Results: A total of 3525 patients were referred for cardiac assessment with 1517 (43%) were CHD patients. Of these 1517, 54.2% were female, 6.7% premature infant and 75.6% less than one year of age. There were 61.6% left to right shunt, 23.3% cyanotic (86 TOF) and 14.8% obstructive lesion (34 Coarctation). Using IPCCC, there were 45.7% septal defect with 498 VSD, 21.9% right heart lesion with 168 PS, 19.6% thoracic arteries and vein with 245 PDA, 4.7% single ventricle with 23 isomerism, 3.3% transposition with 38 D-TGA and only 2.1% left heart lesion with 15 HLHS. There were 192 (12.7%) duct dependent lesion with 143 (74.5%) duct dependent pulmonary circulation. Six hundred and sixty one (43.5%) lesions were considered severe lesion with 123 were large VSD.

Conclusions: The spectrum of CHD seen in our unit is comparable with previously reported studies. Our study also supports that right heart lesion are more common than left heart lesion in Asian population.
IMMUNOGENICITY OF A 3-DOSE PRIMARY SERIES OF THE PNEUMOCOCCAL NON-TYPEABLE HAEMOPHILUS INFLUENZAE PROTEIN-D CONJUGATE VACCINE (PHiD-CV) IN HEALTHY MALAYSIAN AND SINGAPOREAN INFANTS

Fong Seng Lim¹, Kah Kee Tan², Poh Chong Chan³, Chia Yin Chong⁴, Mia Tuang Koh⁵, Yee Leong Teoh⁶, Aurélie Fanic⁷, Ilse Dieussaert⁷, Kristien Swinnen⁷, Dorota Borys⁷

1. NHG Polyclinics, Singapore
2. Hospital Tuanku Jaafar, Seremban, Malaysia
3. National University Hospital, Singapore
4. KK Women and Children’s Hospital, Singapore
5. University Malaya Medical Centre, Kuala Lumpur, Malaysia
6. GlaxoSmithKline Biologicals, Singapore
7. GlaxoSmithKline Biologicals, Wavre, Belgium

**Objectives:** To demonstrate immunological non-inferiority of the PHiD-CV commercial lot versus the phase III clinical lot and to evaluate the immunogenicity, safety and reactogenicity of PHiD-CV in Malaysian and Singaporean infants.

**Methods:** Infants (N=466, aged 6–12 weeks at first dose) were randomised 1:1 to receive either the commercial (Com group) or clinical lot (Clin group) of PHiD-CV at 2-3-5 months of age, each co-administered with GSK’s DTPa-combined and human rotavirus vaccines according to local schedules. Immune responses were measured 1 month post-dose 3 using 22F-inhibition ELISA and opsonophagocytic activity (OPA) assays for vaccine pneumococcal serotypes (VT), and ELISA for protein-D. Non-inferiority was achieved if the UL 95%CI of the adjusted antibody GMC ratio (clinical over commercial lot), measured by ELISA, was below 2 for each VT and protein-D.

**Results:** In both groups, ≥93.6% of infants reached antibody concentrations ≥0.2µg/mL and ≥88.5% reached OPA titres ≥8 for each VT. For cross-reactive serotypes 6A/19A respectively, ≥85.5% and ≥37.7% of infants reached OPA titres ≥8, and ≥60.6% and 54.6% reached antibody concentrations ≥0.2µg/mL; percentages were comparable between groups. All infants except one (Com group) had measurable antibodies against protein-D (≥100 EL.U/mL). Non-inferiority was met for each VT (ULs≤1.45) and protein-D (UL=1.58). Immune responses to co-administered vaccines and safety/reactogenicity profiles were in-line with previous studies.

**Conclusion:** PHiD-CV in a 3-dose primary series was immunogenic for all VT and protein-D in Malaysian and Singaporean infants. Immune responses induced by a PHiD-CV commercial lot were non-inferior to those induced by the phase III clinical lot.
THE ROLE OF ROUTINE RSV SCREENING IN CHILDREN PRIOR TO SURGERY AND CARDIAC CATHETER PROCEDURE IN INSTITUT JANTUNG NEGARA

Leong Ming Chern, Khairul Faizah, Adura Abdul Rauf, Hasri Samion, Mazeni Alwi

Department of Paediatric Cardiology, Institut Jantung Negara, Kuala Lumpur

Background: Respiratory Syncytial Virus (RSV) infection confers higher morbidity and mortality post cardiac surgery.

Objective: We study the role of routine rapid RSV screening in patients going for elective cardiac procedure or surgery.

Methodology: A prospective study of all patients aged less than 2 years, admitted electively from March 2008 till March 2009 was undertaken. RSV screening was done using rapid chromatography and the course of the patient was followed up to assess the length of stay in the ICU, in the hospital and for death. The children were divided into RSV positive and RSV negative.

Results: A total of 130 patients were recruited, of whom 115 were clinically fit to proceed for surgery or intervention. 15.7% of the patients admitted for surgery/procedure were noted to be positive for RSV. Being oxygen / ventilator dependent, having pulmonary hypertension, heart failure and cyanosis did not increase the yield of RSV positivity. There was no statistical difference in the length of hospital stay and ICU stay except patients with heart failure (p=0.044). RSV positive group did not demonstrate increased risk of perioperative mortality (Gp A 10.3% vs Gp B 11.0%).

Conclusion: Routine RSV screening was not indicated for patients undergoing for elective cardiac surgery and procedure.
SAFETY OF MMR VACCINATION. IS ADMISSION NECESSARY?

Muzhirah Aisha Haniffa

Department of Paediatrics, Hospital Selayang, Selayang, Selangor

There has been long standing concern about the safety of measles-mumps-rubella (MMR) vaccine in children with history of egg allergy. We audited our experience in vaccinating such children in our hospital over a year period. Referrals were from nearby health clinics in Gombak area. There were in total 33 patients admitted to paediatric ward from Jan 2009 till December 2009. Patients were observed for 2 hours with hourly vital signs monitoring following MMR (Priorix) vaccination prior to discharge. None developed rash, swelling or anaphylactic reactions following injection. Similar experiences were encountered from other hospitals locally whereby no patients had developed any cardiopulmonary reactions after MMR vaccination. Clinical evidences and published guidelines on the risk of anaphylactic reaction and complications after MMR vaccine is reviewed. A revised approach for the vaccination of children with history of egg allergy is proposed. Local Committee in Infection and Immunization need to address this issue earliest possible and safety profile of this vaccine need to be disseminated to primary care givers.
REAL-TIME QUANTITATIVE PCR CHIMERISM ASSAY AFTER BONE MARROW TRANSPLANTATION (BMT) IN SEX MATCHED SIBLING

Aliza Mohd Yacob¹, Ida Shahnaz Othman², Hishamshah Ibrahim², Raba’atun Sham¹, Fateha Misnar¹, Erniza Assari¹, Maznah Abu Bakar¹, Ezalia Esa¹, Zubaidah Zakaria¹

1. Institute for Medical Research
2. Institute of Paediatrics, Hospital Kuala Lumpur

Objective: To determine absolute level of chimerism for monitoring engraftment in sex–matched related pair by real time quantitative PCR.

Methods: Blood samples were from a male patient Acute Myeloid Leukemia at day 16, 28 and 49 who underwent allo-BMT from his donor brother. DNA was extracted using QIAamp Blood MIDI kit. Before quantification, the donor and recipient were genotyped using 19 biallelic genetic markers. Informative alleles were determined and only one marker was used for each genotype profile. Genotyping and all chimerism quantifications were performed by real-time PCR using LC480, Roche.

Results: SO3 genetic marker located on chromosome 6q was informative in patient and was used in recipient-specific allele amplification. Decrease of recipient genotype % from day 0 to day 49 after allo-BMT was observed with 100% at day 0, 9% at day 16, 6% at day 28 and 4% at day 49 and correlates with clinical findings.

Conclusion: Real-time quantitative PCR chimerism assay is feasible and useful for monitoring engraftment even in sex matched sibling with identified genetic marker and a reproducible sensitivity of up to 1:1000 cells.
POSTER PRESENTATIONS

P2

PREVALENCE OF LATE DIAGNOSIS OF DUCT DEPENDENT AND CYANOTIC CONGENITAL HEART DISEASE IN HOSPITAL SULTANAH AMINAH JOHOR BAHRU

Amelia Alias¹, Chan TL², Mohd Amin Itam¹, Mat Bah MN¹

1. Paediatric Cardiology Unit, Paediatric Department, Hospital Sultanah Aminah Johor Bahru
2. Gleneagles Hospital, Kuala Lumpur

Objective: Late diagnosis of duct dependent and cyanotic congenital heart disease (CHD) may have significant impact on the outcome of CHD. However, there is no study in Malaysia looking at this issue. Therefore we studied the prevalence of late diagnosis of this lesion in our centre.

Methods: This is an observational cross sectional study of all newly diagnosed CHD seen from 1st January 2006 to 30th July 2010. The diagnosis of CHD was made with 2D-Echocardiogram. All data were entered into Pediatric Cardiology Clinical Information System. Non Malaysian Children were excluded.

Results: A total of 1517 CHD patients were seen during the study period, with median age of diagnosis of 90days [Q1 13days Q3 350days]. There were 192 (12.7%) duct dependent lesions with 74.5% were duct dependent pulmonary circulation. The median age of diagnosis of duct dependent lesion were 4.5days [Q1 1day, Q3 23days]. Meanwhile there were 352 (23.2%) cyanotic lesion with 86 (24.4%) were Tetralogy of Fallot. The median age of diagnosis of cyanotic CHD were 12days [Q1 1day, Q3 93days]. There were 39 (11%) cyanotic CHD diagnosed after 1 year of life with 20 Tetralogy of Fallot. Nineteen (9.8%) duct dependent were diagnosed after 3 month of life (3 each for severe coarctation, PAVSD and severe pulmonary stenosis, 2 each for isomerism, mitral atresia, tricuspid atresia, PAIVS, DORV with pulmonary atresia).

Conclusion: There were significant numbers of late diagnosis of cyanotic and duct dependent lesion with prevalence of 11% and 9.8% respectively. Further study is needed to understand the reason behind this.
POSTER PRESENTATIONS

P3

LIPAEMIC BLOOD: A CASE REPORT

Ashikin MN, Teoh SL, Loo HM, Kuan GL

Department of Paediatrics, Malacca Hospital, Malacca

Case report: A previously healthy 3 month old infant, a product of a non-consanguinous marriage presented with incidental finding of gross hepato-spleenomegaly associated with significant anaemia and thrombocytopenia. Initial presentation mimicked a haematological malignancy. However full blood picture, bone marrow aspiration, inborn error of metabolism, primary immunodeficiency screening and infectious disease screening were all normal. One striking feature to note was presence of a lipaemic blood sample. MAK was diagnosed to have Lipoprotein Lipase (LPL) deficiency, a rare autosomal recessive lipid metabolism disorder.
DEMOGRAPHIC PROFILE AND PHYSICAL CHARACTERISTICS OF CHILDHOOD OBESITY IN UNIVERSITY MALAYA MEDICAL CENTER

Azriyanti AZ, Rosmini T, Yazid J, Fatimah H

Department of Paediatric, Faculty of Medicine, University Malaya, Kuala Lumpur, Malaysia

Introduction: The prevalence of childhood obesity in Asian countries is still rising. In Malaysia, health survey in 2008 reports 24% of our children was overweight and obese.

OBJECTIVE: To determine the demographic profile and physical characteristics of overweight and obese children presented to University Malaya Medical Centre (UMMC).

Methods: A total of 310 new patients were seen in Paediatric Obesity clinic from 2005-2010. Only 205 patients’ records were available for evaluation. All children have had their anthropometrics measurements taken, including waist circumference, presence of acanthosis nigricans documented and BMI calculated. The children’s birth weight was also noted. Obesity is defined as BMI >97th centile and, overweight if BMI > 85th centile respectively for age and sex.

Results: One-hundred and ninety (92.7%) children were obese and 15 (7.3%) were overweight. There were 117 (57.1%) male and 88 (42.9%) female children studied. One-hundred and forty (68.3%) were Malays, 14(6.8%) Chinese and 51(24.9%) Indian children included. The mean for age was 9.5± 3 years, BMI 28± 5.7 kg/m², waist circumference for male children was 90.4 ± 12.4 cm and female children were 85.2± 14.2 cm. The mean systolic blood pressures were 114± 15 mmhg and diastolic blood pressure were 69.5± 11 mmhg. Seventy –six percent (n=150) of these children has acanthosis nigricans. Amongst these children, 46 (24.4%) were either born small or large for gestational age.

Conclusions: This study showed that childhood obesity predisposes risk for serious co-morbidities. Referring them early would be beneficial to detect early complications.
POSTER PRESENTATIONS

P5

NUTRIENT DRINKS’ AND HEPATIC IMPAIRMENT IN CHILDREN: A CASE REPORT

Cham Weng Tarng, Tang Swee Ping, Lim Sern Chin

Paediatric Rheumatology Unit, Department of Paediatrics, Hospital Selayang, Selayang

Introduction: Dietary health food supplementation is rampant today with nutrient drinks being the latest fad. Nutrient drinks are touted to boost the immune system and are safe.

Methods: We describe 2 children with autoimmune diseases and previously normal liver function who developed hepatic impairment following consumption of nutrient drinks.

Case report 1: is a 12-year old Chinese boy, with SLE Class IV Lupus nephritis undergoing intravenous Cyclophosphamide therapy. He presented 1 week after his third dose of iv Cyclophosphamide with 2 days of jaundice. His liver was 4cm palpable and had cholestatic jaundice and hepatic impairment (ALT 1419 iu/L, AST 604 iu/L). He started consuming Juice A (a grape juice concentrate) 4 days prior to jaundice onset. Upon stopping Juice A, his elevated transaminases improved only to deteriorate again necessitating a liver biopsy which showed features of ‘drug induced hepatitis’. Further history revealed consumption of Juice B (a cactus juice concentrate with honey). After stopping all juices, his liver impairment gradually improved and resolved after 2 months.

Case report 2: is a 3-year old Chinese boy with undifferentiated autoimmune disease receiving iv Cyclophosphamide therapy. He was asymptomatic but incidentally noted to have elevated transaminases (ALT 223 iu/L, AST 135 iu/L). Further history revealed consumption of Juice B. After stopping Juice B, the elevated transaminases normalized within 1 month.

Conclusion: A specific history of health supplement ingestion must be sought in any child with hepatitis as most parents do not consider these as ‘drugs’. Nutrient drinks can be potentially harmful, causing hepatic impairment especially in the background of other existing diseases or concomittant drug ingestion.
Background: Trauma is a leading cause of paediatric emergencies in Malaysia. Little data exists about paediatric injury patterns in Malaysia. Identification of injury patterns and causes, as well as specific high-risk paediatric population groups may enable development of improved prevention strategies and care.

Methodology: Data from all trauma cases presenting to the Paediatrics Accident and Emergency Unit of the University of Malaya Medical Centre (UMMC) in Kuala Lumpur, Malaysia in November and December 2009 were recorded. Patient demographics, socioeconomic data, causes of injuries, types of injuries, admissions and specialties involved were analyzed for each age group.

Results: A total of 295 paediatric trauma cases presented to UMMC in November and December 2009 constituting 12.8% of the total number of patients seen. Injury rates were higher amongst the younger age groups, and decreased with age, with boys outnumbering girls in all age groups. Either or both parents were the commonest caretakers at time of injury. Falls were the commonest cause of injuries (46% of total causes) while soft tissue injuries were the commonest injuries sustained (26% of total injuries). Injuries at home were more common in younger children, while injuries in public places were more common in older children. Average admission duration was 2.6 days.

Conclusion: The patterns of paediatric injury presenting to UMMC correlated with age and socioeconomic background. Our findings were consistent with studies done elsewhere. Further work needs to be done in the field of injury prevention.
POSTER PRESENTATIONS

P7

A CASE REPORT: HYPERCALCIURIC BARTTER’S SYNDROME WITH NEPHROCALCINOSIS

BH Ching¹, PY Tam¹, Janet YH Hong²

1. Department of Paediatrics, Hospital Pakar Sultanah Fatimah, Johor, Malaysia
2. Department of Paediatrics, Hospital Putrajaya, Malaysia

Introduction: Bartter’s syndrome is a rare autosomal recessive renal tubular disorder affecting sodium, potassium, chloride reabsorption in the thick ascending loop of Henle. It is characterized by hypokalaemia, hypochloraemia, metabolic alkalosis, hyperreninemia and hyperaldosteronaemia with normal blood pressure.

Case report: A baby girl delivered prematurely at 32 weeks following a pregnancy complicated by severe polyhydramnios presented at 10 months old with urinary tract infection. She had delayed development with failure to thrive. Investigations showed persistent hypokalaemia, hypochloraemia, hypercalciuria, metabolic alkalosis with normal blood pressure. Her plasma renin and aldosterone levels were elevated. Karyotyping showed an increase in length of the centrometric heterochromatin of one of the chromosome 6 at 6p11.1. Her third sister who passed away at the age of 10 months old, was also born prematurely following a pregnancy complicated with polyhydramnios. Bartter syndrome was considered. With high dose oral potassium supplements, her growth and mental development improved. On follow up at the age of 5 years, she is developing normally. However, her ultrasound revealed bilateral medullary nephrocalcinosis.

Conclusion: This girl has antenatal form of Bartter’s syndrome. Early diagnosis and treatment is important to prevent life-threatening volume depletion in the neonatal period. As this condition is accompanied by markedly elevated renal and extrarenal Prostaglandin E₂ (PGE₂) production, treatment with Prostaglandin synthetase inhibitors can improve polyuria and hypokalemia besides improving growth.
TREATMENT OF SUBCUTANEOUS PANNICULITIS-LIKE T-CELL LYMPHOMA
WITH CORTICOSTEROID MONOTHERAPY: A CASE REPORT

Chong Lee Ai, Manimalar Naicker, Lalitha Pillay, Hany Ariffin

Departments of Paediatrics and Pathology, University of Malaya Medical Centre, Kuala Lumpur

Introduction: Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) is a distinctive skin lymphoma characterized by infiltration of the subcutaneous tissue by malignant T cells. SPTCL was previously defined as an aggressive T-cell lymphoma requiring multiagent chemotherapy. Recent re-classifications of SPTCL by immunohistochemistry (IHC) have suggested a unique entity warranting changes in conventional treatment.

Case report: A 9-year old boy presented with 2 months history of bilateral cheek swelling which was erythematous but painless and non-pruritic. He also had a month history of intermittent high grade fever, loss of appetite and loss of weight. Examination revealed submandibular lymphadenopathy of up to 1.5cm and hepatomegaly of 2cm below the costal margin. All his blood investigations were normal. CT of his head revealed diffuse soft tissue swelling at the mandibular region extending bilaterally to the maxillary area. Staging CT was normal. Bone marrow and cerebrospinal fluid were also normal. Histopathological examination of his right cheek biopsy confirmed the diagnosis and on IHC staining, the abnormal lymphocytes were CD8+, CD4- and CD56-. He commenced oral prednisolone 2mg/kg/day for 4 weeks and weaned off steroids over another 4 weeks. Complete resolution of the cheek swelling was seen after a week of prednisolone. Hepatomegaly and lymphadenopathy resolved at the end of therapy.

Discussion: SPTCL may be subclassified according to their T-cell receptor (TCR) phenotype. TCRαβ phenotype, usually CD4-, CD8+, CD56-, can achieve long-term remission with systemic corticosteroids and have an excellent prognosis. Our patient, now three months off therapy, remains in complete remission.
POSTER PRESENTATIONS

P9

PENILE TUMOUR IN A 2 YEAR OLD CHILD AT HUSM: A CASE REPORT

Fahisham T, Norsarwany M, Mohd Ashraf MD, Ariffin N

Department of Paediatrics, Universiti Sains Malaysia, Kelantan

Introduction: Penile cancer is extremely rare in paediatrics.
Case report: We present a case of large fungating penile lesion in a 2-year-old Malay boy. The diagnosis was confirmed as infantile fibrosarcoma on histological examination of the lesion. Follow-up after starting on chemotherapy showed partial recovery of penile structure. The diagnostic and therapeutic aspects are discussed in this article.
POSTER PRESENTATIONS

P10

ATYPICAL PRESENTATION OF CHILDHOOD LEUKAEMIA

Florence Wong, Grace Chow Kam Yoke, Tang Swee Ping, Cham Weng Tarng

Department of Paediatrics, Hospital Selayang, Selangor

Introduction: Acute lymphocytic leukaemia (ALL) is the commonest malignancy in children presenting generally with symptoms and signs of bone marrow infiltration and/or extramedullary disease. The full blood count (FBC) may show cytopenias and peripheral leukaemic blasts are seen in 90% of cases. Central nervous system (CNS) involvement is rare (3%).

Case report: We describe a 10-month old Chinese male infant with ALL who presented predominantly with CNS disease with no evidence peripherally of bone marrow infiltration or leukaemia blasts. This infant presented with 2 days history of left periorbital swelling with preceding fever, and was noted to have an acute flaccid paralysis involving both lower limbs with acute urinary retention. His liver was 3 cm palpable, with no splenomegaly or significant lymphadenopathy. 42 hours after admission, he developed paradoxical breathing and required ventilation. All his blood investigations in particular the FBC (Hb 14.4g/dl, WCC 9.36 x10⁹, Neutrophils 69%, Lymphocytes 26.5%, Platelets 540,000) and peripheral blood film (no blasts seen) were normal. Urgent Magnetic resonance imaging (MRI) showed an extra axial left posterior fossa tumour causing obstructive hydrocephalus, multiple extradural masses in the spinal cord especially a large mass at the level of C3 to T6 causing cord compression, and a mass superior to left orbit. He underwent emergency craniotomy, laminectomy and tumour debulking surgery. Biopsy of the cranial mass as well as bone marrow (BM) immunophenotyping confirmed the diagnosis of B lymphoblastic leukaemia.

Conclusion: The absence of cytopenias or peripheral blasts does not necessarily exclude the possibility of ALL.
POSTER PRESENTATIONS

P11
LANGERHANS CELL HISTIOCYTOSIS – A RARE CASE OF SEVERE PULMONARY INVOLVEMENT

Haiza Hani Hamidon, C-Khai Loh, Hamidah Alias
Haemato-oncology Unit, Department of Paediatrics, Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur

Background: Langerhans Cell Histiocytosis (LCH) is a dendritic cell disorder, resulting from proliferation of immunophenotypically and functionally immature Langerhans cells with eosinophils, macrophages, lymphocytes and multinucleated giant cells. The incidence of LCH is 2 to 10 cases per million children aged less than 15 years old. Pulmonary LCH is rare in childhood. It typically affects young adults and associated with cigarette smoking. We describe a case of LCH with severe pulmonary involvement.

Case report: A 9-year-old, Chinese, boy, with 3-month history of constitutional symptoms and gum swelling presented with sudden onset of left sided chest pain and difficulty in breathing. His chest X-ray showed bilateral diffuse cystic lung changes and pneumothoraces. His HRCT thorax revealed multiple diffuse, irregular-shaped cystic lesions of varying sizes in both lung fields suggestive of pulmonary LCH. Biopsy of the gum confirmed Langerhans cell histiocytosis. He developed diabetes insipidus secondary to posterior pituitary infiltration, confirmed by MRI pituitary and was started on oral Minirin. He subsequently was started on multisystem risk regime as per LCH III UK Protocol. He received 4 weeks of Prednisolone (60mg/m²/ daily) and Vinblastin bolus (6 mg/m²) one week a part, 6 doses in total. It was a challenge for us to manage the recurrent tension pneumothorax which required chest tubes in-situ for prolong period of time. In view of this, he underwent bilateral thoracotomy and mechanical pleurodesis.

Conclusion: We presented a case of severe pulmonary LCH with challenge in managing his recurrent bilateral pneumothoraces.
**POSTER PRESENTATIONS**

**P12**

**LI-FRAUMENI SYNDROME IN MALAYSIAN FAMILIES**

Hany Ariffin, Chong Lee Ai, Zarina A Latiff, Mahfuzah Mohamed, Eni Juraida

Malaysian Society of Paediatric Haematology and Oncology (MASPHO)

**Objective:** The Li-Fraumeni syndrome (LFS) is a rare autosomal familial cancer syndrome characterized by an increased incidence of malignancies in family members aged less than 45 years. In 1990, the association between LFS and germline mutations of the TP53 tumour suppressor gene was demonstrated. We aimed to identify TP53 mutations in patients who fulfilled the phenotypic criteria for LFS to document its incidence and for counselling purposes.

**Methods:** Genomic DNA was extracted from peripheral blood leucocytes of the affected patients. Pre-test and post-test counselling was given to all affected families. Exons 2 to 11 including splice junctions of the TP53 gene on chromosome 17 was amplified following parameters available from www-p53.iarc.fr followed by DNA sequencing. Sequencing results were analysed by the study’s central geneticist. All PCR products were sent to the International Agency for Research on Cancer (IARC/WHO) for verification and confirmation.

**Results:** Three affected families, treated at the various paediatric oncology centres in Malaysia have been identified, including one with a previously unreported duplication of a six base-pair motif (GGCGTG) in Exon 10.

**Conclusions:** Through coordinated effort and sharing of facilities using the MASPHO platform, we may be able to determine the incidence of LFS in Malaysia. This information will also allow us to make recommendations for cancer screening in asymptomatic carriers of TP53 mutations for early diagnosis.
POSTER PRESENTATIONS

P13

CASE REPORT: AGROBACTERIUM RADIOBACTER INFECTION IN PAEDIATRIC ONCOLOGY PATIENT, HUSM EXPERIENCE

Hayati J\(^1\), Norsarwany M\(^1\), Siti Asma’ H\(^2\), Fahisham T\(^1\), Ariffin N\(^1\)

1. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia
2. Department of Microbiology, School of Medical Sciences, Universiti Sains Malaysia

**Introduction:** Agrobacterium radiobacter is an aerobic, non-spore forming Gram negative bacillus belongs to genus Agrobacterium. They are widely distributed in soil environment. Human infection by this organism is rare and mostly occur in immunocompromised patients.

**Case report:** We report a case of Agrobacterium radiobacter septicaemia in a child patient with the diagnosis of Acute lymphoblastic leukemia (ALL) and described the clinical presentations, treatment strategies and outcome of Agrobacterium infections based on our experience.

**Conclusion:** Removal of the central venous catheter and administration of antimicrobial therapy led to favorable outcomes in this patient.
POSTER PRESENTATIONS

P14

MOWAT WILSON SYNDROME CAUSED BY DE NOVO CHROMOSOME 2 PERICENTRIC INVERSION

YL Heng, WT Keng, Roziana Ariffin, Farah Amalina, Aminah Mekesat

Department of Genetics, Hospital Kuala Lumpur

Introduction: Mowat Wilson Syndrome is characterized by dysmorphic features particularly large and uplifted ear lobes and a prominent but narrow and triangular pointed chin as well as moderate to severe intellectual disability including severe speech delay. Nearly all have microcephaly and epilepsy. Congenital anomalies, including Hirschsprung disease, congenital heart disease, hypospadias, genitourinary anomalies, agenesis of the corpus callosum, eye anomalies and short stature are common. MWS is caused by heterozygous mutations or deletions in the Zinc finger E-box-binding homeobox 2 gene, ZEB2, previously called ZFHX1B (SIP1) on chromosome 2q22-q24.1. The majority of MWS cases reported so far were sporadic, therefore the recurrence risk is low.

Case report: We report a Mowat Wilson Syndrome patient who has a de novo pericentric inversion involving chromosome 2.

Conclusion: We hypothesized that the break point on the long arm of chromosome 2 has disrupted ZFHX1B gene, causing Mowat Wilson Syndrome in this patient. Further work will be performed to map the break point.
POSTER PRESENTATIONS

P15

OCHOA SYNDROME: SMILE TO DIAGNOSE A NEUROGENIC BLADDER

Indra Ganesan
Sabah Women and Children’s Hospital, Sabah

Case report: We report on an 8 year old girl who had presented with acute retention of urine at age 3 years requiring bladder catheterization. The neurological examination of the lower limbs were normal and she did not have spinal dysraphism. She subsequently had recurrent urinary tract infections. Renal ultrasound showed bilateral hydronephrosis, and micturating cystourethrogram revealed a trabeculated bladder with grade III VUR into the left kidney and a non refluxing right kidney. Urodynamic studies documented high opening pressures with a urinary retention pattern. MRI of spinal cord was normal. As for her urinary habits, she is unable to void fully on her own. Urinary stream is poor, and there is a constant dribbling of urine. She was subsequently started on clean intermittent catheterization. The initial renal impairment with elevated serum creatinine (130 umol/l) subsequently normalized. She has a strikingly peculiar facies in which there is an inversion of the facial expression where she has the appearance of frowning and looking sad when she smiles.

Conclusion: This is a well described urofacial syndrome also known as Ochoa syndrome.
POSTER PRESENTATIONS

P16

SHORT STUDY OF ACUTE GASTROENTERITIS IN PUTRAJAYA HOSPITAL, FROM APRIL TO JUNE 2010

Janet YH Hong¹, Sofia ANM², Fuziah MZ¹

1. Department of Paediatrics, Putrajaya Hospital
2. Cyberjaya University College of Medical Sciences

Introduction: Putrajaya Health and Morbidity Survey 2006 showed that the prevalence of acute gastroenteritis (AGE) in children (< 17 years old) was 6.3%.

Objective: To study the clinical and biochemical characteristics of AGE cases admitted to paediatric ward, Putrajaya Hospital.

Method: The electronic medical records were reviewed retrospectively from 1st April to 30th June 2010.

Results: A total of 42 patients were included of which 57.1% were first admissions. There were 61.9% boys. There were 42.9% children who were less than 12 months. Majority (78.6%) of the patients had working parents. Patients presented with severe vomiting (45.2%) of up to 6 – 10 times per day and 54.8% had severe diarrhoea of 6 – 10 times per day. Abnormal skin turgor was noted in 2.3%, sunken eyes in 71.4% and dry mucus membrane in 28.6% of patients. Most of them (66.7%) had decreased feeding and 19.0% had decreased urine output. Dehydration was seen in 69.0% of patients, of which 69.0% had <5% dehydration, 27.6% had 5% dehydration and 3.4% had 10% dehydration. Hyponatraemia and hypokalaemia were noted in 64.3% and 40.5% of patients respectively. Marginally elevated blood urea level occurred in 4.8% of patients. Most of the patients with dehydration received intravenous fluid therapy.

Conclusion: Majority of children admitted to Putrajaya Hospital with AGE had mild dehydration, indicating that parents sought early medical treatment. Most of the patients were taken care by caregivers. Hence, education on proper food preparation and personal hygiene should be emphasized to all caregivers.
POSTER PRESENTATIONS

P17

TRENDING OF CHILD ABUSE CASES PRESENTED TO PUTRAJAYA HOSPITAL

Janet YH Hong¹, AB Ismail¹, Jabbar Al¹, JSL Wong¹, Ashwin Kaur JS¹, Fuziah MZ¹, Nora’i MS²

1. Department of Paediatrics, Putrajaya Hospital, Putrajaya
2. Department of Administration, Putrajaya Hospital, Putrajaya

Introduction: Child abuse cases are on the rise, with the highest incidence reported in Selangor and Kuala Lumpur in 2008 by Jabatan Kebajikan Masyarakat Malaysia.

Objective: To observe the pattern of child abuse cases and to identify associated features and possible contributing factors.

Method: The electronic medical records were reviewed retrospectively from 1st January 2007 to 31st December 2009.

Results: A total of 56 children were included for the study period (13 in 2007, 20 in 2008, 23 in 2009), with a predominance of male (60.7%) and children less than 2 years old (76.8%). Patients consisted of Malay (89.3%), Chinese (5.4%), Indian (1.8%) with age ranged from 0 – 14 years. Majority (62.5%) of the incidence occurred outside Putrajaya. There was an increasing tendency for abuse cases to occur outside home over the years (from 46.2% to 54.5%). The types of injuries ranged from head injury (73.2%), bruises (14.3%), fractures (7.1%), sexual abuse (1.8%) and others (3.6%). Of head injury cases, 39.0% had intracranial bleed. The prevalence of mortality was 30.8%, 60.0%, 39.1% in 2007, 2008 and 2009 respectively, whereby only 40.0% had post-mortem performed.

Conclusion: The increasing trend of child abuse may reflect the increasing population in and around Putrajaya consisting of mostly working parents who depend on child care services. Children younger than 2 years old had higher risk of being abused and should be given more attention. Intracranial bleed is the leading cause of death and public awareness on prevention is warranted.
POSTER PRESENTATIONS

P18

PARENTS’ PERCEPTION AND SATISFACTION OF QUALITY FAMILY-CENTERED CARE IN PEDIATRIC WARDS AT HOSPITAL UNIVERSITI SAINS MALAYSIA

Khairun Nisa Mohammad, Jayah K Pubalan

School of Health Sciences, Health Campus, Universiti Sains Malaysia, Kelantan, Malaysia

Background: The care of hospitalized children becomes a family – centered issue and it is the best way to deliver care to children in health care centers.

Methods: This cross-sectional study was conducted to determine the parents’ perception and satisfaction of quality family-centered care varied across the pediatric wards. Convenience sampling was applied and information gathered from 65 parents of hospitalized children using Parent Questionnaire on Family – Centered Care using five Likert scales.

Results: The mean level of parents’ perception of family-centered care was 94.72 (SD = 20.12) with score of 72.76%. The differences of parents’ perception of family-centered care among health issue of medical, surgical and oncology were not significantly different (F = 0.137, p > 0.05). The mean level of parents’ satisfaction of family-centered care was 113.97 (SD = 13.37) with score of 41.53%. The differences of parents’ perception of family-centered care among health issue of medical, surgical and oncology were not significantly different (F = 0.39, p > 0.05). The differences of parents’ satisfaction of family-centered care among health issue of medical, surgical and oncology were not significantly different (F = 0.39, p > 0.05). Study also showed that there were no significant different between overall perception and overall satisfaction in pediatric wards (chi square = 1.377, p > 0.05).

Conclusion: The six elements of family-centered care that being explored in this study showed positive perception. Thus, more positive perception of family – centered care affects more satisfaction of family – centered care and increases its quality.
POSTER PRESENTATIONS

P19

PARENTS VIEWS OF LUMBAR PUNCTURE IN CHILDREN

Wong SLJ, Yeoh AAC, Ooi TC, Lye CS

Department of Paediatrics, Hospital Seberang Jaya

Introduction: A descriptive study was carried out to obtain the views of parents regarding lumbar punctures.

Methods: A total of 58 children were recruited over a one month period. 12% of these children were admitted for febrile seizures, 3% had epilepsy with breakthrough seizures and the rest were admitted for a range of diagnosis which includes pneumonia, acute gastroenteritis and urinary tract infection. Either one of the parents was asked to complete a questionnaire during admission.

Results: The ethnicity of the patients was Malay (85%), Chinese (9%), Indians (5%) and others (1%). 20% of parents have not heard of lumbar puncture (LP) before. 46% of parents said they will be agreeable for LP for their children if requested by the doctor, 31% will refuse and 22% are unsure. LP was requested and done in only one patient in this study. The main reasons for refusal were fears that it may cause paralysis (48%), mental retardation (6%) and relatives/friends influence (16%). The main reason given by those who would consent to LP were because they think it is therapeutic (17%), to aid in the diagnosis (11%) and based on doctor’s advice (5%). The main sources of information on LP for the parents were their relatives/friends.

Conclusion: In conclusion, LP refusal is a common problem in local setting and there are still misconceptions and apprehensions on it. Appropriate measures must be carried out to educate the public on the indication and safety of the procedure.
MITOTANE RELATED VAGINAL BLEEDING IN A GIRL WITH RECURRENT ADRENOCORTICAL CARCINOMA

Karmila A, Nurshadia S, Jalaludin MY, Harun F

Department of Paediatrics, Faculty of Medicine, University Malaya

**Introduction:** Mitotane is the only adrenal-specific agent available for treatment of residual adrenocortical carcinoma (ACC) after surgery. It exerts a specific, direct cytotoxic effect on adrenal cell mitochondria and impairs adrenal steroidogenesis. Mitotane has been associated with increased binding capacity of steroid hormone binding globulin (SHBG) and modulates their disposal for target cell. We report a rare case of mitotane associated vaginal bleed in a girl with recurrent ACC.

**Case report:** A 7-year old girl, with underlying left ACC Stage III diagnosed in May 2007 presented with tumour recurrence two years later. During her initial presentation, she had a course of mitotane after the left adrenalectomy. She experienced bilateral breast enlargement without vaginal bleeding, which resolved upon completion of chemotherapy. In early 2010, she had recurrence of tumour with liver and lymph nodes metastases. Tumour was resected and another course of daily oral mitotane was commenced. Vaginal bleed was noted 8 weeks after starting mitotane. She was thin and had no rapid height gain. Her areola was pigmented but there was no breast enlargement, nor axillary or pubic hair. Pelvic ultrasound showed thickened endometrium with no ovarian abnormality. Hormonal study confirmed prepubertal response (baseline LH <0.1mIu/ml, FSH <0.3mIu/ml and oestradiol <37pmol/L with no rise of LH/FSH during LHRH stimulation test). Vaginal bleed ceased with the introduction of progestogen.

**Conclusion:** Vaginal bleeding can occur during mitotane therapy due to increased binding capacity of SHBG. Hormonal investigation is usually consistent with prepubertal response. Symptomatic treatment and reassurance is important in managing such cases.
Objective: To determine the incidence of RSV in Acute Lower Respiratory Infections (ALRI) and to identify the seasonal pattern and clinical features of RSV infection in children admitted to Sabah Women and Children’s Hospital (SWACH).

Methods: Nasopharyngeal aspirates for respiratory viruses were taken for all children admitted with ALRI from June 2008 to July 2010. Case notes of those positive for RSV infections were retrieved and analyzed retrospectively.

Results: The incidence of RSV infection is 23.4% during the study period. A distinct peak in RSV infection was seen from October to February in 2008 - 2010 and this occurs 2 to 3 months after the onset of rainy season. A total of 75 children with RSV infection during the study period were reviewed, and there were 47 (62.7%) males. Majority 43 (57.3%) were less than 6 months old, 17 (22.7%) were 6-12 months and 15 (20%) of them were more than 1 year old. 8 (10.6 %) patients had underlying medical condition whereby 7 (9.3%) had history of prematurity and 1 patient is syndromic.

The most common presenting features were cough (96%) and rapid breathing (81.3%). The average length of stay was 4.5 days. Eight patients (10.7%) required intensive care for severe RSV disease, where 4 had underlying medical condition. Four patients were ventilated and there were no deaths from RSV disease during the study period.

Conclusions: RSV is a significant pathogen of acute lower respiratory tract infection of children in SWACH. A definite RSV season is seen consistently from October to February, 2 to 3 months after the rainy season. Majority of the patients were less than 6 months of age and a significant proportion require ICU care.
POSTER PRESENTATIONS

P22

INTRAVENOUS IMMUNOGLOBULIN IN INFLUENZA A H1N1-ASSOCIATED ACUTE NECROTIZING ENCEPHALOPATHY: A CASE REPORT

Koh Lee Min, Tang Swee Ping, Lim Sern Chin, Cham Weng Tarng

Department of Paediatrics, Hospital Selayang, Selangor

Introduction: Acute necrotizing encephalopathy of childhood (ANEC) affects healthy children commonly following respiratory infections including influenza A infections. Treatment is largely symptomatic although high dose corticosteroids are recommended for severe cases.

Case report: We describe a 3-year old Malay boy who presented with status epilepticus following 3 days of fever, cough and rhinorrhea during the recent Influenza A H1N1 pandemic. Physical examination revealed a drowsy child with Glasgow coma scale of 12/15 with generalized hypertonia, hyperreflexia and bilateral upgoing plantar reflexes. He had neutrophilic leucocytosis (WCC 31,000, neutrophils 84%), liver impairment (ALT 3075 iu/L, AST 7632 iu/L) and renal impairment (creatinine 89umol/L). Throat swab for influenza A H1N1 PCR was positive. His brain computerized tomography scan showed ventriculomegaly, cerebral edema and bilateral thalamic hypodensities classical for ANEC. He was treated with oral oseltamivir, iv ceftriaxone, iv acyclovir, anticonvulsants, mannitol, inotropic agents and ventilation. On day 2 of admission, he was started on intravenous immunoglobulin (IVIg) 1g/kg/day x 2 days, and subsequently a further third dose given. He was extubated after 1 week and had signs of pseudobulbar palsy and mixed spastic dystonic quadriparesis with truncal ataxia. He was discharged home on day 20 on nasogastric feeding and unable to walk. At 2 weeks after discharge, he was able to walk aided unsteadily and was feeding by mouth. Six months after his illness, he showed almost complete recovery with minor neurological sequelae.

Conclusion: Influenza A H1N1 associated ANEC which normally has a poor prognosis may be successfully treated with IVIg.
POSTER PRESENTATIONS

P23

NON-ACCIDENTAL OR ACCIDENTAL HEAD INJURIES IN MALAYSIAN INFANTS: THE CLINICAL CLUES

Meera Thayalasingam¹, Irene Cheah Guat Sim², Abhi Veerakumarasivam¹

1. Faculty of Medicine & Health Sciences, University Putra Malaysia, Selangor
2. Institute of Paediatrics, Hospital Kuala Lumpur, Kuala Lumpur

Introduction: The search for a single reliable diagnostic symptom, sign or test that can absolutely confirm child abuse remains elusive. Understanding and identifying the differences between infants with non-accidental (NAHI) and accidental head injury (AHI) may help identify markers of abuse.

Objective: We aimed to distinguish the markers of abuse by comparing the socio-demographics, presenting complaints, clinical features and the extent of hospital investigation carried out in infants who sustained NAHI versus AHI.

Method: A retrospective review of all infants < 1 year of age with a diagnosis of head injury or abnormal head computed tomography scans at Hospital Kuala Lumpur between 2005 and 2006 was conducted.

Results: From a total of 81 children, 53.1% had a discharge diagnosis of NAHI. Infants with NAHI were more likely to be symptomatic, under the supervision of caretakers, and presented with no accurate mechanism of injury compared to infants with AHI. Retinal and subdural haemorrhages were more common in the NAHI group. However, 16 cases that were diagnosed with accidental subdural haemorrhage did not undergo ophthalmology examination or skeletal survey.

Conclusion: The history, the mechanism of injury, the presenting signs and symptoms as well as the nature of the injuries sustained are all valuable clues as to whether a head injury sustained during infancy is likely to be accidental or not. This review reinforces the need for an interdisciplinary and comprehensive assessment on any infant who has sustained any kind of head injury and this includes a referral to the child protection team.
Objective: This study evaluated the management of patients with cerebral palsy receiving care in the Paediatric Clinic, Hospital Tuanku Ja’far Seremban. The standard of care was based on the clinical practice guidelines by the Royal Children’s Hospital Melbourne, Australia. The standard is 80% adherence to the management aspect of the guidelines and 100% for type, classification and cause of cerebral palsy.

Methodology: All patients attending the Cerebral Palsy Clinic from February to July 2010 were included into the study. Patients with other causes of developmental delay without disorders of motor and tone were excluded. The documentation in the Paediatric Clinic medical records was then evaluated using a checklist and data processed using SPSS 11.0 programme. A focus group discussion was done to discuss remedial measures to be undertaken.

Results: There were 87 patients included in this study. The documentation for the type of cerebral palsy and classification was 100%. There were 86 patients (99%) had a cause or were investigated for the cause of cerebral palsy. In terms of the management of the associated disabilities, guidelines were adhered to in only 27 (31%) patients. Remedial measures suggested during the focus group discussion were having a proper guideline for management, having a checklist for guidance, continuous medical education and post clinic discussions.

Conclusion: There is still room for improvement in the management of cerebral palsy patients in our clinic and remedial measures will be undertaken to ensure acceptable quality of management for this group of patients.
POSTER PRESENTATIONS

P25

IMMATURE BRAIN TERATOMA IN AN INFANT IN HOSPITAL
UNIVERSITI SAINS MALAYSIA: REPORT OF A CASE

Norsarwany M1, Ariffin N1, Mohd Iqbal I1, Hasnan J2, Rohaizan Y3, Abdul Rahman G4, Stewart K5

1. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia
2. Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia
3. Department of Radiology, School of Medical Sciences, Universiti Sains Malaysia
4. Department of Neurosurgery, School of Medical Sciences, Universiti Sains Malaysia
5. Paediatric Oncology, Children Westmead Hospital

Introduction: The head is an uncommon site for teratomatous tumours. This is a report of an infant with an immature teratoma of the brain.

Case report: We reported a 3 months old child presented with sign of increase intracranial pressure. Patient underwent cranietomy with subtotal removal of the tumour. The resection of the tumour was incomplete due to it’s critical site in the brain. A pathological study revealed an Intracranial Immature Teratoma Grade 3. An alpha-fetoprotein was elevated. There was a dilemma of commencing adjuvant chemotherapy due to a very young age. ICE courses were delayed till 7 months of age. Patient completed 4 courses of ICE and remained in remission after more than 3 years follow-up.
POSTER PRESENTATIONS

P26

NETHERTON SYNDROME: A CASE REPORT

Suhaila Omar, Muzhirah Aisha Md Haniffa

Department of Paediatrics, Hospital Selayang, Selangor

Introduction: Netherton Syndrome is a rare hereditary disorder which presents with generalized erythroderma at birth or soon after. The objective of this case report was to highlight the importance of the other associated comorbidities that are attributed to this rare medical entity. Failure to recognize this may result in severe failure to thrive and high postnatal mortality.

Case report: We present the case of a boy, born borderline premature at 2.87 kg to non consanguineous parents who, upon birth was noted to have generalized erythrodermic skin lesions. Histopathology of skin revealed classical findings of Netherton Syndrome and ultrastructural analysis of hair showed trichorrhexis invaginata. He had severe failure to thrive in the first 6 months of life that was attributed to chronic diarrhea and recurrent infections and hence admitted during this time for weight monitoring and optimization of calorie intake. He had recurrent hypoglycaemia while in the ward and was not able to tolerate special diet prescribed by a Paediatric Gastroenterologist. Intravenous line cannulation was extremely difficult and attempts for central line was unsuccessful. Parents were counseled repeatedly and many family conferences were held in regards to patient’s condition and progress. Parents however decided to bring him home against medical advise. Patient succumbed to death soon after. In summary, children with ichthyosis and growth failure may have uncompensated caloric needs because of an impaired skin barrier (unpublished data). We would like to highlight the importance of recognition of growth failure and malnutrition in these children as well as prompt treatment of any infections.
POSTER PRESENTATIONS

P27

CHRONIC IMMUNE THROMBOCYTOPENIC PURPURA IN CHILDHOOD: A CLINICAL PROFILE

Abdul-Wahab1, Nargis M1, Aye Aye1, Haymond P2, Naznin M3

1. Paediatric Department, Kulliyyah of Medicine, IIUM, Kuantan, Pahang
2. Paediatric Department, HoSHAS, Temerloh, Pahang
3. Basic Medical Sciences Department, Kulliyyah of Medicine, IIUM, Kuantan, Pahang Malaysia

Objective: The aim of this study was to examine the clinical and laboratory profiles of chronic immune thrombocytopenic purpura (ITP) patients attending Paediatric Clinic of Tengku Ampuan Afzan Hospital, Kuantan and Sultan Haji Ahmad Shah Hospital (HoSHAS), Temerloh.

Method: All chronic ITP patients on follow-up in the year 2009 were identified. Information was collected from the case-record using a predesigned questionnaire.

Results: A total of 15 patients were identified. The age at which the patients first presented ranged from 2.3 – 10.5 years. Two patients were asymptomatic at presentation. Almost half of the patients had history of prior infection. In all except for 2 patients the platelet count was below 30 x 10⁹/L. Screening for connective tissue disease was positive on follow-up in one patient and none had concurrent autoimmune haemolytic anaemia. Eight patients had bone marrow examinations performed. Intravenous immunoglobulin and / or oral corticosteroid was started in all but two patients. There were no episodes life-threatening bleed. Duration of follow-up ranged from 1.5 – 9 years. One third had sustained normalization of the platelet count.

Conclusion: Although no episodes of life threatening bleed were documented in this small review, the daily functions and quality of life of these patients need to be looked into as some patients continued to require treatment to increase or sustain the platelet count.
POSTER PRESENTATIONS

P28

VEIN OF GALEN MALFORMATION IN NEWBORN WITH HEART FAILURE

Norazah Zahari, Lim Chin Theam

Department of Paediatric, University Malaya Medical Centre, Kuala Lumpur

Introduction: Extracardiac arteriovenous malformations are an uncommon cause of severe cardiac failure in infancy. Aneurysm of the vein of Galen may lead to diagnostic confusion because the presenting signs point to cardiac cause as a cause of cardiac failure. A neonate with such a condition is reported here.

Case description: A newborn boy with birth weight of 3.4 kg was limp and cyanosed at birth. Despite maximum positive pressure ventilation the oxygen saturation ranged from 75 to 78%. Physical examination revealed swelling of the neck bilaterally, which is soft and expansile. Loud bruit was detected on the head and neck, and a loud systolic murmur all over the precordium. There were gross cardiomegaly and hepatomegaly. The initial blood gas showed severe respiratory and metabolic acidosis and poor oxygenation. Echocardiography demonstrated normal cardiac anatomy, and a patent ductus arteriosus. The brachiocephalic artery and left common carotid artery were dilated. Cranial ultrasound confirmed the presence of aneurysm of vein of Galen. Despite optimal support the infant succumbed to the condition before embolisation could be performed.

Conclusion: Clinical diagnosis of cerebral arteriovenous fistula in newborn maybe difficult. In an infant with unexplained heart failure extracardiac arteriovenous malformation must be considered.
RIGHT ADRENAL PHAEOCHROMOCYTOMA IN A BOY WITH HYPERTENSIVE CRISIS INITIALLY SUSPECTED TO BE DUE TO AN INTRACRANIAL LESION

Nurshad S1, Karmila A1, Lucy C2, Ramanujam T3, Lucy LCS1, Fatimah H1, Jalaludin MY1

1. Department of Paediatrics, Faculty of Medicine, University Malaya, Kuala Lumpur
2. Department of Anaesthesiology, Faculty of Medicine, University Malaya, Kuala Lumpur
3. Department of Surgery, Faculty of Medicine, University Malaya, Kuala Lumpur

Introduction: Hypertension is uncommon in children. It is even rare for children to present with hypertensive crisis due to excessive adrenergic stimulation. We report a case of hypertensive crisis secondary to right adrenal phaeochromocytoma and its management.

Case report: A 14-year old Malay boy presented with worsening symptoms of hot flushes, sweating and palpitation for 3 years with 2 syncopal attacks 3 months prior to presentation. He was hypertensive (BP 180/100 mmHg) and tachycardic (110-120/min), with blurring of vision. Bilateral papilloedema was noted. Right subarachnoid cyst was found on CT brain and later decompressed. However, hypertension persisted and was difficult to control. Further investigations showed marked elevation of urine normetanephrine (91.15 umol/L). Right suprarenal mass (4.1 x 4.3 x 4.8cm) was found on CT scan. MIBG (methyl- iodobenzylguanidine) scan was positive. Short (prazosin) and long-acting (phenoxybenzamine) alpha blockers successfully controlled the hypertension. Normal saline infusion was given for five days prior to operation to sustain his intravascular volume. Intra-operatively he had a brief hypotensive episode which responded to noradrenaline infusion. He developed heart failure secondary to fluid overload 48 hours post-operation and was successfully treated with diuretics. His blood pressure normalised five days post-operation. Histopathological report confirmed malignant phaeochromocytoma of right adrenal. A repeat 24-hour urine test showed normal levels of catecholamines and its metabolites.
POSTER PRESENTATIONS

P30

INFLUENZA TYPE B VIRUS INFECTION ASSOCIATED ACUTE NECROTIZING ENCEPHALOPATHY OF CHILDHOOD

Putri Y¹, Nur Hafiza N², Norashidah AW², Norliah O¹, Hamid MZA¹

1. Department of Pediatric, Faculty of Medicine and Health Sciences
2. University Putra Malaysia, Selangor Department of Pediatrics, Hospital Serdang, Selangor

Introduction: Acute necrotizing encephalopathy of childhood or ANEC is clinical neurological entities that predominant affect young children in Far Eastern countries mainly in Taiwan and Japan. The clinical presentation consists of a rapidly deteriorating encephalopathy in a previously healthy child after an acute episode of upper respiratory illness and their prognosis is usually poor. The characteristic findings of this novel entity are multifocal brain lesions symmetrically distributed in the thalami, brainstem, tegmentum and cerebellar medulla, demonstrated by CT scan or MRI. The pathogenesis and the etiology of ANEC remain unclear and there are no specific treatments or prevention in this disease.

Case report: We report an uncommon case of Influenza type B virus infection in a 7-year-old Malay boy who presenting as a acute necrotizing encephalopathy of childhood. The child also had upper GI bleeding and hematological features of disseminated intravascular coagulation. He was treated with steroids and he made a full neurological recovery. We also review the current aspect of treatment of acute necrotizing encephalopathy of childhood.
TWO PREGNANCIES RESULTING IN 2 DAUGHTERS WITH ABERRANT CHROMOSOME X IN A MOTHER WITH VARIANT TURNER SYNDROME

Sathyabama R¹, Keng WT¹, Vigneswari G², Roziana A²

1. Department of Paediatrics, Penang Hospital, Penang
2. Department of Genetics, Hospital Kuala Lumpur, Kuala Lumpur

Case reports: We report 2 sisters who presented with learning difficulties. NSQ, 12 years old, had feeding problems at birth, history of frequent febrile seizures from 10 months till 2 years, global developmental delay, behavioral problems and dysmorphic features [full cheeks, right auricular pit, tapering long digits, mild hypertelorism and lateral deviation of toes]. Her IQ assessment result was 80, demonstrating both receptive and expressive language disorder. Her younger sister, 9 year old NSW had learning difficulty with IQ of 83, mild hyperactivity and subtle dysmorphic features [mild clinodactyly, short stature and lateral deviation of third toe]. Chromosomal analysis of both sisters revealed 46XX,add(X). Their mother’s chromosomal analysis showed 45 X, (13) /46 X, add (X) (17), which was essentially consistent with variant Turner Syndrome. Mother has premature menopause but no learning problems or dysmorphism. The abnormal chromosome X in both sisters was thus inherited from their mother. Further analysis of the abnormal chromosome X showed that the additional material was duplicated Chromosome X. Fertility among women with Turner syndrome have been reported especially among those with 45,X/46,XX mosaicism. This family was very unusual in that 2 spontaneous pregnancies in a mother who has 45X/46 X,add(X) ended with both children bearing abnormal chromosome X.
Objective: The objective of this study was to describe the epidemiologic features of child abuse detected and reported in Sabah Women And Children’s Hospital (SWACH) in the first six months of year 2010.

Method: This retrospective study was conducted by obtaining data from the Department of Social Welfare in SWACH. The data collected were all children aged sixteen and younger suspected and confirmed as child abuse. The victims’ medical records were retrieved and a standardized form was used to collect demographic data, type of child abuse and the perpetrator description.

Results: There were 77 cases of child abuse from January to June 2010, about 142% increment as compared to the same duration the previous year, 2009 (31 cases). Sexual abuse was the highest (79.2%) followed by physical abuse (14.2%) and child neglect (6.5%). Majority of child sexual abuse perpetrators were mainly from persons known to the victims (95%) such as boyfriend and father. In child physical abuse cases, mothers were the main perpetrators (45.4%).

Conclusion: The incidence of child abuse is increasing. Sexual abuse constituted the majority and the main suspects were persons known to the victims.
A REPORT OF TWO LIFE-BORN INFANTS, HOMOZYGOUS FOR SOUTHEAST ASIAN OVALOCYTOSIS

Surini Yusoff, Narazah Mohd Yusoff, Keng Wee Teik, Chng Gaik Siew, Winnie Ong Pei Tee, Yeoh Seoh Leng, Nur Hasnah Ma’amor, Masafumi Matsuo, Hans Van Rostenberghe

1. Department of Paediatrics, Universiti Sains Malaysia
2. Advanced Medical and Dental Institute, Universiti Sains Malaysia
3. Department of Genetics, Hospital Kuala Lumpur, Kuala Lumpur
4. Department of Paediatrics, Hospital Pulau Pinang
5. Human Genome Centre, Universiti Sains Malaysia, Malaysia
6. Department of Paediatrics, Kobe Graduate School of Medicine, Kobe, Japan

Introduction: Homozygosity of Southeast Asian Ovalocytosis (SAO) has been thought to cause death in utero. Life births have not been reported so far. We report two cases of live-born infants with homozygous codon 400-408 deletion of the erythrocyte band 3 gene.

Case presentations: Probands 1 and 2 were boys, born to mothers aged 25 and 35 years respectively via emergency lower segment Caesarean section for foetal distress. Proband 1 was born with hydrops fetalis and presented with pallor, oedema, hepatosplenomegaly and a globular heart on chest X-ray. There was severe anaemia (Hb 4.9 g/dL) at birth with haemolytic picture on the blood film. The infant died at 7 hours of life. Proband 2 was born to a mother who had previously three spontaneous abortions. This case was diagnosed antenatally to have severe anaemia and received 2 antenatal transfusions. He was ventilated after birth for respiratory distress. Haemolytic anaemia complicated by pathological jaundice was ongoing and he required exchange transfusion at day 2 and a packed transfusion at day 5 of life. The infant was clinically bronzed with large hepatomegaly (4-5cm) and palpable spleen (1cm). The DNA analysis revealed that both probands were homozygous for the codon 400-408 deletion in band 3 gene and the parents were heterozygous.

Conclusion: This is the first report of two live-born infants with homozygosity of the band 3 gene (SAO). One of the children received antenatal treatment for foetal anaemia and managed to survive up to at least two months of age.
POSTER PRESENTATIONS

P34

CHRONIC PAIN SYNDROMES: A MISSED OR MISDIAGNOSIS?

Tang Swee Ping

Paediatric Rheumatology Unit, Department of Paediatrics, Hospital Selayang, Selangor

Introduction: Chronic pain syndromes (CPS) are common amongst children but often unrecognised and misdiagnosed resulting in inappropriate investigations and management with unnecessary exposure to potentially toxic drugs. A case series of 3 children is described highlighting this issue.

Case report 1: 13-year old Indian girl who developed generalized muscle, back and joint pains with stiffness soon after a diagnosis of diabetes mellitus. She had non restorative sleep, poor appetite, dizziness, school absenteeism and needed help for activities of daily living (ADL). She was previously diagnosed as reactive arthritis, spondyloarthropathy and possible systemic lupus erythematosus, and treated with various NSAIDs.

Case report 2: 10-year old Chinese boy with progressive severe bilateral ankle, knee, arm and back pains with stiffness and inability to ambulate in the mornings. He was diagnosed as acute rheumatic fever and later possible arthritis, and given aspirin, prednisolone and etoricoxib with no improvement.

Case report 3: 13-year old Malay boy who developed finger joint pains following a viral infection. This progressively worsened over months to generalized joint and muscle pains with stiffness. He was wheelchair bound, dependent for his ADLs, had sleep disruption and school absenteeism. He was diagnosed as juvenile idiopathic arthritis and treated with several NSAIDs, high dose prednisolone and even 2 months of Methotrexate.

All 3 patients subsequently received appropriate pain management upon diagnosis of CPS, and Case 2 and 3 have done very well whilst Case 1 still has intermittent pain flares.

Conclusion: Prompt recognition of CPS is important to avoid unnecessary investigations and treatment, and to ensure timely appropriate management for optimal outcome.
POSTER PRESENTATIONS

P35

INFLIXIMAB THERAPY IN SEVERE REFRACTORY KAWASAKI DISEASE

Tang Swee Ping¹, Hung Liang Choo²

1. Paediatric Rheumatology Unit, Department of Paediatrics, Hospital Selayang Selangor
2. Department of Paediatrics Institute of Paediatrics, Hospital Kuala Lumpur, Kuala Lumpur

Introduction: Kawasaki Disease (KD) is the second commonest vasculitis in childhood. Majority respond to a single dose of intravenous immunoglobulin (IVIg). Resistant cases may require a second dose of IVIg and/or intravenous methyl prednisolone (IV MTP).

Case report: We report a 4-month old Malay male infant with severe refractory KD and bilateral coronary artery (CA) aneurysms who was successfully treated with iv infliximab, a chimeric anti-Tumour Necrosis Factor (TNF) monoclonal antibody, the first reported case in Malaysia. This infant initially presented with 9 days of fever with symptoms and signs typical of KD. He did not respond to 2 courses of IVIg (each 2g/kg) and 3 doses of IV MTP 30 mg/kg/day. He was referred to us for persistent fever with rashes. On day 31 of illness, he was still irritable, febrile with red lips, conjunctival injection, rashes and mild arthritis. He had leucocytosis, thrombocytosis, and raised inflammatory markers. He was given one dose of iv infliximab 5 mg/kg and within hours, his fever settled and was clinically better. His blood inflammatory markers post infliximab improved and an echocardiogram 3 days later showed dilatation of all CA with a thrombus at the right CA ostia. He also had evidence of transient asymptomatic myocardial ischaemia. He was treated with Aspirin, Clopidogrel and subcutaneous Enoxaparin. Follow-up echocardiogram a week later showed a resolving thrombus. At the last review 15 months after initial diagnosis, he remained well with regressing CA aneurysms.

Conclusion: Infliximab therapy may be an effective treatment option for patients with severe refractory Kawasaki disease.
POSTER PRESENTATIONS

P36

CLINICAL FEATURES AND PREDICTORS OF RENAL DISEASE IN JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS

Lim Sern Chin, Tang Swee Ping, Wan Jazilah Wan Ismail

Department of Paediatrics, Hospital Selayang, Selangor

Objective: To describe renal involvement in Juvenile Systemic Lupus Erythematosus (JSLE) at presentation and during the course of disease; and assess for clinical predictors of renal disease.

Methodology: A retrospective review of medical electronic records of all patients with JSLE seen at the paediatric rheumatology and nephrology units in Hospital Selayang from 2003-2009. Data collected included demographics, clinical and laboratory features.

Results: There were 66 patients with JSLE (62 females, 4 males), with mean age at presentation of 9 years (3.9 - 13.4 years) and mean follow-up of 31.4 months (1-75 months). Majority (53.0%) were Malays, 42.4% Chinese and 4.5% Indians. 52 patients (78.8%) had renal involvement which was present in 35(67.3%) at presentation (group A) and another 17(32.7%) during follow-up (group B). Up to 42.8% (15/35) of Group A and 52.9% (9/17) of group B went into renal remission with treatment. Only 14/52(26.9%) had renal biopsies and the commonest patterns were Class IV Lupus Nephritis (6 patients) followed by Class III and Class II (3 patients each). Hypoalbuminemia at presentation had significant correlation with renal disease in these patients (p<0.05, 95% confidence interval). However, there were no other identifiable clinical predictors for the development of subsequent renal disease or persistence of renal disease despite standard treatment.

Conclusion: Majority of JSLE patients had renal involvement either at presentation or subsequently in the course of disease but no clinical predictors for the development of lupus nephritis could be identified in our cohort unlike other adult studies.
POSTER PRESENTATIONS

P37

GAUCHER’S DISEASE TYPE 2: A CASE REPORT

Tengku Putri Zainab, Soo Min Hong, Ngu Lock Hock

Department of Paediatrics, Hospital Sungai Buloh, Selangor

**Introduction:** Gaucher's disease is the most common of the lysosomal storage diseases with an incidence of about 1 in 50,000 to 100,000 live births.

**Case presentation:** We report here a 7 month old girl presented with poor sucking and recurrent respiratory tract infections from birth associated with post tussive vomiting. She had limb rigidity and gross hepatosplenomegaly. She was later diagnosed with Gaucher’s Disease Type 2.

**Discussion:** To the best of our knowledge, there is no case report published for Gaucher’s Disease Type 2 in Malaysia.

**Conclusion:** We are adding a new case to stress the importance of early recognition by clinical manifestations of Gaucher’s Disease in Malaysia.
L-ASPARAGINASE INDUCED SAGITTAL SINUS THROMBOSIS IN CHILDHOOD ACUTE LYMPHOBLASTIC LEUKAEMIA: A CASE REPORT

Teoh SL, Kuan GL
Department of Paediatrics, Malacca Hospital, Malacca

Case report: A 3-year-old girl with acute lymphoblastic leukaemia (ALL) who was undergoing induction phase of chemotherapy that consist of dexamethasone, vincristine and L-asparaginase, developed acute left sided seizure and hemiparesis. A diagnosis of sagittal sinus thrombosis with right frontoparietal infarct was confirmed by magnetic resonance imaging (MRI). The thromboembolic event could not be attributed to leukaemic infiltration or leukostasis. Until this event, the girl had been treated already with 6 doses of L-asparaginase. We conclude that the thrombosis was likely L-asparaginase induced. She was commenced on low molecular weight heparin (LMWH). A computed tomography (CT) done 24 hours later due to recurrent seizure revealed haemorrhagic transformation within the infarct. The LMWH was, however, continued for a total of 6 weeks. She made full neurological recovery within 48 hours after presentation. A magnetic resonance venography (MRV) performed 10 weeks later showed complete resolution of the thrombus. The chemotherapy protocol was incessant with omission of L-asparaginase.
POSTER PRESENTATIONS

P39

POLYARTERITIS NODOSA – A RARE PRESENTATION

Tina Sivalal\textsuperscript{1}, Tang Swee Ping\textsuperscript{2}, Cham Weng Tarng\textsuperscript{2}, Soo Min Hong\textsuperscript{1}

1. Department of Paediatrics, Sungai Buloh Hospital, Selangor, Malaysia

2. Department of Paediatric Neurology, Selayang Hospital, Selangor, Malaysia

Introduction: Polyarteritis nodosa (PAN) is a vasculitis of medium-sized or small vessels usually involving medium-sized arteries of the skin, peripheral nerves, the gut, and the kidney vasculature. It is a disease of the older age group, and is uncommon in children.

Objective: To create awareness of a rare presentation of arthritis such as PAN.

Methods: A 3 year old girl was admitted with fever and non specific symptoms of cough, runny nose and vomiting for 5 days. She also had difficulty in walking for 2 days.

On examination, the only abnormalities were an antalgic gait, patchy, reddish pigmentation, 1-2 cm in diameter, over left foot. Her blood pressure was 113/65 mm Hg, and motor strength of 4/5 in both hamstring and quadriceps muscles, with no other abnormality.

Results and discussion: All preliminary investigation results were normal, she was provisionally diagnosed as an upper respiratory tract infection with infected impetigo, and treated with intravenous penicillin and gentamycin and discharged a few days later. On subsequent review, investigation results were ESR - 115, white blood count(WBC) more than 40,000, increasing platelet count, C-reactive protein – 15. The child had been unwell for more than 2 weeks, with persistent fever associated with lower limb pain but able to walk. She was treated with nifedipine, aspirin, prednisolone and ranitidine. The child’s symptoms are resolving and she is currently being tapered off prednisolone.

Conclusion: It is important to consider PAN in children with arthritis and a atypical rash.
A CASE REPORT OF PENTALOGY OF CANTRELL

Yeoh Chiou Yen, Sunita, Yong Lai Peng, Nachal RMN Nachiappan, Yogeswari Sithamparanathan

Department of Paediatrics, Hospital Tengku Ampuan Rahimah, Selangor

Case report: We report a case of Indonesian infant boy with a rare Pentalogy of Cantrell which consists of defects of the lower sternum, anterior diaphragm, midline supraumbilical abdominal wall and diaphragmatic pericardium with ectopia cordis. He first presented to HTAR on day 15 of life with supraventricular tachycardia and subsequent admission at 3 months of life with respiratory distress requiring prolonged ventilation. He had an omphalocele at birth and during second admission he was found to have a large central tendon defect of diaphragm with absence of pericardium and intrathoracic herniation of liver into mediastinum pushing the heart to the left. He had surgical repair of the congenital diaphragmatic hernia, abdominoplasty and right herniotomy done at 5 months of age. The Pentalogy of Cantrell must be actively sought in every patient with an omphalocele.
POSTER PRESENTATIONS

P41

DAPSONE SYNDROME – FIRST PAEDIATRIC CASE REPORT IN MALAYSIA

Z Zurina¹, O Elizawaty², S Thevarajah³, O Norlijah¹

1. Paediatric Department of Faculty of Medicine and Health Sciences, University Putra Malaysia
2. Paediatric Department, Hospital Serdang
3. Dermatology Department, Hospital Kuala Lumpur

Introduction: Dapsone syndrome is a potentially fatal hypersensitivity reaction characterised by high fever, skin rash, hepatitis, lymphadenopathy and haemolytic anaemia appearing several weeks to as late as six month following the use of dapsone.

Case Report: We report a 12-year-old girl who developed high grade fever associated with intensified jaundice, generalised erythematous maculopapular rash and hepatomegaly after five weeks of starting the multi-drug regimen (dapsone and clofazimine) for the treatment of Hansen's disease. Laboratory investigations revealed presence of anaemia, leucocytosis with eosinophilia, deranged liver enzymes and abnormal coagulation profile. Histological examination of the skin biopsy showed perivascular lymphocytic infiltrates in the dermal layer. Immediate cessation of the offending drug and administration of steroid was proven successful.

Conclusion: A high level of clinical awareness is important for early diagnosis of dapsone syndrome as initiation of a prompt treatment may lead to rapid recovery.
P42

BRONCHIAL ARTERY EMBOLISATION IN BRONCHIECTASIS WITH NEOVASCULARISATION AND HEMOPTYSIS

Teh Keng Hwang, Choong Pheik Sian

Department of Paediatrics, Hospital Sultanah Bahiyah, Kedah

Case report: We report a 9 year old patient with global developmental delay and bronchiectasis since 6 years ago who presented with massive hemoptysis and was ventilated on high PEEP. Bleeding continues in bouts and he needed blood transfusion and inotropes. CT angiography showed extensive neovascularisation involving the right upper lobe with the feeder from the Right subclavian and bronchial artery. Embolisation was performed on the 6 hospitalisation day and extubated 3 days later. Embolisation as a therapeutic option will be discussed in this patient with bronchiectasis and hemoptysis secondary to severe neovascularisation.
Objective: Childhood obesity has become a serious public health problem. The main objective of this study is to determine the prevalence and risk factors associated with childhood obesity among 7 and 8 year old primary school students in Kota Kinabalu. The secondary objective is to determine the prevalence of high blood pressure among primary school students who are obese.

Methodology: A total of 891 students were included from 5 zones of Kota Kinabalu. This study was conducted between February to April 2008. Questionnaires were used for data collection. Weight and height were measured and BMI (body mass index) was calculated. Blood pressure measurements were taken at least twice.

Results: The overall prevalence of obesity among 7 and 8 year old primary school students in Kota Kinabalu was found to be 2.5%. Low birth weight (birth weight less than 2.5kg), high birth weight (birth weight 4.0kg and above) and high family income (total family income per month more than RM 5000) were not associated with obesity in this study. No breast feeding and maternal gestational diabetes increased risk of obesity, but were not statistically significant. Non exclusive breast feeding (infant formula started before 6 month old) was associated with doubling the risk of obesity (odds ratio=2.4, 95% confident interval= 1.02 to 5.63, p=0.038) and this was statistically significant. Obese students were associated with 7 times increased risk of hypertension (odds ratio=6.62, 95% confident interval= 2.8 to 15.63) and p<0.05.