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YIA1

COMPARISON OF EFFECTIVENESS AND SAFETY OF KETAMINE AGAINST KETAMINE WITH MIDAZOLAM IN PROCEDURAL SEDATION FOR LUMBAR PUNCTURE IN PAEDIATRIC LEUKAEMIA PATIENTS

Ahmed Faisal¹, Mohd Suhaimi bin Ab Wahab², Norsarwany Mohamad¹, Saedah Ali³

1. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
2. Department of Pharmacology, School of Medical Sciences, Universiti Sains Malaysia
3. Department of Anaesthesiology, Hospital Universiti Sains Malaysia, Kelantan, Malaysia

Children with leukaemia undergo several invasive procedures. Sedation is used to make these procedures less painful as it is necessary for successful outcome. However sedatives can have devastating effects. In our centre as well as others, combination of ketamine with midazolam has been used for years without specific protocol. A double blinded crossover clinical trial in 29 leukaemia patients was conducted to compare effectiveness and safety of ketamine 2 mg/kg (K2) against combination of ketamine 1 mg/kg with midazolam 0.1 mg/kg (KM) for lumbar puncture (LP). Twenty seven patients (93 percent) were successfully sedated in both groups K2 and KM. Mean time taken for sedation (7.56 minutes), and mean time to be fully conscious after sedation (132 minutes) were significantly less in K2 (P<0.05). Though statistically not significant (McNemer Test 0.250) adverse effects were more common in K2 and could be clinically relevant. In both K2 and KM difference in mean time to complete LP were not statistically significant. All the adverse effects were easily reversible. Even though individual response varies widely Ketamine could be used safely and effectively as a single agent for sedation in lumbar puncture in a controlled environment.
YOUNG INVESTIGATOR AWARD PAPERS

YIA2

BLOOD STREAM INFECTIONS (BSI) IN HAEMATOPOIETIC STEM CELL TRANSPLANT (HSCT) RECIPIENTS: A 20-YEAR REVIEW

Ngian Geok Hoon\textsuperscript{1}, Chan Lee Lee\textsuperscript{2}

1. Department of Paediatrics, Hospital Pakar Sultanah Fatimah, Muar
2. Department of Paediatrics, University of Malaya Medical Centre (UMMC), Kuala Lumpur, Malaysia

\textbf{Background:} BSIs are common in HSCT with crude mortality rate of 50\%. Multiple factors determine the rate and outcome of BSI. A variety of microbiological agents predominate as causative organisms for BSI depending on the different phases of transplantation.

\textbf{Objectives:} 1. To determine the rate of blood stream infections (BSI) and the causative organisms in paediatric HSCT recipients in the first 100 days post-transplant. 2. To identify the risk factors for the development of BSI in HSCT recipients.

\textbf{Design:} Retrospective cohort study. The inception cohort is from 1st March 1987 to 28th Feb 2007, of all paediatrics HSCT recipients in UMMC.

\textbf{Result:} Of 321 HSCT during the study period, 128 (40\%) transplants had a total of 181 episodes of BSI. There were a total of 200 isolates, 50\% were Gram positive organisms, 40\% were Gram negative rods, 6\% were fungi and 4\% viral and parasitic infections. Coagulase negative staphylococcus (CONS) were the most common isolates (29.5\%), followed by S. aureus (9.5\%). Prolonged neutropenia of more than 30 days and presence of infection within 1 week prior to conditioning till day of HSCT were significant predictors of BSI, with hazard ratio of 2.3 each.

\textbf{Conclusion:} This study confirmed a high BSI rate in paediatric HSCT recipients. CONS was the most common isolate.
PAEDIATRIC MELIOIDOSIS: SYSTEMIC VERSUS LOCALIZED

Wong Ke Juin, Fong Siew Moy, Soo Thian Lian

Paediatric Department, Sabah Woman and Children Hospital, Kota Kinabalu, Sabah, Malaysia

Objective: To determine the demographic data, clinical presentation and outcome of paediatric melioidosis in Sabah, Malaysia.

Method: A retrospective study of culture-positive cases of melioidosis in children age less than 15 years admitted to Hospital Likas from 2003 to 2010. Patients’ case records were retrieved, analyzed and recorded on standardized forms.

Results: There were total of 24 cases of melioidosis during the 7 years period and 23 cases notes were traced. Thirteen patients were male. The mean age of presentation was 7 years (3 weeks to 14 years). All but one patient had fever on presentation, followed by 11 patients had arthralgia, arthritis and soft tissue abscess respectively. Majority of them were Kadazan Dusun, followed by Bajau. Sixteen patients (70%) had medical comorbidity with thalassaemia being the commonest underlying medical condition. Seventeen patients (74%) had systemic melioidosis and 6 patients had localized melioidosis. In systemic group, 88% had pneumonia, 47% needed ventilation and 65% required inotrops. Mortality rate was 70% among the systemic melioidosis and no relapse was documented in these patients. In localized group, all of them had soft tissue abscess and joint involvement but only one patient had pneumonia. None of the patients required intubation or inotrops. Survival was 100% in the localized group but 2 patients (33%) had relapse of melioidosis. The overall case fatality rate was 52.2%.

Conclusion: Majority of melioidosis in our series had medical comorbidity with thalassaemia major being the commonest underlying medical condition. Systemic melioidosis had high mortality rate compared to localized melioidosis.
INVASIVE PNEUMOCOCCAL DISEASE IN PAEDIATRIC INTENSIVE CARE UNIT, HOSPITAL SULTANAH AMINAH, JOHOR BAHRU

Pazlida P, Mat Bah MN
Paediatric Department, Hospital Sultanah Aminah, Johor Bahru, Malaysia

Objectives: To review the epidemiology, morbidity and mortality rate, and antibiotic susceptibility in patients with invasive pneumococcal disease admitted to our Paediatric Intensive Care Unit (PICU).

Methodology: Retrospective review of our PICU database and patients’ record from January 2008 till March 2011 for invasive pneumococcal.

Results: Fifteen patients with invasive pneumococcal disease were identified, they accounted as 1% of total patients admitted to PICU during the study period. None of them received any pneumococcal vaccine and only one patient (6.7%) had underlying chronic illness. Of these 15 patients, 60% were less than 2 year old, 86.7% were Malay and male to female ratio were 2:1. Six presented with meningitis, 6 with pneumonia and 3 as bacteremia. The morbidity rate was 86.7% with septicaemic shock as the most common morbidity (77%). Penicillin resistance was noted in 26.7% and none showed resistance towards Ceftriaxone and Cefotaxime. There were 7 (46.7%) death with highest mortality rate in meningitis (5 patients) followed by bacteremia (2 patients). Of these 7 deaths, 42.9% died within 48 hours of admission and 4 were Penicillin sensitive Streptococcus pneumonia.

Conclusion: Invasive pneumococcal disease in children has significant morbidity and mortality. Targeting vaccination only on high risk group children would be futile since majority of the affected child were healthy without underlying illness. Hence, inclusion of pneumococcal vaccine inside the future national immunization program should be considered.
YOUNG INVESTIGATOR AWARD PAPERS

YIA5

RISK FACTORS ASSOCIATED WITH THE EMOTIONAL STATES OF PARENTS ADMITTED TO THE PAEDIATRIC INTENSIVE CARE UNIT

Noor Zehan Rahim¹, Maniam Thambu², Tang Swee Fong¹

1. Department of Paediatrics, Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur, Malaysia
2. Department of Psychiatry, Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur, Malaysia

Objective: To determine risk factors associated with the emotional states of parents whose children were admitted to a Paediatric Intensive Care Unit (PICU).

Method: This was a prospective questionnaire study of parents whose children were admitted to the PICU. The emotional states of parents were assessed using the Depression, Anxiety and Stress Scale 21 (DASS 21).

Results: A total of 132 parents participated. Twenty percent of fathers and 29% of mothers experienced depression and a third of both fathers and mothers experienced moderate to extreme anxiety and stress. There was no significant difference in the occurrence of the different emotional states between mothers and fathers. There was also no significant association between depression and anxiety with age, ethnicity, employment status, education level, type of admission and readmissions. Higher educated parents experienced higher stress levels.

Conclusion: This study shows that a significant proportion of parents not only experience stress and anxiety but also depression when their child is admitted to the intensive care unit. Mothers and fathers have similar emotional states and higher educated level parents experience more stress.
YOUNG INVESTIGATOR AWARD PAPERS

YIA6

PLASMA LEAD LEVEL AMONG PAEDIATRIC THALASSAEMIA PATIENTS IN KOTA BHARU, KELANTAN

Mohd Iqbal I1, M Suhaimi A W2, Norsarwany M1, Mariani M1

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

Introduction: The route of human lead exposure is primarily via inhalation or ingestions. However, Bearer et al has shown that blood transfusion can be a source for lead exposure to premature infants. As majority of Thalassemic patients are transfusion dependant, thus hypothesis been made that regular blood transfusion can represents a significant source of lead.

Objective: To determine plasma lead level in Thalassaemic patients with regular blood transfusion.

Method: This was a cross sectional study, conducted at Hospital Universiti Sains Malaysia (HUSM) from March 2009 until August 2010. A total of 90 patients were included, 45 were Thalassaemic transfusion dependant patients and the other 45 were age and sex matched. The blood lead samples were taken pre-transfusion for Thalassaemic patients group and at random for control group. Samples then analyzed using standard Atomic Absorption Spectrometer.

Results: This study revealed that overall mean plasma lead level was 2.13 ± 1.72µg/dL. Lead levels in Thalassaemic patients group (1.14 ± 0.85µg/dL) were significantly lower than the control group (3.12 ± 1.81µg/dL). No correlations of age or serum ferritin level to blood lead level were found.

Discussion: Surprisingly Thalassaemic showed lower lead levels than controls. This could be explained by the presence of high serum iron in Thalassemics which will reduce lead absorption and lead uptake of by erythrocytes.

Conclusion: This study shows that our Thalassaemic transfusion dependant patients do not have extra exposure to lead from their regular blood transfusion. Lower blood lead level in Thalassemic patients requires further study.
ORAL PRESENTATIONS 1

OP1

PREVENTION OF CHILDHOOD DROWNING IN MALAYSIA

Amar-Singh HSS1,2, Pui-San Tan2, Lina Hashim2

1. Paediatric Department, Raja Permaisuri Bainun Hospital, Malaysia
2. Clinical Research Centre Perak, Malaysia

Introduction: Injuries are a major cause of death in children aged 1-18 years. Drowning deaths are an area that has received insufficient attention as an important cause of mortality locally. The World Health Organization commissioned this study with the support of Ministry of Health Malaysia. The objective was to “To collaborate data on childhood drowning in Malaysia and review existing drowning prevention measures in place”.

Methodology: This was a cross-sectional study using secondary data obtained from various sources on drowning events. Sources included Government: Ministry of Health, Ministry of Education, Ministry of Sport, Police, Policymakers, Ministry of Transport, Department of Statistics and NGOs (Life Saving Society Malaysia, newspaper sources, Fire & Rescue Volunteers Malaysia). Drowning deaths data were collected from Year 2000-2007. Data obtained were then analysed to provide understanding of the epidemiology of drowning incidents in Malaysia, risk factors contributing to it and preventive measures that need to be drawn up or revision of current preventive measures in addressing this problem. Drowning outcome was divided into three categories: Death, Morbidity, No morbidity.

Results: Notified fatalities due to drowning amounted to between 600-700 per year for the year 2000-2007. Of these between 250-300 (44.5%) were children under 18 years. An additional 200 children drowned but survived. Hence the burden of drowning in children is approximately 500 yearly. The national average drowning fatality rate in children was 3.05 per 100,000 children over the 8 year period. The national average drowning rate (fatal and non-fatal) in children was 5.0 per 100,000 children. An average of 26.3% of childhood drowning deaths were non-medical certified. There was no reduction in drowning fatalities from 2000 to 2007. Most drowning took place in east coast regions (Terengganu and Kelantan) from November-March. It was 3 to 4 times more common in boys than girls and was most prevalent in the 10 to 14 years age group. Most prevalent sites of all-age drowning were seas and rivers. Limited water safety regulations or legislation are currently available in the country. The Life Saving Society Malaysia has various water safety courses for the public, but is not widely implemented to the whole nation as yet. Although between 250-300 children die each year due to drowning (3 per 100,000 children), an approximately additional 200 or more drowned but survived (approximately 2 per 100,000 children). Hence the real burden of drowning in children is approximately 500 yearly (5 per 100,000 children).

Discussion: The average Malaysian childhood drowning fatality rate was below the global rate as well as the Western Pacific Region LMIC rate but this may still be due to underreporting. The Malaysia's rate was closer to the HIC region's rate. This report was based on reported sources, hence it may underestimate overall drowning rates. Very limited data was available on the drowning and mechanisms of events are not available. It was only possible to capture snapshots of detailed drowning information from local newspaper archives. Some government agencies did not respond to requests for information or data despite numerous requests in various forms. This however is the first comprehensive national study in Malaysia on drowning in children and serves to highlight the magnitude of the problem.

Recommendations:
1. Finding of this study need to be presented to senior manager in Health, Safety and other relevant agencies to plan intervention strategies.
2. Finding of this study need to be communicated to the general public to sensitise the general public. Health education material produced by the MOH need to incorporate clear messages on drowning safety.
3. Establish a national drowning database or registry to facilitate accuracy of rates as well assist in monitoring the effect of any interventions.
Objective: To assess the effect of pacifier use versus no pacifier use in healthy full-term newborns whose mothers have initiated breastfeeding and intend to exclusively breastfeed, on the duration of breastfeeding, other breastfeeding outcomes and infant health.

Method: We searched the Cochrane Pregnancy and Childbirth Group's Trials Register (31.12.2010) for randomised and quasi-randomised controlled trials comparing pacifier use versus no pacifier use in healthy full-term newborns who have initiated breastfeeding. Two authors independently assessed the studies for inclusion, assessed risk of bias and carried out data extraction. We used relative risks (RR) and 95% confidence intervals (95%CI).

Results: We included 2 trials (involving 1302 infants). Meta-analysis of the two combined studies showed that pacifier use had no significant effect on the proportion of infants exclusively breastfed at 3 months RR 1.00; 95%CI (0.95-1.06), and at 4 months of age RR 0.99; 95%CI (0.92-1.06), on partially breastfed at 3 months RR 1.00; (95%CI 0.97-1.02), or at 4 months of age RR 1.01; 95%CI (0.98-1.03).

Conclusion: Pacifier use in healthy term breastfeeding infants started from birth or after lactation is established, did not significantly affect the prevalence or duration of exclusive and partial breastfeeding up to 4 months of age. Evidence to assess the short-term breastfeeding difficulties faced by mothers and long-term effect of pacifiers on infants' health is lacking.
LESS CONVENTIONAL USE OF INTRAVENOUS METHYLPREDNISOLONE IN CHILD NEUROLOGY

Nor Azni Yahaya, Muslim Muhammad

Department of Paediatrics, Hospital Raja Perempuan Zainab II, Kota Bharu, Malaysia

Objective: High dose steroid in the form of intravenous methylprednisolone (IVMP) is an established treatment in child neurology, and is the mainstay treatment in some inflammatory, autoimmune and demyelinating diseases such as acute disseminated encephalomyelitis (ADEM), transverse myelitis and cerebral lupus vasculitis. However, its uses in other similar conditions are rather limited, with clinicians preferring other immunomodulatory modalities such as oral steroids and immunoglobulin. We aim to share our experience in treating a number of neurological conditions using IVMP in children whereby IVMP itself is not the usual choice of treatment in practice, although its usefulness in those conditions has been described in literatures.

Method: We present a case series consisting of different neurological diagnoses in which the instituted treatment included the use of IVMP either as a first choice or add-on therapy.

Results: Five cases were described; a 10 year-old child with severe rheumatic chorea, an 8 year-old girl with Guillain-Barre syndrome (GBS), a 5 year-old girl with protracted post varicella cerebellitis, and 2 infants with infantile spasms due to lissencephaly. In all these cases, varying degrees but significant clinical improvements were observed following the treatment.

Conclusion: In cases that we described, in which the pathogenesis was postulated to involve autoimmune or inflammatory mechanisms, as well as in severe symptomatic epilepsies in young infants, IVMP therapy could be beneficial, especially when an aggressive mode of treatment is preferred.
ORAL PRESENTATIONS 1

OP4

MIDTERM OUTCOME OF VENTRICULAR SEPTAL DEFECT CLOSURE USING PFM LE VSD NIT OCCLUDER DEVICE IN HUSM

Mohd Rizal Mohd Zain, Wan Pauzi Ibrahim, Mohd Nikman Ahmad, Saedah Ali, Raja Salihatunnisa Ibrahim, Noormin Hida Adenand, Abdul Rahim Wong

1. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
2. Anaesthesiology Department, Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia
3. Paediatric Department, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia

Objective: To analyse safety, efficacy and early and mid-term follow-up results of percutaneous closure of VSD using Pfm Le Vsd nit occluder device.

Background: The previous percutaneous closure of VSD using other occluder device have been reported before, however it was complicated by complete AV block. However the data on the Pfm Le Vsd nit occluder device are still limited.

Methods: Between January 2008 till April 2011, 41 patients underwent percutaneous closure of VSD at our center. All the patients were using Pfm Le Vsd nit occluder device for the closure.

Results: The mean age at closure was 13.2 ± 4.2-year-old (range 6 to 22-year-old). The attempt to place the device was successful in 36 patients. Total occlusion rate was 42% at completion of procedure and rising to 73% at 18 months of follow-up. No major complications except for one in which the embolisation of the device requiring removal of the device via surgical exploration. No case of AV block noted and no worsening of AR.

Conclusions: Percutaneous closure of VSD using Pfm Le Vsd nit occluder device can be performed safely and successfully and can be a good alternative to surgical closure and previous VSD device.
SEPARATE CARE FOR NEW MOTHER AND INFANT VERSUS ROOMING-IN FOR INCREASING THE DURATION OF BREASTFEEDING: A COCHRANE REVIEW

Sharifah Halimah Jaafar¹, Kim Seng Lee², Jacqueline J Ho³

1. Ipoh Specialist Hospital, Malaysia
2. Fatimah Hospital, Ipoh, Malaysia
3. Penang Medical College, Malaysia

Separate care for new mother and infant may affect the duration of breastfeeding, breastfeeding behaviour and may have adverse effect on neonatal and maternal outcomes.

**Objective:** To assess the effect of mother-infant separation versus rooming-in on the duration of breastfeeding (exclusive and total duration of breastfeeding).

**Methods:** We searched the Cochrane Pregnancy and Childbirth Group's Trials Register (7th April 2010) for randomised or quasi-randomised controlled trials (RCTs) investigating the effect of separate mother-infant care versus rooming-in after hospital birth or at home on the duration of breastfeeding, proportion of breastfeeding at six months and adverse neonatal and maternal outcomes. Two review authors independently assessed the studies for inclusion. Results were expressed as mean difference (MD) and relative risk (RR) with a 95% confidence interval (95%CI).

**Results:** There were 20 reports from 18 potential trials. After assessment, one trial (involving 176 women) was included. The mean duration of any breastfeeding was slightly lower in the separate care group compared to the rooming-in group but this difference was not significant (MD -0.35; 95%CI -1.47 to 0.77). The rate for exclusive breastfeeding before discharge from hospital (at day 4 of life) was significantly lower in the separate care group compared to the rooming-in group (RR 0.52; 95%CI 0.36 to 0.75).

**Conclusion:** We found little evidence to support or refute the practice of mother-infant separation. Therefore, we see no reason to practise it. Rooming-in should be considered the norm until further evidence is available. We recommend a well designed RCT to investigate all of the primary and secondary outcomes suggested in this review.
ORAL PRESENTATIONS 1

OP6

INFLUENZA A (H1N1): CHARACTERISTICS AND TREND OF PAEDIATRIC HOSPITALIZATION IN UKM MEDICAL CENTRE BETWEEN YEAR 2009 – 2011

AL Hasniah¹, SW Wong¹, AJ Nordiah², I Ilina, SF Tang²

1. Paediatric Department, Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur, Malaysia
2. Microbiology & Immunology Department, Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur, Malaysia

Objective: To describe the characteristics and trend of children hospitalized with H1N1 infection between 2009 and 2011.

Methods: This was a medical chart review of all children with influenza-like illness (ILI) symptoms with positive real-time polymerase chain reaction for influenza A (H1N1), hospitalized in the Paediatric Ward, UKM Medical Centre between July 2009 and April 2011.

Results: A total of 107 children with confirmed H1N1 infection were included in this study. The median age was 2.8 (range 0 – 18) years. Sixty-eight cases were identified during the worldwide pandemic period from July to September 2009, while another 39 cases were confirmed during the post-pandemic period (September 2009 - February 2011). During the post-pandemic period, a surge in cases was seen in January 2011 (13 of 39 cases). The prevalence of co-morbid illnesses among these children was high i.e. 70.6% (48/68) and 59.0% (23/39) in pandemic and post-pandemic period respectively; the most common being bronchial asthma (20.6%). The clinical presentation was similar in both groups with fever (97.2%), cough (94.4%), coryza (65.4%), respiratory distress (43.9%), gastrointestinal symptoms (19.6%) and seizures (11.2%). About 50% of patients presented with pneumonia, 35.5% had upper respiratory tract infection, 7.5% had febrile seizures and 3.7% had acute gastroenteritis. From the total cohort, nine patients (8.4%) required mechanical ventilation and three patients died (2.8%). There was no significant difference in the need for respiratory support in both groups. Median duration of hospital stay for both pandemic and post-pandemic period was 4 days.

Conclusion: Children hospitalized for H1N1 infection were mainly toddlers and most had associated co-morbid illnesses. H1N1 continued to be a cause of ILI even after the pandemic with a surge seen during January 2011. The clinical characteristics and severity of illness were similar during the pandemic and post-pandemic phases. There should be continued surveillance for this infection in patients presenting with influenza-like illness especially in those with co-morbidities.
URINARY TRACT INFECTION IN PROLONGED NEONATAL JAUNDICE

Mohd Zaidi Yusoff¹, Hans Van Rostenberghe², Teh Keng Hwang³, Jeyaseelan A/L Nachiappan⁴

1. Information not available
2. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
3. Information not available
4. Information not available

Introduction: Thorough clinical assessment is warranted in all cases of prolonged jaundice, but the threshold and range of investigations remain debatable.

Objective: This study was undertaken to determine the incidence of UTI (Urinary Tract Infection) among the prolonged jaundice patients in Alor Setar, Kedah and correlate the presence of UTI with other symptoms, signs and risk factors.

Methodology: It is a cross sectional descriptive study. Neonates with jaundice that persist after 14 days of life were included in the study. Premature babies and patients who were seriously ill, requiring admission were excluded from the study. A detailed history was taken and a thorough physical examination was done to look for risk factors or other signs and symptoms of UTI other than jaundice. A work-up for prolonged jaundice was done. UTI will be confirmed by suggestive urine microscopy together with positive urine culture and confirmation by 2 consultant paediatricians.

Results: Total numbers of recruited patients are 101. UTI was diagnosed in 2% of the prolonged jaundice patients. Both of the patients with UTI had poor weight gain.

Discussion: Incidence of UTI in prolonged jaundice patients is quite low in our region and each patient with UTI had also poor weight gain.

Conclusion: If other studies would confirmed our data, not all prolonged jaundice patients may need to be screened for urinary tract infection via urine culture.
ORAL PRESENTATIONS 2

OP8

KNOWLEDGE ON PEDIATRIC PALLIATIVE CARE BETWEEN MALAYSIAN AND JORDANIAN PEDIATRIC NURSES

Fadi Musa Abusuqair¹, Ariffin Nasir¹, Azriani Ab Rahman², Khaled Khader³, Hans Van Rostenberghe¹

1. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
2. Department of Community Medicine, School of Medical Sciences Universiti Sains Malaysia, Malaysia
3. Palliative care unit, King Hussein Cancer Center, Amman, Jordan

Objective: To compare the knowledge of palliative care between Malaysian and Jordanian Pediatric nurses.

Methods: A comparative cross sectional study was conducted among Pediatric nurses in hospital Universiti Sains Malaysia, King Hussein Cancer centre and Al Bashir Hospital in Jordan between June to September 2010. Self administered validated PCQN questionnaire consisting of 20 items assessing knowledge on palliative care was used. Independent t-test and multi factorial ANOVA was used to compare the mean score of palliative care knowledge between Malaysian and Jordanian Pediatric nurses.

Results: A total of 91 HUSM nurses and 92 Jordanian nurses were studied. We found that there was no significant difference in adjusted mean score of palliative care knowledge between Malaysian and Jordanian nurses.
ORAL PRESENTATIONS 2

OP9

MUTATION SPECTRUM OF DYSTROPHIN GENE IN MALAYSIAN PATIENTS WITH DUCHENNE/BECKER MUSCULAR DYSTROPHY

Abdulqawee M Rani¹, Teguh H Sasonko¹, David J Bunyan², Abdul R Salmi³, Bin A Zilfalil³, Zabidi AMH Zabidi–Hussin³

1. Human Genome Centre, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
2. National Genetics Reference Laboratory (Wessex), Salisbury Hospital NHS Trust, Salisbury, Wiltshire, UK
3. Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

The gene for Duchenne/Becker Muscular Dystrophy (DMD/BMD), is the largest gene in the human genome. Data on the mutation spectrum of this gene in Malaysian patients with DMD/BMD are limited. In this study, we identified the mutation spectrum of dystrophin gene in Malaysian patients with DMD/BMD. 35 patients with symptoms and signs of DMD or BMD were screened for mutations in the 79 exons of the dystrophin gene using multiplex ligation-dependent probe amplification (MLPA) followed by direct sequencing of selected exons. 28 patients (80%) showed mutations of the Dystrophin gene; two of them were novel (c3709A>T and c.4742delA). The MLPA technique also found 23 patients (66%) with confirmable deletions of one or more exons and one patient (3%) with a single exon duplication. Confirmatory direct sequencing on patients found by MLPA to carry an apparent one exon deletion revealed a patient with a single nucleotide deletion (c.4742delA). Further direct sequencing on selected exons detected 3 cases with nonsense mutations and one case with missense mutation. We could not find Dystrophin mutations in the rest 7 patients (20%). According to previous studies, this detection rate is reasonable since 20% of Dystrophin mutations may be occurred as small mutations beyond the studied exons. Our study showed that the majority of the mutations (61%) were in the distal hotspot. Although most of our clinical and molecular diagnoses showed compliance to the reading frame rule, we found two unrelated DMD patients with an in-frame deletion. We found the MLPA approach to be simple, rapid and reliable, although it does need to be supported by other independent methods in certain cases. This study showed the mutation spectrum of Dystrophin among Malaysian patients with DMD/BMD and conforms to other studies, which reported that the distribution of mutations were concentrated in the distal hotspot of the gene, although the frequency of the mutations along the gene may vary.
ORAL PRESENTATIONS 2

OP10

CLINICAL FEATURES AND SHORT TERM OUTCOMES OF CONGENITAL DIAPHRAGMATIC HERNIA IN CHILDREN

Rozita Abdullah, Noraida Ramli

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

Objectives: This study aims to determine the case fatality rate and associated risk factors for mortality.

Methodology: This was a descriptive study of children with congenital diaphragmatic hernia admitted to Neonatal Intensive Care Unit and general Paediatric ward conducted in Hospital Universiti Sains Malaysia and Hospital Sultanah Nur Zahirah from January 1998 until June 2010. For retrospective collection of cases, the medical records of all children presented with diaphragmatic hernia starting from January 1998 were retrospectively reviewed. The prospective data collections were started from 1st October 2009 until end of June 2010.

Results: A total of 73 patients were included in the study, 68 retrospective cases and 5 prospective cases. Using univariate analysis, factors associated with mortality were presence of PPHN, pneumothorax, lung hypoplasia and congenital anomalies; usage of HFOV, delayed timing of surgical repair, place of delivery, blood gases parameter and Apgar score. Using multivariate analysis only two independent predictors for mortality were found i.e. presence of PPHN and delay surgical operation (p value 0.021 and 0.019 respectively). Delay surgical operation was strongly associated with survival rate while presence of PPHN significantly associated with death.

Conclusion: This study suggests additional predictors for outcome in CDH, which were PPHN as a strong predictor for mortality and delay timing of operation as strongly associated with survival.
ORAL PRESENTATIONS 2

OP11

CHILDHOOD CANCER: PARENTS’ PERCEPTION ABOUT VISITING ‘BOMOH’ FOR TREATMENT

Azizah Othman, Fahisham Taib, Ariffin Nasir, Norsarwany Mohammad

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

Diagnosis of childhood cancer is a great challenge to the parents. The condition imposes them to look for all possible ways to help and protect the children. Whilst the conventional medical treatment for childhood cancer is promising, a majority of parents would seek for non-conventional treatment, in preference to or complimenting medical intervention. One of the most commonly seek non-conventional treatment in the community is to visit ‘bomoh’ – a famous traditional healer in a Malay community. The present study examines parents’ perception and knowledge about visiting ‘bomoh’ for treating cancer in their children. We interviewed parents seen at pediatric oncology ward and clinic Hospital Universiti Sains Malaysia (HUSM). Demographic information and series of questions related to the use of ‘bomoh’ treatment were asked using a semi-structured questionnaire developed by the researchers. Parents’ perceived knowledge, conviction, as well as their involvement with ‘bomoh’ in the attempts to treat the children are explored. Parents’ use of other alternative treatments is also looked into.
POSTER PRESENTATIONS

P1

EPIDEMIOLOGICAL ANALYSIS OF PAEDIATRIC MEASLES OUTBREAK IN SUNGAI BULOH HOSPITAL

Faisal Mohd Fadzli1, Yap Hsiao Ling2, Syed Jefrizal Syed Jamal1, Najian Saiful Bahari3

1. University Teknologi MARA, Malaysia
2. Sungai Buloh, Ministry of Health, Malaysia
3. University Kebangsaan Malaysia, Bangi, Malaysia

Objective: To describe demographic and clinical findings of paediatric measles patients during a recent outbreak (13th to 20th of March 2011).

Methods: Several paediatric patients were admitted to Sungai Buloh Hospital, in the space of 1 week, with confirmed measles. The method of confirmation was by enzyme-linked immunosorbent assay on serum samples to detect measles specific IgM. Demographic parameters were established for every patient. These parameters were then analysed in order to determine any significant correlation to the measles positivity.

Results

<table>
<thead>
<tr>
<th>MEASLES</th>
<th>GENDER</th>
<th>AGE</th>
<th>ADDRESS</th>
<th>HOUSEHOLD MEMBERS</th>
<th>MMR VACCINATION</th>
<th>POSSIBLE MEASLES CONTACT*</th>
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</tbody>
</table>

*Contact with people with skin rash

Gender and number of household members were found to be significant risk factors for developing measles. In addition, this cohort of patients was also found to have a higher risk of developing secondary complications from measles.
Conclusion: Measles is now re-emerging as a significant disease amongst the paediatric population. In cases where there is a doubt to the diagnosis, patients have a higher possibility of having measles if they are female and live in a large family. Health professionals need to be extra vigilant as paediatric measles patients are more likely to develop secondary complications.
POSTER PRESENTATIONS

P2

KAWASAKI DISEASE PRESENTING WITH JAUNDICE AS THE INITIAL PRESENTATION: CASE REPORT OF 2 CASES

Andy Rahardja¹, Kok Juan Loong²

1. Paediatric Department, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak, Malaysia
2. Paediatric Department, Sarawak General Hospital, Malaysia

Jaundice is usually a minor presentation of Kawasaki Disease (KD) and includes hepatobiliary dysfunction or gallbladder hydrops. We are reporting 2 cases of unusual Kawasaki Disease with fever and jaundice as the initial presentation. In one patient, other signs typical of the Kawasaki disease occurred a few days later and permitted diagnosis, while in the other there were no typical signs of Kawasaki Disease. Both patients improved after given intravenous immunoglobulin. Echocardiography revealed heart abnormalities in both patients. Our case report suggests that KD should be added to the differential diagnosis of a febrile and icteric young patient.
SUCCESSFUL TREATMENT WITH IMATINIB IN A CHILD WITH FIP1L1-PDGFRA NEGATIVE HYPEREOSINOPHILIC SYNDROME

Teoh YL, Yeoh SL, Nallusamy R

Paediatric Hematology Oncology Unit, Penang Hospital, Pulau Pinang, Malaysia

Background: Hypereosinophilic syndrome (HES) is a heterogenous disease characterized by marked hypereosinophilia associated with eosinophilic infiltration of multiple organs. The subsets of HES include FIP1L1-PDGFRA associated HES, chronic eosinophilic leukemia (CEL), lymphocytic-HES (L-HES), myeloproliferative-HES (M-HES), idiopathic-HES and organ-restricted eosinophilic disease. The classification is important for the choice of treatment. HES with positive FIP1L1-PDGFRA fusion gene generally respond well with tyrosine kinase inhibitor, imatinib mesylate.

Case: We describe a 7 years old girl who presented with persistent fever, leukocytosis (60 x 10^9/L) with marked eosinophilia (31 x 10^9/L), hepatosplenomegaly, and non-specific cutaneous lesions. Her bone marrow smear revealed hypercellular marrow with eosinophilia, liver biopsy showed chronic active hepatitis, ECHO was normal and molecular study confirmed negative FIP1L1-PDGFRA fusion gene. She was treated with corticosteroid and hydroxyurea but the eosinophilia persisted. Three months later she was admitted to paediatric intensive care unit for severe acute respiratory distress and was ventilated for 14 days. Vincristine was added with some improvement. However, her clinical response was not sustained with rebound eosinophilia within a month and she developed multiple colonic ulcers. Finally she was given a trial of imatinib in which she responded well with normalizing eosinophil count and regressing hepatosplenomegaly.

Discussion: Treatment with imatinib among HES patients with negative FIP1L1-PDGFRA gene has not well been established. The response to imatinib of such patients suggests unidentified cytogenetic rearrangement that leads to acquisition of imatinib-sensitive autonomous tyrosine kinase activity. Further molecular study is needed to improve the outcome in patients with HES.
POSTER PRESENTATIONS

P4

GANGLIONEUROMA

Noor Shafina Mohd Nor¹, Syed Jefrizal Syed Jamal¹, Zahilah Filzah Zulkifli²

1. Faculty of Medicine, Universiti Teknologi MARA, Selangor, Malaysia
2. Paediatric Department, Hospital Sungai Buloh, Selangor, Malaysia

Ganglioneuromas are rare tumours arising from neural crest cells. Although rare, ganglioneuromas can cause significant morbidity due to its mass effects. We report a case of a 3-year-old boy who presented with abdominal pain and refusal to walk. The patient had not passed any urine for 36-hours prior to presentation. On examination, there was a paravertebral swelling at the lumbar region which was claimed to be present since birth. The patient had reduced power of both his lower limbs with absent reflexes. He was also noted to be hypertensive. The mass was non-tender, firm in consistency, measuring 6cm x 7cm with no associated skin changes. MRI-spine was performed and showed a paraspinal mass at the level of T10 to L4 with epidural extension and cord compression. The appearance was suggestive of neuroblastoma. The patient subsequently underwent laminectomy. Due to the mass being highly vascularised, only partial excision was possible. Histology confirmed the mass as ganglioneuroma. The lower limb power subsequently improved and the patient was discharged home with continuing rehabilitation. Although ganglioneuromas are uncommon in the paediatric population, it should be kept in mind since surgical removal can significantly improve morbidity.
A CHILD WITH RECURRENT RESPIRATORY INFECTION AND TELANGIECTATIC RASHES DUE TO FUCOSIDOSIS: PICTORIAL ASSAY OF FIRST CONFIRMED CASE IN MALAYSIA

Law HN, Chieng CH, Toh TH, Wong SC, Siaw KNS, Zabedah MY, Ngu LH

1. Paediatric Department, Sibu Hospital
2. Radiological Department, Sibu Hospital
3. Specialized Diagnostic Centre, Institute for Medical Research, Kuala Lumpur
4. Clinical Genetics Department, Kuala Lumpur Hospital

Introduction: Fucosidosis is a rare lysosomal storage disease (LSD) caused by deficiency of alpha-L-fucosidase enzyme. We described the clinical and radiological features of the first confirmed case of fucosidosis in Malaysia.

Case report: Our patient was a 5 ½-year Iban girl who had recurrent respiratory infection since infancy requiring frequent hospital admissions. Her developmental milestones were normal in the first year but subsequently delayed: only walked independently at 3 ½-year and had never acquired speech. Mild coarse facies, large tongue, hepatosplenomegaly and bilateral sensorineural hearing loss was noted when she was 2 ½-year. At 4 years 11 months old, she had a severe pneumonia requiring prolonged ventilatory support and tracheostomy, resulted in chronic lung disease. At this time diffuse telangiectatic rashes over her palms, soles, trunk and umbilicus was noted. She also had insomnia, frequent blepharospams, hypohidrosis and kyphosis. Skeletal survey showed features of dysotosis multiplex but her urinary glycoaminoglycan analysis was normal. Urinary oligosaccharides analysis revealed presence of abnormal oligosaccharides bands. Lysosomal enzymes studies in peripheral blood leucocytes found markedly reduced alpha-L-fucosidase level (<0.1, normal 0.4-2.7 nmol/min/mg protein). Her cranial MRI showed reduced cerebral white matter volume, enlarged CSF spaces, signal abnormalities of globus pallidi, external capsules and posterior limbs of internal capsules, and a well-defined lesion in the right thalamus. At 5 ½-year, she had evidence of neuro-regression, spasticity and required tube feeding. Some of the telangiectatic rashes had evolved into angiokeratoma.

Discussion & Conclusion: The early diagnosis of rare LSD could be difficult and challenging. The presence of coarse facies, neurological deterioration, skeletal dysostosis, visceromegaly and unusual skin rashes should prompt diagnostic evaluation for LSD. A definite diagnosis will enable accurate genetic counseling.
A RARE CASE OF TWO CLOSELY SPACED NONSENSE MUTATIONS IN THE DMD GENE IN A MALAYSIAN FAMILY

Abdul Qawee Rani¹, Rusdy Ghazali Malueka², Teguh Haryo Sasonoko¹, Hiroyuki Awano², Tomoko Lee², Mariko Yagi², Bin Alwi Zilfalil³, A B Razak Salmi³, Yasuhiro Takeshima², Z.A.M.H. Zabidi-Hussin¹, Masafumi Matsuo²

1. Human Genome Centre, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia
2. Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan
3. Department of Pediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

Identification of one nonsense mutation in the DMD gene has been considered an endpoint of genetic diagnosis. We reported two closely spaced nonsense mutations the DMD-causing gene. Our mutation analyses identified two nonsense mutations (p.234SNX and p.249QNX) separated by only 44 nucleotides within exon 8 of the gene in an 18-year-old Malaysian DMD patient. Further analyses on the rest of the family members showed that the proband's mother carried both mutations on one allele. To our knowledge, this is the first example of two nonsense mutations identified in the DMD gene. Multiple mutations suggest hypermutability of the inherited allele and may explain the occasional discrepancies between genotype and phenotype in dystrophinopathy.
POSTER PRESENTATIONS

P7

STROKE IN INFANCY: A RARE COMPLICATION OF HIB MENINGITIS

A Zainal¹, Nor Azizah A¹, Khuzaih R¹, Hargeet K², Nor Ain M²

1. Faculty of Medicine, Universiti Teknologi Mara (UiTM), Selayang Campus, Selangor, Malaysia
2. Department of Paediatrics, Hospital Selayang, Selayang, Selangor, Malaysia

The introduction of vaccine for Haemophilus Influenzae type b (Hib) has lead to a dramatic decrease in the incidence of Hib meningitis in Malaysia. Nonetheless, bacterial meningitis remains a potential threat. Stroke, although a rare entity in children, is one of the most devastating complication of bacterial meningitis. We report a case of a baby girl suffered from Hib meningitis presented with stroke.
POSTER PRESENTATIONS

P8

CARDIAC RHABDOMYOMA: A CASE REPORT


Neonatal Intensive Care Unit, Department of Paediatrics, School of Medical Sciences,
Universiti Sains Malaysia, Kelantan, Malaysia

A term baby boy, a product of non consanguineous marriage, was born to a young primigravida mother at term plus 12 days gestations. He was born via emergency lower segment caesarean section for poor progress of labour with the birth weight of 2.49kg. He was intubated following direct suction for born not vigorous with thick meconium stained liquor and ventilated with HFOV with the diagnosis of meconium aspiration syndrome. On examination, he was small for the age but did not appear dysmorphic. He was tachypnoeic on ventilator with SPO2 of 100%(pre ductal) and 88%(post ductal) with high ventilator setting. Chest examination revealed reduced breath sound on the left side.

Chest radiograph showed cardiomegaly with normal pulmonary vasculature. Echocardiography revealed huge anterior mass compressing and covering the heart. CT scan thorax was done showed large hypodense enhancing intracardiac mass seen within the right ventricle. The patient remained unstable with persistent hypoxemia. He further deteriorated and became asystole at almost 24 hour of life. Biopsy was performed and confirmed as cardiac rhabdomyoma. The mother had classical features of tuberous sclerosis.
A CASE OF CONGENITAL TUBERCULOSIS

Mohd Iqbal I, Nor Rosidah I, Hans Van Rostenberghe, Noraihan I, Noraida R

Neonatal Intensive Care Unit, Department of Paediatrics, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia

We reported a term, 3.7 kg baby boy, who was admitted to the Neonatal ward at day 4 of life for fever and abdominal distension. He continued to have persistent high grade fever, progressive massive hepatosplenomegaly with severe neonatal hepatitis, pancytopenia and poor weight gain. Multiple blood and urine cultures were negative for bacterial and fungal but the C-reactive protein were elevated > 100mg/dL. There were no response to empirical antibiotics and antifungal therapy. There was no history to suggest primary or secondary immunodeficiency. History from mother revealed that she had chronic cough with constitutional symptoms during third trimester but resolved a month prior to delivery.

Ultrasound abdomen revealed multiple splenic micro abscess. The diagnosis of neonatal tuberculosis was made at day 21 of life once gastric lavage were positive for acid fast bacilli for 3 consecutive days. Unfortunately cultures for mycobacterium tuberculosis were unsuccessful. Treatment in this baby posed a challenging task due to severe hepatitis. Prior to treatment initiation, the AST was 773 IU/L, ALT 268 IU/L and total bilirubin 357 umol/L (direct 70%). He was started on isoniazide, pyrazinamide, rifampicin and prednisolone. At day 5 of treatment, isoniazide was withheld due to worsening hepatitis and replaced by amikacin for 2 weeks. Isoniazid was restarted at a lower dose when the liver enzyme improved. The baby slowly responded to the antituberculosis drug and was discharged at day 95 of life. He completed 12-month course of treatment with complete recovery of his bone marrow and liver function.