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PLENARY 1

NUTRIENTS AND THE DEVELOPING BRAIN

Zabidi Azhar Hussin

School of Medical Sciences, Universiti Sains Malaysia, Kelantan

Recent advances in neuroimaging, neurophysiology and neurochemistry have made it possible for scientists to decipher more precisely the essential role of nutrients in the developing brain. It has been known for a long time that some essential nutrients notably zinc, copper, iodine, selenium choline and folate exert their influence on the developing brain by affecting myelination, altering neurotransmitter synthesis and affecting hippocampal energy metabolism. Copper and zinc in particular affect cerebellar function and the autonomic nervous system. Deficiencies depend on the timing of gestation as different parts of the brain undergo different rates of synapses and myelination at different phases of the pregnancy. Early insults result in reduction of cell numbers through reduced proliferation, while later insults adversely affect cell differentiation and synaptogenesis. Deficiencies are also determined by the areas of the brain most vulnerable at the time of the deficiency. A particularly critical observation in a state of nutritional deficiencies is the effect on neurochemical and receptor synthesis, which may result in long term alteration in neuronal performance.

These new knowledge further highlights the importance of optimum maternal nutrition during pregnancy. Some of the effects on the developing brain are known to be irreversible even after repletion of the deficient nutrient despite the well established understanding of the neuroplasticity concept. Maternal health and well being should therefore receive greater emphasis in the training of Paediatricians for the reasons described above.
PLENARY 2

NEW GENETICS AND ITS IMPLICATIONS FOR PAEDIATRIC PRACTICE

Thong Meow Keong

Genetics & Metabolism Unit, Department of Paediatrics, Faculty of Medicine, University of Malaya, Kuala Lumpur

Genetic disorders and birth defects collectively form an important component of the health burden and a main cause of chronic illnesses in childhood. Three percent of newborns have major birth defects and represents a major contributor to under-five mortality in Malaysia. While single gene disorders are important from the family’s perspective, other multifactorial conditions such as neural tube defects and cleft lip and palate involving the interplay between genetic and environmental factors are important from the public health’s perspective. In addition, conditions with strong genetic susceptibility such as cancers, diabetes and asthma have become increasingly prominent and many families are turning to their paediatricians for advice and information.

Prevention of genetic diseases may be achieved through empowering individuals with information about their genetic risks and enabling them to make informed choices about their reproductive options. Reducing genetic morbidity and mortality would be a secondary goal of prevention. The paediatrician can assist the clinical geneticist in playing an active role in this respect. This include public education regarding genetic diseases, birth defects and strategies to avoid teratogens, providing basic genetic counselling for at-risk families, promotion of periconceptional folic acid supplementation and advising pre-pregnancy management of maternal illnesses and early antenatal visits once pregnancy is confirmed. These strategies require an understanding of the natural history of the disorders, genetic testing techniques and their limitations, and the basic knowledge of genetic counselling.

A good family history will help in identifying family members at risk for genetic diseases. Genetic counselling is defined as a communication process that deals with the human problems associated with the occurrence or risk of occurrence of a genetic disorder in a family. Genetic counselling helps the individual or family to understand the diagnosis, burden of disease and the available management, the risk of recurrence in relatives and makes the best possible adjustment to the disorder in an affected family. The paediatrician must be able to differentiate between merely giving genetic information and performing genetic counselling.

Providing timely information on the avoidance of teratogens and encouraging all women of child bearing age to consume periconceptional folic acid supplementation also reduce birth defects such as neural tube defects. Population screening and counselling for genetic diseases may empower carriers to make appropriate decision regarding the choice of life partners and prenatal diagnosis. Technology has also changed the roles of the paediatricians, for example accessibility to internet information and the introduction of newer techniques leading to over-medicalisation and raising parental anxiety are some of the implications in paediatric practice today. The paediatrician must be aware of the ethical, legal, social and religious implications of this ‘new genetics’.

The paediatrician’s evolving roles of this ‘new genetics’ will become increasingly crucial for the health of children in the decade to come.
An outbreak of acute respiratory illness caused by novel swine-origin influenza A(H1N1) virus was identified in Mexico in late March 2009. This novel virus subsequently caused widespread infections in many countries and prompted WHO to declare an influenza pandemic on 11 June 2009. This pandemic had presented an opportunity to define the local epidemiology, clinical features, laboratory investigations and outcome among hospitalised children in Malaysia. The first case of 2009 pandemic influenza A(H1N1) among children in Malaysia was documented on 18 June 2009. The number of cases surged significantly from epid week 30 to peak at epid week 33, followed by a gradual decline from epid week 43 onwards. A total of 1362 children with laboratory-confirmed 2009 pandemic influenza A(H1N1), diagnosed by RT-PCR, were documented between 18 June 2009 to 1 March 2010. 861(63.2%) were below 5 years, while 536(39.4%) were below 2 years. The illness was predominantly respiratory, although extrapulmonary involvement was occasionally documented. 101(7.4%) children were intubated and received ventilatory support. 461(35.3%) had a co-morbid disorder. The illness was generally mild and the case fatality rate was 3.7%, comparable to that reported elsewhere. Presence of a co-morbid illness was associated with a four-fold increased risk of death in our cohort. In conclusion, the 2009 pandemic influenza A(H1N1) illness was generally a mild disease in our local children. Early antiviral therapy and targeting young children below 2 years with an underlying co-morbid illness for prompt treatment may have been beneficial.
Growth is a complex physiological process that requires the interaction of various factors including genetic constitution, nutrition, hormones, normal metabolism and psychosocial wellbeing of the individual. Stature is a measure of previous growth. Short stature thus maybe due to any abnormalities of the above factors which result in slow growth rates. Occasionally short stature could also be a result of previous rapid growth which causes premature ephyseal fusion, preventing further growth. Since 1985, there is an abundant supply of growth hormone (GH) by recombinant technology, and GH is not only indicated in individuals with GH deficiency, but it has been shown to be effective in short individuals who were born small for gestational age (SGA), those with Turner syndrome and a small subset of those with idiopathic short stature. GH treatment is expensive and is effective in improving final adult height only if it is given in life. The challenge is to accurately determine the underlying cause of short stature and identify early those who will benefit from GH treatment and not wait till in their adolescent years. This lecture will focus on the practical approach in evaluating a short child based on a focused history, physical examination and relevant investigations.
Puberty is the result of increasing gonadotropin releasing hormone (GnRH) release by the hypothalamus followed by a complex sequence of endocrine changes with functioning of negative and positive feedbacks, and associated with the development of sex characteristics, a growth spurt and reproductive competence. Delay in puberty can be the result of a lag in normal pubertal maturation of the hypothalamic-pituitary-gonadal axis or it may represent an underlying abnormality. Abnormality may also be present if puberty has begun but does not progress appropriately. Therefore, appropriate evaluation is needed so that treatment can be started in a timely manner. Hypogonadotropic hypogonadism (HH) is often the result of GnRH deficiency. The clinical picture is characterized by the absence of pubertal development and infertility. It is difficult to differentiate HH from constitutional delayed puberty. Androgen remains the mainstay of hormonal therapy in boys, though other alternatives are GnRH pulse therapy and hCG therapy. There are various forms of estrogen preparation for the pubertal induction in girls. It is important to induce puberty gradually to mimic normal physiological process for optimal outcome. Occasionally there are reasons to delay the induction of puberty, such as in patients on growth hormone therapy. This has to be balance with the risk of a lifelong diminution of bone mineral density, and the possibility of accentuating the social isolation and stigmatization of patients. The approach is to customize therapy according to individual condition and psychological needs of each patient.
SYMPOSIUM 1

GROWTH AND PUBERTAL DISORDERS

S1.3

CONGENITAL HYPOTHYROIDISM

Fuziah Md. Zain

Department of Paediatrics, Hospital Putrajaya, Selangor

With the changing priorities in health care, the improvement in economy and lifestyle will lead to preventive issues that includes national screening programs which have been well established in many industrialized countries. Congenital hypothyroidism (CH) is difficult to diagnose at birth as newborns lack the typical clinical features in the first few weeks of life. Diagnosis is usually made by neonatal screening studies. A delay in diagnosis and treatment causes mental retardation, developmental delay, growth failure and some neurologic sequelae. Screening programs for congenital hypothyroidism (CH) were started in USA, Canada and Western Europe in 1974 with a prevalence of 1:4000 births. In October 1998, Ministry of Health (MOH) commenced newborn screening for CH in Malaysia. The incidence from pooled studies from South East Asian countries is 1:3,093. The screening methods practiced are primary TSH screening and screening for low thyroxine level. Blood is taken from the heel on day 5 of life or from the umbilical cord at birth. The latter is being practiced in Malaysia as babies are discharged soon after delivery. A total thyroxine level of <6 µg/dL or TSH >25 mU/L is usually indicative of CH, but values differ by laboratory. Since 1998, 104 hospitals have carried out newborn screening for CH with over 2 million babies screened giving an incidence of 1:3,914 (data from MOH). Physical growth, IQ level, mental and motor development of infants with CH usually are normalised with early levo-thyroxine therapy. In conclusion, newborn screening for CH is an essential preventive public health program in early detection and treatment. However, even experienced screening programs are not perfect as about 8 -10% of cases is still missed. Hence, doctors must still apply their clinical acumen. In the 21st century, the challenge of maintaining excellence in the existing programs, developing new screening test including molecular genetic defects in CH, assuring optimal follow-up and management must be met for all children.
Dental caries affecting preschool children has become a major health care problem in many countries. In the USA, The national health and nutrition examination survey (NHANES ) revealed that 8.4% of 2 year old children had at least one decayed or filled tooth and by the age five almost 40.4% of children were affected. Dental caries in primary teeth is one of the major reasons for hospitalisation of children and is costly to treat. In Malaysia the prevalence of dental caries in children below the age of 5 years is 87.1% (Ministry of Health 1995). Early childhood caries (ECC) is the term used to describe dental caries in the preschool child (below 71 months of age). ECC if not managed early may result in rapid destruction of tooth structure and involvement of the dental pulp. This may cause pain, infection and result in early loss of the tooth at a very young age.ECC occurring in medically compromised children can further complicate their medical condition and put them further at risk. Indeed many of these children will end up needing to be treated in hospitals using General anaesthesia .The hallmark of management of ECC is prevention. It is therefore imperative that all health care providers especially those treating children should be aware of this infectious and highly debilitating disease and play an important role in preventing its occurrence and severity. This lecture will give a general overview of early childhood caries and its management.
DENTAL MANAGEMENT OF CHILDREN WITH MEDICAL PROBLEM

Azillah Mohd Ali

Klinik Pakar Pergigian, Hospital Sultanah Bahiyah, Kedah

Dentistry is safe provided the patient is healthy and the procedure is not dramatically invasive. Having a chronic medical condition can directly affect the provision of dental care and some, where the consequences of dental disease or even dental treatment, can be life-threatening. Many health personnels fail to acknowledge that oral assessment is an important part of the examination of the child’s overall health. This 25 minutes session will cover common oral manifestations of patient with medical problem and early recognition and appropriate referral will be highlighted. As dental disease can have grave consequences on these patients and so rigorous prevention is important. Planning for dental care should be an aggressive and well planned preventive regimen which includes dietary counselling, fluoride therapy, fissure sealant, oral hygiene instruction and regular monitoring clinically and radiographically. The importance and effectiveness of early intervention is greatly emphasised.
SYMPOSIUM 2

DENTAL SYMPOSIUM 1

S2.3

DENTO-FACIAL TRAUMA IN CHILDREN

Thevadass K. Palany

Klinik Pakar Pergigian, Hospital Raja Permaisuri Bainun, Perak

Trauma is one of the most common presentations of children to a pediatric dental surgeon. The management of dento-facial trauma in children is distressing for both child and parent and often difficult for the dental practitioner. The majority of injuries involve the dentition and its supporting structures. Facial fractures are rather uncommon in children. The principles of management of traumatic injuries in children may differ from that in adults due to certain anatomical and patho-physiological factors specifically related to children. Careful assessment with proper treatment of dento-facial injuries can have a significant effect on outcome and prognosis. Children require long-term follow up to monitor potential complications including growth abnormalities. This presentation would give an overview of the common manifestations of different types of dento-facial trauma in children with particular reference to classification, assessment and formulation of treatment strategies.
Improved outcomes of critically ill children managed in paediatric intensive care units has increased the pressure to transfer such patients to a tertiary centre. However the transport of critically ill children can be hazardous with inherent risks related to deterioration from the primary illness, complications of treatment and the transfer process itself. It has been shown that up to 75% of patients transferred by non-specialised teams can suffer serious clinical complications. The solution would be to send a team of experienced staff to stabilize and then transport the patient. Britto et al \(^1\) showed, in a prospective study, that critically ill children can be transferred with minimal morbidity and mortality related to transport, when the transport was performed by a specialized paediatric retrieval team. More recently, in another prospective cohort study, Orr et al \(^2\) showed that the use of a specialized transport team was associated with improved survival rates and fewer unplanned events during transport. After adjustment for illness severity, only the use of a nonspecialised team was independently associated with an unplanned event and death. Thus, transport of critically ill children to a paediatric tertiary care centre can be conducted more safely with a paediatric critical care specialized team than with teams lacking specific training and expertise in paediatric critical care and paediatric transport medicine.
The Neonatal and Paediatric Retrieval Unit for Perak was set up in September 2000 under the ‘Dasar Baru’ programme. The aim of the retrieval system is to ensure that ill neonates and children are transferred from district hospitals to referral centres by highly trained and dedicated staff, in stabilised and optimal condition thereby reducing mortality and morbidity. The team is equipped with equipment for resuscitation and stabilisation of the critically ill children. From the pioneer team of 4 paramedical staff in 2000, currently there are 12 staff, comprising of staff nurses and assistant medical officers providing service from 7 am to 9 pm. The staff were trained over a period of one year, consisting of tutorials on important paediatric topics, and performing core procedures in paediatrics. Hands-on practical on equipment use and drug management are also covered. They have to complete their log book of procedures before being certified and credentialled through various assessment. In 2009, the team performed 489 transfers, 301 were internal transfers and 188 external transfers. Of these 382 or 78.11% were patients on IPPV. All team members are NRP instructors and PALS providers and are actively involved in BLS, NRP and PALS training at hospital and state level. The future plan is to expand the service providing 24-hour coverage and to act as a call centre coordinating ventilator beds for the state. Challenges include shortage and turnover of staff and lack of equipment.
The regionalisation of Paediatric Intensive Care Units is the rational strategy to make accessible these specialized and equipment/expertise intensive services throughout the country. An important component of this strategy is the ability to transport critically patients from the referring front-line hospital to the referral centre. The British Paediatric Association has stated that patients during the period of transportation should receive a level of care that is equivalent to that of intensive care. Yet the irony is that the referring hospital is transferring the case precisely because it is not capable of delivering intensive care, yet that level of care must be provided for the patient during the process of transfer. Hence the transport of such children should not be conducted by escorting teams from the referring hospital, but by dedicated and appropriately trained retrieval teams dispatched from the referral hospital. The Paediatric Emergency Transport System of Sabah Women & Children’s Hospital (SWACH) is led by a medical officer, assisted by a staff nurse or assistant medical officer. Although paramedical staff can be well-trained in stabilization and resuscitation procedures, they may nevertheless be unable to cope with the complexity of the patients’ conditions and range of pathophysiological problems that could arise during the period just before and during the transportation. For this reason it was felt that the PETS should best be led by a medical officer rather than a paramedical staff.

The key tasks in the transfer of critically ill patients are:

1. assess the cardiopulmonary status of the patient and ascertain the need for resuscitation
2. carry out the resuscitation effectively and stabilise the patient before considering transferring the patient
3. ensure a secure airway – intubate with an appropriate sized endotracheal tube if necessary, and ensure good anchoring of the tube
4. assess if there is respiratory failure and the need for assisted ventilation, and ensure the provision of the means for maintaining the assisted ventilation along the journey (appropriate ambu bag, extra ET tubes, filled oxygen tanks, suction apparatus and tubes)
5. secure good intravenous access and maintain the appropriate fluid infusion for the journey
6. provide for the necessary medications that are required for the journey
7. close monitoring of the airway, ventilation and cardiovascular status during the transfer
8. anticipate and rapidly treat critical incidents that may arise along the journey.
With the regionalization of Pediatric and neonatal intensive care services it follows logically to develop a regional transport services. This will be developed in phases and would be in accordance with the development of Pediatric and neonatal Intensive care services. Training of the Pediatric intensivist and Neonatologist will be intensified to lead such teams. In future each state will have a PICU and level 3 NICU and with that a regional transport team. This would need a specialized pediatric transport team where the personnel are adequately trained and equipped to provide critical care interventions in the out-of-hospital setting capable of continuing and intensifying goal-directed therapy for a wide range of critical conditions. They are mobile ICU teams, delivering state-of-the-art critical care in the transport environment and will need to fulfill stringent qualifications. A better outcome would be achieved with an early goal directed therapy approach in which pediatric transport team is an extension of the ICU setting. The scoop and run mentality which was practiced before will have to be abandoned in favour of the goal directed therapy which has been shown to improve survival by nine fold.

Other aspects that will be looked into include proper ambulances which are well equipped, adequate monitoring, training of staff and maintenance of their skill, certification in relevant pediatric resuscitation course, BLS, NRP, PALS and APLS. In future transport course will be introduced. Safety issues and challenges of transport from remote areas and islands will be discussed. Audit and research will also be an important component.
The incidence of type 1 diabetes mellitus (T1DM) is rising at an average annual rate of 3-5% and it is the third most common illness in childhood following asthma and cerebral palsy. Type 2 diabetes mellitus (T2DM) is an emerging and serious clinical problem in children. Sedentary lifestyles, obesity and insulin resistance contribute to an explosive increase in the incidence of T2DM among children and adolescents worldwide. The NHMS III survey (2006) revealed a national prevalence of overweight children below 18 years old as 5.4%. Diabetes management is lifelong and unpredictable as it changes throughout childhood and adolescence alongside with age, psychological development, physiology and maturity. The goals of antidiabetic treatment in paediatric patients are no different from the adults. Treatment instituted must achieve certain targets in order to minimize long-term diabetic complications. Multiple complications are present in almost one fifth of patients with diabetes as a result of maintained hyperglycaemia in the younger years. There is global evidence to suggest that metabolic control deteriorates during the period of adolescence. In the United Kingdom, only 15% of under-fifteens with diabetes are currently achieving recommended blood glucose levels as defined by glycosylated haemoglobin of 7.5%. In Malaysia, a total of 240 diabetic patients were reported to the Diabetes in Children and Adolescents Registry (DiCARE) from 2006-2007. Latest data from DiCARE will be presented. Researchers conducted in the western countries have shown that there were many factors affecting adolescent metabolic control of diabetes such as cultural and societal influences that include gender, developmental levels, illness perceptions, school support, peers and the family.
SYMPOSIUM 4

CHILDHOOD DIABETES

S4.2

HOW TO MANAGE TYPE 1 DIABETES MELLITUS?

Yazid Jalaluddin

Department of Paediatrics, University Malaya Medical Centre, Kuala Lumpur

Type 1 diabetes mellitus (T1DM) is the consequence of a progressive autoimmune destruction of pancreatic beta cells triggered by environmental factors in genetically susceptible individuals. Over the last decade, the age of onset of type 1 diabetes has decreased. Managing diabetes in this group of children poses its own unique challenges. The surge of growth and sex hormones together with behavioral issues in adolescents posts a great challenge in managing T1DM adolescents. The only treatment available currently is by insulin injections. Multiple insulin regimen therapy ie twice daily, three times daily, multiple daily injections (MDI) aka as basal-bolus, or insulin pump therapy are now available with its own advantages and disadvantages. The Diabetes Control and Complications Trial (DCCT) had demonstrated that intensive therapy of T1DM delays the onset and progression of life-threatening microvascular complications. In light of these facts, optimal insulin delivery within the context of normal child development remains a significant challenge. Twice or three times daily insulin injections have repeatedly shown to be at a disadvantage compared to MDI in relation to metabolic control. However, MDI of insulin can be cumbersome and unnecessarily complicates the lifestyle of most children. Insulin pump therapy is considered to be the most physiologic method of insulin administration. Insulin pumps allow a quick response to rapidly fluctuating insulin needs and can administer very small doses to highly insulin sensitive children. Timely dose adjustment using pumps allows flexibility in activity and behaviours, and enables a quick response to insulin absorption and hormonal variation. These management tools translate into decreased hemoglobin A1C and hypoglycaemia avoidance. Diabetic ketoacidosis (DKA) results from absolute or relative deficiency of circulating insulin and the combined effects of increased levels of the counter-regulatory hormones: catecholamines, glucagon, cortisol and growth hormone. Absolute insulin deficiency occurs in previously undiagnosed T1DM and when patients on treatment deliberately or inadvertently do not take insulin, especially the long-acting component of a basal-bolus regimen. Patients who use an insulin pump can rapidly develop DKA when insulin delivery fails for any reason. Relative insulin deficiency occurs when the concentrations of counter-regulatory hormones increase in response to stress in conditions such as sepsis, trauma, or gastrointestinal illness with diarrhea and vomiting. The combination of low serum insulin and high counter-regulatory hormone concentrations results in an accelerated catabolic state with increased glucose production by the liver and kidney (via glycogenolysis and gluconeogenesis), impaired peripheral glucose utilization resulting in hyperglycaemia and hyperosmolality, and increased lipolysis and ketogenesis, causing ketonemia and metabolic acidosis. Hyperglycaemia that exceeds the renal threshold (~10mmol/L) and hyperketonemia caused osmotic diuresis, dehydration and obligatory loss of electrolytes, which often is aggravated by vomiting. These changes stimulate further stress hormone production, which induces more severe insulin resistance and worsening hyperglycemia and hyperketonemia. Prompt treatment with exogenous insulin together with fluid and electrolyte replacement would prevent fatal dehydration and metabolic acidosis.
Traditionally diabetes mellitus in childhood is synonymous with type 1 diabetes. This spectrum has changed in last two decades with better understanding of patho-physiology of diabetes and the classification has become more elaborate. The epidemiology of type 2 diabetes mellitus (T2DM) in adolescents in the United States and worldwide has been reviewed multiple times. These reviews confirm the rise in the global incidence of T2DM in children and adolescents over the last two decades and suggest a close relationship between an increase in obesity in the general population and the appearance of the disorder in adolescents. However the true prevalence of T2DM in complicated by the uncertainties in diagnosis and variations in definitions. The current recommendations on criteria of diagnosis will be presented. T2DM is often associated with risk factors for cardiovascular disease that may already be present at time of diagnosis, making normalisation of blood glucose levels and diagnosis and treatment of hypertension and dyslipidaemia important.
SYMPOSIUM 5
DENTAL SYMPOSIUM 2
S5.1
GINGIVOSTOMATITIS AND OTHER GINGIVAL LESIONS IN CHILDREN
Laila Abdul Jalil
Klinik Pakar Pergigian, Hospital Tuanku Jaafar, Seremban

The objective of this lecture is to be able to identify, compare and contrast the common gingival lesions in children. In clinical practice, very often an infection of viral origin is mistaken for bacterial thus leading to unnecessary antibiotic prescription. Gingivostomatitis, a common infection in young children is an example of a case in point. Diagnosis and management of the case are discussed. A list of possible lesions of the gingival from causes other than infection will also be discussed in this lecture.
In many clinical settings, the services of a professional medical photographer may not be available. If a standardized approach is followed those clinicians who take their own clinical photographs can achieve acceptable results. This lecture offers guidance to the clinician on exposure, scale, depth of field and basic techniques of using a digital camera to accurately record a patient’s medical condition.
SYMPOSIUM 5a

PAEDIATRIC SLEEP SYMPOSIUM: IDENTIFYING AND MANAGING SLEEP PROBLEMS IN INFANTS

S5a.2

SLEEP IN INFANTS IN MALAYSIA – FINDINGS OF MALAYSIAN SLEEP SURVEY

Noorashidah Abdul Wahab

Department of Paediatrics, Hospital Serdang, Selangor

997 Malaysian participated in the sleep survey conducted in 2007. The parents of children aged 0 till 36 months completed the brief infant sleep questionnaire (BISQ) on-line asking specific questions on infants daytime and nighttime sleep patterns as well as their sleep related behaviors. 53% of the respondents were Chinese suggestive that most of the participants in this survey stayed in the urban areas with easy access to computers and internet facilities. 49% of the infants and toddlers in the survey were boys. The majority of Malaysian children in the survey sleep in their parents’ room (84%) in which 44% is on the parents’ bed. Most of the children went to bed very late at about 10.47 at night with nighttime sleep of 9 hours which is far more shorter comparing with western children. The total sleep time was 12 hours 27 minutes (day and night). Most of them woke up around 7.30 in the morning. 63.6% of respondents reported that their children sleep well and almost 52% claimed to have bedtime routine. However, 44.7% of respondents reported sleep problems in their children with 20% had bedtime difficulties suggestive that the parents of Malaysian children aged 0 to 36 months have higher perception of sleep problems.
SYMPOSIUM 5a

PAEDIATRIC SLEEP SYMPOSIUM: IDENTIFYING AND MANAGING SLEEP PROBLEMS IN INFANTS

S5a.3

CROSS-CULTURAL DIFFERENCES IN SLEEP IN YOUNG CHILDREN

Jodi A Mindell

The Children's Hospital of Philadelphia, Saint Joseph's University Philadelphia

Sleep problems are highly prevalent in young children, especially bedtime problems and night wakings. However, substantial differences exist in sleep patterns and sleep practices in young children across culturally diverse regions. Results will be presented from a global study of cross-cultural sleep patterns and sleep problems in a large sample of children ages birth to 36 months in multiple predominantly-Asian and predominantly-Caucasian countries. Differences in sleep patterns, sleep practices, and sleep problems, as well as predictors of sleep outcomes and parents’ perception of sleep problems, will be presented. Clinical management recommendations as a result of these findings will be discussed.
Symposium 5a

Paediatric Sleep Symposium: Identifying and Managing Sleep Problems in Infants

S5a.4

The Importance of Identifying and Managing Sleep Problems
(Clinical Perspectives & Practical Screening Tools)

Daniel YT Goh

Department of Paediatrics, Yong Loo Lin School of Medicine,
National University of Singapore, Singapore

Sleep seemingly passive phase of the diurnal cycle. Sleep is important for optimal growth and development as well as learning. Sleep deprivation can be associated with significant health problems and diseases including hypertension, diabetes and even predispose to the development of obesity. These issues are of particular significance in the young where the foundations of health are laid for adulthood and the future. The spectrum of sleep disorders and problems in childhood will be discussed, together with simple approaches to screening patients for sleep problems in clinical practice. From this talk, you will understand the importance of good sleep in achieving good health and how you can achieve these in your patients through a quick assessment in your busy clinical is a basic vital activity in life. Many physiological processes occur in this practice.
SYMPOSIUM 5a

PAEDIATRIC SLEEP SYMPOSIUM: IDENTIFYING AND MANAGING SLEEP PROBLEMS IN INFANTS

S5a.5

SLEEP-RELATED BREATHING DISORDERS (IDENTIFYING AND MANAGING)

Arthur Teng

Department of Sleep Medicine, Sydney Children’s Hospital, Australia

- Sleep-related breathing disorders (SRBD) in infants and children can range from primary snoring to severe obstructive sleep apnoea (OSA) and respiratory failure. Routine childhood health screening should include questions about sleep and snoring. Children at high risk of SRBD should be considered for Sleep Studies where available.
- Snoring is a cardinal symptom of OSA but children can have severe SRBD without very loud snoring.
- Snoring most nights, pauses in breathing during sleep and an increase in work of breathing are the symptoms pointing most strongly towards a diagnosis of OSA in children.
- Children with OSA are often hyperactive and have difficult behaviour during the day. This is in contrast to adults who have excessive daytime sleepiness as a major symptom. They may also have poor attention and/or poor concentration and learning difficulties. Even children at the mild end of the spectrum can have significant consequences.
- The overnight Sleep Study or Polysomnography (PSG) is the gold standard for the investigation of sleep disorders in children and adults. The PSG also picks up other sleep disorders which might have a significant impact on the child’s nocturnal and daytime symptoms. These might include parasomnias, nocturnal seizures and periodic limb movements.
- Adenotonsillectomy (AT) is the treatment of choice for most children with OSA. However children at the severe end of the SRBD spectrum are at high risk of post-operative compromise. There is probably a higher than expected “residual rate” of OSA despite AT and a high index of suspicion is needed especially in the high risk group.
- Nasal mask continuous positive airway pressure (CPAP) or bi-level pressure support (BPAP) is used for the treatment of severe childhood and infantile SRBD where surgical treatment is contraindicated or unsuccessful.
Is it estimated that between 3 - 12% of preschool children snore. The majority of these children is healthy, without other symptoms and has primary snoring. But, about 2% of snoring children have obstructive sleep apnoea syndrome (OSAS), a sleep-disordered breathing (SDB) characterized by upper airway obstruction that disrupt normal ventilation during sleep leading to serious health morbidity. The differentiation of primary snoring and obstructive sleep apnoea syndrome (OSAS) is difficult on clinical grounds. Full polysomnography (PSG) is the gold standard procedure in the diagnosis of OSAS, however it’s used in clinical setting is limited by several factors. The procedure required trained personnel in the paediatric sleep medicine, is labour intensive, time-consuming and very costly. Selection of snoring patients who require further investigation is based on the evaluation of detailed sleep symptoms, clinical findings and associated risk factors related to SDB. Other screening modalities will be discussed.
Cough is very common in children of all age groups. It is a mechanism to clear secretions from the airway. Prevalence of chronic cough in children varies from 5-15%. Definition of chronic cough varies from 2-4 weeks between the prevalence studies. Chronic cough in children can be classified into three groups i.e. expected or normal cough, non-specific cough and specific cough. Symptoms suggestive of specific cough are cough accompanied by chest pain, daily productive cough, difficulty in breathing, feeding difficulties and haemoptysis. Symptoms suggestive of underlying diseases like immune deficiency, neuromuscular diseases and heart disease need further evaluation of the chronic cough. Presence of signs such as clubbing, chest deformity, failure to thrive, tachypnea and abnormal auscultatory findings are suggestive of specific chronic cough and require further investigations. Common causes of chronic cough in children are asthma, post infective cough, protracted bronchitis, upper airway cough syndrome and gastroesophageal reflux disease. Chronic cough in children: when to worry.
Wheezing is a common presenting symptom of respiratory disease in children especially in the preschools. Epidemiologic studies conducted worldwide have shown that 10 to 15 percent of infants wheeze during the first year of life, and as many as 25 percent of children younger than five years of age present to their physicians with wheezing respiratory illnesses. Most children with recurrent wheezing are very likely to have asthma, regardless of the age of onset, evidence of atopic disease, precipitating causes, or frequency of wheezing. However, other diseases can present with wheezing in childhood, and patients with asthma may not wheeze. Therefore, the initial evaluation of a wheezing child should be directed toward the exclusion of alternative diagnoses, followed by a therapeutic trial of bronchodilators if asthma is suspected. The differential diagnosis of wheezing that is chronic or recurrent is broad and includes structural abnormalities of the tracheobronchial tree or other thoracic structures. Nonstructural causes of chronic wheezing include aspiration syndromes, bronchopulmonary dysplasia, bronchiolitis obliteran, vocal cord dysfunction, infective causes, cystic fibrosis and abnormal immunological defense mechanisms. Clinical history, physical examination, laboratory investigations, and response to treatment all play a role in establishing the underlying etiology of wheezing. A diagnostic approach to wheezing in childhood is presented here.
There has been an increased in the recognition among paediatricians on the importance of assessment of nutritional status in children who required hospital care. Hospitalized children who are malnourished have higher rates of infectious complications and delayed recovery, require longer hospital stay, have increased rate of readmission and increased mortality. It has been estimated that between 6-50% of hospitalized children are undernourished. Causes of malnutrition include inadequate intake, increased losses, malabsorption, or abnormal nutrient requirements. It is thus imperative to monitor the nutritional status of hospitalized children regularly. Nutritional assessment entails a detailed examination of metabolic, nutritional or functional variables by an expert clinician or a dietician, which may include a detailed medical history and physical examination, anthropometric and biochemical measurements. Medical history should include dietary history and dietary assessment, paying special attention to conditions that may interfere with food intake and absorption. Physical assessment includes assessment of growth, determination of loss of fatty tissue and muscle strength, signs of malnutrition or micronutrient deficiency. These include a thorough examination of skin, hair, nails, oral cavity, teeth and bones. Anthropometric measurements should also include assessment of weight, as well as recent weight changes if applicable, and assessment of linear growth. Various biochemical parameters, such as albumin, transferrin, retinol-binding protein have been used to assist in the assessment of nutritional status.
Breastfeeding remains the gold standard in infant feeding. In recent years, there have been an increase in the number of different types of infant formulas and claims of their efficacy in various conditions such as gastroesophageal reflux, protein allergy, colic and lactose intolerance. Paediatricians and general practitioners have been showered with requests from parents to switch formulas to treat various conditions. In many cases frequent formula changes have occurred which may not be beneficial to the infant. Sometimes even breast-feeding is stopped without justifiable reasons to do so. The purpose of this presentation is to look at some of the available evidence and recommendations for the use of special infant formulas.
SYMPOSIUM 8

IDIOPATHIC NEPHROTIC SYNDROME

S8.1

GENETICS OF NEPHROTIC SYNDROME

Indra Ganesan

Sabah Women & Children’s Hospital (SWACH), Sabah

The recent developments in the genetics of nephrotic syndrome/FSGS have focused on the Glomerular Filtration Barrier as it is the target of injury in these syndromes. The Glomerular Filtration Barrier (GBM) separates the blood from the urinary spaces. It selectively permits the ultrafiltration of water and solutes but prevents leakage of large molecules such as albumin. Glomerular Filtration Barrier consist of inner endothelium, glomerular basement membrane and outer podocyte. Last 3-4 decades, the GBM was thought to play a major role in the pathogenesis of proteinuria. However, in the last 7-9 years, in familial NS & FSGS, the discovery of several genes expressed in the podocytes have shifted to focus from GBM to the podocyte. In the autosomal recessive inheritance, there are congenital nephrotic syndrome of the Finnish type and childhood FSGS which are due defective production of nephrin and podocin respectively. Mutations in WT1 gene which is a tumour suppressor gene results in Denys Drash syndrome. ACTN4, TRPC6 and CD2AP gene defects account for the adulthood FSGS. Understanding the genetic basis of familial Nephrotic syndrome and FSGS has significantly improved our understanding on renal permselectivity. The indication for genetic testing include familial cases of nephrotic syndrome and steroid resistant nephrotic patients who are difficult to treat.
The US Children’s Nephrotic Syndrome Consensus recently published a children’s primary nephrotic syndrome clinical guideline in journal Pediatrics Aug 2009. Their recommendations will be examined in line with our National Paediatric Protocol. Definitive therapy for idiopathic nephrotic syndrome in children is still a trial of steroid at first presentation. Only children who have atypical presentation of nephrotic syndrome or those resistant to a trial of steroid therapy are subjected to renal biopsy. The rational approach to steroid therapy for different presentations of idiopathic nephrotic syndrome (initial presentation, infrequent relapse, frequently relapsing, steroid dependent) and the role of secondary alternatives like alkylating agents, calcineurin inhibitor, levamisole and mycophenolate mofetil will be discussed. The complications of nephrotic syndrome are associated with disease activity and therapy and include infections (an important cause of mortality), thromboembolism, obesity and short stature, acute renal failure, dyslipidemia. Symptomatic management include adequate management of oedema, hypertension and the complications of nephrotic syndrome. Older studies on long-term outcome of children with idiopathic nephrotic syndrome suggested > 90% of children achieve long-term remission without further relapses by puberty. However, this has recently been challenged by surveys indicating a rate of relapse during adulthood as high as 27-42%.
Management of steroid resistant nephrotic syndrome remains a clinical challenge. The clinician has to balance toxicity of medications and unknown long term prognosis. The patients’ response to initial steroid treatment appears to be the best predictor of disease progression. In long term cohort studies of children and adults with primary FSGS renal survival has been directly associated with degree of proteinuria control. Focal segmental glomerulosclerosis (FSGS) is a histologic finding that may result from a variety of insults. The Malaysian Registry of Renal Biopsy reported that FSGS is the histological finding in 25% of renal biopsies in our children over the period 1998 – 2008. FSGS contributed 8% of Malaysian children on dialysis. The lecture focuses on current therapeutic approach toward children with primary FSGS. Therapy of FSGS incorporates conservative and immunosuppressive protocols to control proteinuria.

Corticosteroids
Corticosteroids have been the mainstay of treatment for childhood nephrotic syndrome regardless of its aetiology. A response to corticosteroid is generally consistent with a more favourable response. Corticosteroids remain a key component of many therapeutic regimens for FSGS usually in various combinations with other drugs. A few paediatric protocols have advocated high doses of intravenous methylprednisolone with varying degree of success.

Cyclosporine A
Cyclosporine A and Tacrolimus are both potent calcineurin inhibitors. Activated T cells produce lymphokines such as interleukin 2 that mediate glomerular basement damage. Cyclosporin A acts by preventing full activation of T helper cells. A major concern is the potential for nephrotoxicity. A second concern is the high relapse rate after drug withdrawal.

Tacrolimus
The newer and more potent tacrolimus have been shown in anecdotal reports to have favourable responses in the treatment of children with FSGS. One retrospective study of 16 patients including 13 children with biopsy proven FSGS documented reduction in protein excretion. There were two small prospective studies that also showed a positive response.

Mycophenolate mofetil
Mycophenolate mofetil (MMF) was initially introduced in the 1990s as an immunosuppressive agent for organ transplantation. MMF blocks de novo synthesis of both T and B cell lymphocytes through non-competitive reversible inhibition of the enzyme inosine monophosphate dehydrogenase. Choi et al reported 18 children with FSGS with statistically significant decrease in proteinuria in patients receiving MMF. Catrnan et al reported an open label 6 month trial of MMF in 18 patients with FSGS. All had received various combination of therapy previously. Four out of 18 maintained complete remission. MMF is showing early promise as a steroid sparing therapy but questions remained about length of therapy and long term malignancy risk. MMF is also potentially teratogenic.

Plasmapheresis
Plasmapheresis has been considered as a rescue option. The rationale for its use is for the removal of circulating factor from the plasma that alters glomerular barrier function. It is an invasive procedure with significant risks of hypocalcaemia, infection and bleeding.
Rituximab

Current concepts indicate that not only T cells but B cells are actively involved in the pathogenesis of FSGS. A number of case reports on the use of rituximab on a selected group of children with complicated nephrotic syndrome. Rituximab is a chimeric monoclonal antibody inhibiting CD 20 mediated B cell proliferation and differentiation. All children remained on concomitant treatment with prednisolone and or calcineurin inhibitors. The overall report is rather promising but one must be aware of publication bias as only positive outcome would be reported.

Progress in this therapeutic field remains a priority in an attempt to normalize protein excretion so as to prevent renal failure. The search continues for successful regimens with minimal toxicity.
Cardiac failure is a clinical syndrome where the heart is unable to provide enough output required to meet the metabolic demands of the body. The causes and mechanisms of cardiac failure are significantly different between each cause. The management of congestive heart failure (CHF) is difficult and sometimes dangerous without knowledge of the underlying cause. Consequently, the first priority is acquiring a good understanding of the etiology.

The causes of heart failure other than congenital heart defects are less common in children. The causes would include rheumatic heart disease, cardiomyopathy which could be idiopathic, or secondary to myocarditis of various aetiologies such as viral, bacterial or drugs such as anthracycline. Chronic anaemia, hypocalcaemia, prolonged, recurrent or uncontrolled arrhythmias can also cause heart failure.

Knowing the aetiology will very much help in treating or controlling the heart failure, which include removal of the causative agents or treat the cause. Controlling the arrhythmias could mean controlling the heart failure in cases with persistent arrhythmias.

Medical treatment has not changed much in treating heart failure in children. The main and basic drugs being used are still the diuretics such as loop diuretics and aldosterone antagonists, and angiotensin converting enzyme inhibitors (ACE inhibitors). If the heart failure is associated with hypocontractile ventricles, inotropes such as dopamine and digoxin may have some roles. Digoxin may also act as an anti-arrhythmic. More recent drugs being used in paediatric population are angiotensin receptor blockers (ARBs) and beta-blockers especially for those patients with cardiomyopathy. Of course in certain centres, cardiac transplant is being offered to patients with advanced cardiomyopathy.
SYMPOSIUM 9
CARDIOLOGY

9.2
NATIONAL CONGENITAL HEART DEFECT REGISTRY

Mohd Nizam Mat Bah
Department of Paediatrics, Hospital Sultanah Aminah, Johor

Congenital Heart Disease (CHD) is the commonest congenital malformation seen in children with reported prevalence of 8-10 cases per 1000 live birth. A study on under five deaths in Malaysia in the year 2006 showed that 10% of mortality was directly related to CHD. Despite the significant role of CHD causing mortality, to date, there is limited study in Malaysia described the epidemiology, clinical presentation and outcome of CHD. This presentation will describe the purposes of Malaysian Congenital Heart Disease Registry (MyCHDR). It also describes the inclusion and exclusion criteria for patient selection. Various part of MyCHDR such as CHD diagnosis, CHD intervention and CHD surgery will be discussed briefly. In summary, MyCHDR will function as a tool to improve care of patient with CHD, aid in research, teaching, practice management, physician driven resource utilization and outcome analysis. Therefore, nationwide involvement and contribution is needed to achieve this.
Inborn Errors of Metabolism (IEM) are individually rare but collectively numerous. There are more than 100 IEMs presenting in the newborn period, of which about 20 are amenable to treatment. Despite the advent of newborn screening by tandem mass spectrometry, many IEMs remain undiagnosed. IEMs are classified into three categories: 1. Disorders leading to ‘intoxication’, 2. Disorders involving energy metabolism and 3. Disorders of complex molecules. Clinical illness can present anytime from the newborn period to adulthood. The presence of ketones is pathological in a neonate and highly suggestive of an underlying metabolic aetiology. The key to suspecting IEM is the disparity between the clinical response to appropriate conventional therapy. Adequate diagnostic approach initially is based on the proper use of few routine screening tests. Once a diagnosis is suspected, it is imperative that emergency treatment is commenced while awaiting diagnostic confirmation. A good clinical outcome is dependent on the early recognition and timely commencement of IEM treatment.
Long Term Care and Management of Genetic and Metabolic Disorders: What Role Can the Pediatrician Play

Shanti Balasubramaniam

Institute Pediatrics, Hospital Kuala Lumpur, W.P

The intent of this statement is to describe the breadth of issues that have special pertinence to pediatricians who care for children and families affected by genetic disorders and other chronic health conditions. Early diagnosis, excellent dietary management, biochemical interventions, and good general medical care have begun to erase the previous limits, with a steadily increasing number of patients leading healthier and longer lives. Pediatricians are the more highly qualified to serve them, by virtue of their training and experience, and to provide them a “medical home.” This statement is designed to assist pediatricians in the treatment of these patients by describing their potential roles in relationship to their patients’ changing needs, as they work with various members of the health care team. Pediatricians have a critical role in diagnosis, interdisciplinary planning, acute care delivery, and long-term treatment of children and adolescents with genetic disorders and other chronic health conditions. During the last decade we have witnessed dramatic changes in the diagnosis and treatment of genetic disorders in children that have fundamentally altered the way pediatricians view such conditions. Using the latest scientific information, pediatricians are in the unique position of assisting children who have genetic conditions to reach their full potential. This goal is best achieved by facilitating the integration of the child and family into the community while minimizing the effects of the genetic condition on the child’s overall growth and development. Individuals with genetic and other chronic health conditions and their families confront a seemingly endless series of stressors in their daily lives which may burden families emotionally, socially, and financially. Pediatricians have a key role in the provision of family-oriented, community-based services that recognize the need for the provision of continued multidisciplinary care that will mitigate the adverse long-term physical, developmental, educational, and psychosocial consequences of genetic and other chronic conditions. Pediatricians must stay abreast of the impressive scientific advances that have been made in genetic services while retaining their perspective on the art of the daily practice of medicine to meet the needs of children with genetic or other chronic health conditions and the needs of their families.
The multi ethnic, multi cultural and multi religious society of Malaysia poses a challenge for the clinical genetics practice in this country. Ethical issues ranging from prenatal genetic diagnosis to withdrawal of treatment and end of life issues are often unique and specific to the country. How these issues affect the progress of clinical genetics practice in the country and how they are managed will be discussed during the lecture.
Bedwetting or nocturnal enuresis is common. The prevalence in Malaysia has been shown to be similar to that of the West. It is often classified as primary and secondary forms but modern research indicates that the classification into monosymptomatic (MNE) - with no appreciable daytime voiding symptoms and non-monosymptomatic (NMNE) is clinically more appropriate. The majority of children have at least one subtle daytime symptom thus making NMNE the more common type. Three major pathogenetic mechanisms have been established as crucial in these children. These are nocturnal polyuria, small bladder capacity +/- detrusor overactivity and an increased arousal threshold. Co-morbid conditions such as constipation, obesity with obstructive sleep apnoea and neuropsychiatric disorders such as attention deficit hyperactivity disorder contribute and potentially lead to therapy resistance. The mainstay of the evaluation of a child with enuresis is a good case history with focus on voiding habits and a simple physical examination together with an understanding of the familial tolerance and child’s motivational level. Minimal investigations (if any) are required in the majority of patients. Primary therapy should include education and advice on general lifestyle in particular daily fluid intake as well as achieving good bowel and bladder habits and specific treatments such as enuretic alarms for small bladder capacity and /or desmopressin for polyuric forms. Expected success rates are 60-80%. Therapy resistant cases should be evaluated and handled by a specialist in the field as they may require second line medications and /or combination therapy that include anticholinergic and tricyclic medications. Results of other complementary therapies (hypnosis, acupuncture, homeopathy) are currently not validated. Although the majority of children will outgrow the problem, studies confirm that enuresis has an impact on a child’s self esteem hence treatment is indeed justified.
SYMPOSIUM 11
VOIDING DISORDERS
S11.2
DYSFUNCTIONAL VOIDERS- AETIOLOGY AND ASSESSMENT
Susan Woo
Department of Urology, Hospital Kuala Lumpur, W.P

Dysfunctional voiding is one of the commonest conditions described under the category of non neurogenic voiding dysfunction. Although a common condition, it is often underappreciated by primary care-givers. The aetiology is multifactorial. Children commonly present with urinary incontinence and recurrent urinary tract infections. The association of dysfunctional voiding, recurrent urinary tract infections and vesicoureteral reflux is well known. It is also important to exclude associated constipation as part of the dysfunctional elimination syndrome. The aim of assessment is to determine the diagnosis and exclude neurogenic causes in order to plan subsequent management. Initial evaluation includes detailed voiding and bowel history, physical examination, bladder diary, urinalysis, urinary flow and post-void residual volumes evaluation and genitourinary ultrasound. More invasive investigations such as micturating cystourethrogram, urodynamics and cystoscopy may be indicated in complicated urinary tract infections or those who fail conventional therapy.
SYMPOSIUM 12

NEUROLOGY

S12.1

APPROACH TO THE FIRST UNPROVOKED SEIZURE

Vigneswari

Penang Hospital, Penang

The International League Against Epilepsy defines the FIRST seizure as ‘one or multiple seizures within 24 hours with recovery of consciousness between seizures’. About 5% of paediatric medical emergencies in the UK are due to a first seizure. A first seizure has been reported to be a very stressful and worrying experience for families. The first afebrile seizure may be a non-epileptic event or an acute symptomatic seizure or an unprovoked epileptic seizure. A practical approach to managing the first seizure would be to identify the possible underlying aetiology, as this would subsequently guide therapy, as well as counseling about prognosis and risk of seizure recurrence. A first seizure that is acute symptomatic or provoked is unlikely to recur (recurrence risk ~ 3-10%). Meta-analyses studies show that there is about 30-50% risk of recurrence after occurrence of a first unprovoked seizure and 70-80% recurrence after the second unprovoked seizure. This lecture will highlight the important differential diagnosis that can mimic seizures, the relevant investigations required, especially the indications for EEG and neuroimaging in the first unprovoked seizure. Evidence-based practice recommendations on decision to commence treatment after the first unprovoked seizure would also be presented.
Headaches are among the top 5 health problems in childhood. Among 7 year old Finnish children prevalence rose from 14% in 1974 to 52% in 1996. In a national survey, 26.6% of Canadian 12-13 yrs old and 31.2% of 14-15 yrs old children had experienced a headache at least once in the previous week. In Malaysia headache related complains constitute 6% of all new referrals in local pediatric neurology services. The majority of these are primary headache like migraine and tension type headaches. However in the emergency situation it is important to exclude secondary headaches due to intracranial pathology. This can be done confidently by a systematic assessment of the headache and a full neurological examination. Only a small number of children with headache will benefit from neuroimaging. Management of primary headaches should start with non drug approaches. Only a few drugs are of proven value in the prophylactic treatment of primary headaches and they will benefit children with troublesome symptoms.
A child who presents with an abnormal gait may have an underlying musculoskeletal or neurological disorder. Acute painful and limping gait often is due to orthopaedic or rheumatological problem. However, for a child who has a progressive gait difficulty, neurological or neuromuscular disorders are more likely. Observational gait analysis remains clinically useful in this situation. However, one needs to be systematic in the assessment and be aware of the potential pitfalls in order to arrive at the correct diagnosis or differential diagnosis. The common gait problems due to neurological causes are hemiplegic, diplegic, waddling, high stepping and ataxic gaits. A number of video clips will be used as illustrations. Toe walking could be due to spasticity or contracture of the Achilles tendons but may also be behavioural in nature. Ataxic gait may be due to peripheral neuropathies or cerebellar dysfunction. Waddling gait and Gower’s sign indicate proximal muscle weakness and is often due to muscular dystrophy or myopathies. However, one needs to be aware that they may also occur in spinal muscular atrophy Type III as well as treatable condition such as dermatomyositis. High stepping gait with absent tendon reflexes often points towards a neuropathic condition which may be hereditary or acquired. Careful physical examination could often differentiate the various groups of neuromuscular disorders. Further investigations into these various conditions will be mentioned briefly in the presentation.
There are various pathologies that have been described to cause airway distress in the pediatric age group. Our overview of the various pathologies encountered at the ORL Department, Hospital Sultanah Bahiyah, Alor Setar under the management of the Peds ORL team and Paediatric department are described. The aetiological causes of upper airway obstruction most frequently cited are foreign bodies, trauma, infections, severe laryngomalacia, laryngeal cysts, laryngeal papilloma, subglottic stenosis, tracheoesophageal fistula, laryngeal cleft, choanal atresia, congenital pyriform aperture, neuromuscular deficiency which including drooling of saliva. A proportion of the surgical airway conditions management can be addressed endoscopically and the rest via open surgical technique. The topic will display clinical and endoscopic evaluation followed by the surgical treatment in the presenter’s clinical setting.
MANAGEMENT OF EXTERNAL EAR ANOMALIES: CONGENITAL MICROTIA AND CANAL ATRESIA

Goh Bee See, Asma Abdullah, Mazita Ami, Lokman Saim

Department of Otorhinolaryngology, Faculty of Medicine, Universiti Kebangsaan Malaysia

External ear anomalies in children or microtia occur once in about every 7,000 to 8,000 births in the general population. And it is fairly common for microtia to be associated with other congenital abnormalities. Among associated malformations, facial cleft and cardiac defects are the most common followed by anophthalmia or microphthalmia, limb reduction defects, renal malformation, and holoprosencephaly. Congenital aural atresia is a birth defect characterized by hypoplasia of the external auditory canal and often associated with microtia or dysmorphic features of the auricle and middle ear with occasional abnormalities of inner ear structures. Parents of newborn with microtia and congenital aural atresia often develop great anxiety. Therefore it is important for clinician to provide proper counseling and realistic plan of management. Initial management of microtia and congenital aural atresia should be directed towards determining the auditory function rather than cosmesis of the abnormal pinna. In unilateral cases, if the hearing in the contralateral ear is normal, these children usually develop normal speech and normal intellectual growth without any surgical intervention. For bilateral cases, the audiologist should manage these children from the beginning. Amplification by bone conduction hearing aid should start as early as possible. It is important to be selective in consideration for microtia surgery and canal reconstruction. High resolution CT scan of the temporal bone is important in patient selection for surgery. We limit canal reconstruction to ears with normal cochlear function and normal or near-normal pneumatization of the tympanic cavity. Good hearing results can be achieved by removing most or all of the bony atresia. The effectiveness of skin grafting after the bony canal has been drilled is the most important factor in the success of canal reconstruction. Facial asymmetry due to abnormal facial and temporal bone growth is a relative contraindication for canal reconstruction. There is a higher risk of facial nerve injury and poor hearing outcome. Pinna reconstruction may be performed without canal reconstruction in these cases. At the Department of Otorhinolaryngology, Universiti Kebangsaan Malaysia Medical Centre, canal and pinna reconstruction is usually performed after the age of 10 years except for cases of canal cholesteatoma when surgery is performed earlier. Management of congenital microtia and canal atresia needs full evaluation by ORL surgeon, audiologist, paediatrician and may be psychologist for proper counseling of realistic plan and optimize the functional outcome.
UNIVERSAL VERSUS HIGH RISK NEONATAL HEARING SCREENING AND RELEVANCE TO REHAB AND COCHLEAR IMPLANT PROGRAMME

Siti Sabzah Mohd Hashim¹, Norzi Gazali²

1. Department of Otorhinolaryngology, Hospital Sultanah Bahiyah, Kedah
2. Department of Otorhinolaryngology, Hospital Sultanah Bahiyah, Kedah

WHO has defined Hearing Impairment as one of the major public Health and social problems and estimates that there are 250 million people worldwide who are deaf and have impaired hearing at birth (WHO report 2005). In Malaysia, The National Hearing Disorder survey 2005 displayed a prevalence of 17.4% with an estimated population of 3,962,879. In the Disease Burden Report 2000 (Ministry of Health Statistics based on the total DALY status and rank order) hearing related problems is one of the top ten reported diseases in Malaysia. Permanent Congenital Hearing Loss is one of the most frequent congenital abnormalities at birth. It is cited as 1-4 in 1000 life birth. Screening is therefore the avenue through which access to quality intervention is made available. Universal and High Risk Newborn hearing Screening (UNHS) has been introduced in many countries in order to allow early diagnosis and intervention to congenital hearing impairment. It has been proven worldwide in established studies that early intervention in hearing impairment children will improve language outcomes and subsequent school and occupational performance. Screening will ensure that the age of identification of hearing loss is reduced and the outcome of intervention is better. Implementation of hearing screening requires responses and collaboration from a myriads of diverse group; ORL, Paediatricians, Obstetricians, Family Physicians, Public Health Specialists, Geneticist, Administrators, Politicians, the Hearing Loss Communities and the General public. In the Ministry of Health Hospitals, Universal Hearing Screening (Screening all newborn) is long due. High risk hearing screening (hearing loss potential children with identified risk factors) is available in some hospitals. High risk Screening only support 50% of detection of overall congenital hearing loss in children. Screening programme is therefore a pressing need and relevant in the Malaysian Healthcare settings in order to support the existing hearing loss burden in Malaysia. This programme will provide positive impact to the outcome of the existing hearing rehabilitation services as well as the Cochlear Implant Programme in Malaysia.
Congenital hearing loss needs early diagnosis and intervention. The aim of management of these children is to improve and optimize the hearing so that they will have normal speech and language development. The two most common congenital defects that can be managed surgically are congenital sensorineural deafness and congenital aural atresia. Congenital profound sensorineural loss can be treated by cochlear implantation after failure of hearing aid trial. The success of cochlear implant does not rely on surgical technique alone but the team work and commitment of the parents. The bone conduction hearing aid is a useful alternative to patients who need hearing rehabilitation but are unable to wear an air conduction hearing aid. It can also be effectively utilised by patients who are not suitable surgical candidates for correction of their deficits. A bone-anchored hearing aid (BAHA) or Baha implant is defined as a hearing aid with percutaneous transmission of sound vibrations to the skull. The use of Baha implant gives superior audiometric results when compared to the conventional bone conduction hearing aid. Percutaneous transmission is 10–15 dB more efficient than transcutaneous transmission; however, Baha has to be surgically implanted to obtain skin penetrating coupling. The UKM Cochlear Implant Programme has performed more than 250 cochlear implant surgeries since 1995. Our hospital also pioneered Baha surgery in Malaysia started in the year of 2000. We have a total of 30 patients using Baha system for hearing rehabilitation. The overall mean functional gain was 34dB. The surgical procedure will be described and its complication were reviewed and discussed. Canalplasty is also an option for canal atresia but not widely recommended as the risk of restenosis is high. Our centre still performs canalplasty in the case of favourable middle ear on HRCT and hence provide alternative for rehabilitation in canal atresia. The outcome of this programme will be presented.
Atopy refers to the genetic predisposition to develop allergic diseases (such as asthma, eczema, food allergy and allergic rhinitis) involving the capacity to produce IgE in response to common environmental proteins like house dust mite, grass pollen, and food allergens. Therefore, primary prevention of atopy refers to the prevention of immunological sensitisation, that is, the development of IgE antibodies (to be distinguished from secondary prevention of an allergic disease following sensitisation and tertiary prevention which is the treatment of the disease). And primary prevention of asthma involves reduction in asthma incidence by identifying and eliminating risk factors (versus secondary prevention of asthma which is the alleviation of established disease and involves disease detection, management, and control and tertiary prevention, the reduction of disease complications). Primary prevention needs to be based on consistent definitions and phenotypic classifications. The polymorphic nature of the disease and evolving definitions make it difficult to assess the effectiveness of various interventions. Currently available studies are largely epidemiological and evidence-based recommendations are hampered by a lack of blinded interventional studies. Disease prevention requires an understanding of the cause(s) of the disease. Many factors, usually on a background of genetic predisposition, have been implicated in causing asthma. Exposure to aeroallergens like house dust mite induces production of IgE in predisposed individuals and high IgE levels are linked with higher incidence of asthma. Timing and load of exposure are also important factors in preventing or increasing sensitization. There seems to be a window of opportunity for both sensitisation and tolerance. Various levels and times of exposure to pet dander and cockroach have also been studied. Cigarette smoke should be avoided. The hygiene hypothesis based on epidemiological studies launched a gamut of studies exploring, amongst other things, endotoxin exposure, early infections, antibiotics, immunisations and anti-pyretics.

Diet is another important area of intervention. Breast feeding exclusively for at least four months together with weaning between 4-6 months is currently recommended. However, maternal diet and dietary avoidance in breastfeeding mothers is no longer recommended as a means of preventing allergy in the child. If breast feeding is not possible hypoallergenic formula may be useful especially in those babies at high risk. Other modalities like prebiotics and probiotics and pharmacological intervention may have a role in prevention. Any intervention recommended must be practical and cost-effective. Selective prevention focusing on groups whose risk of developing asthma is above average would be more fruitful than universal prevention. The timing of these preventive strategies is crucial. The truth is out there – but it is an evolving truth.
SYMPOSIUM 14

ALLERGY IN CHILDREN

S14.2

FOOD ALLERGY IN CHILDREN: WHAT CHOICE DO WE HAVE?

Sabeera Begum Bt Kader Ibrahim

Institute of Paediatrics, Hospital Kuala Lumpur, W.P

Food allergy is defined as an abnormal immunological reaction to food proteins, which causes an adverse clinical reaction. Food allergies have increased significantly in the past decade. An accurate history is crucial in approaching the management. At the outset, food intolerance must be distinguished from food allergies and, furthermore, these allergies should be classified into either an IgE, Non-IgE, or a mixed response. The clinical features vary from life-threatening anaphylaxis to milder IgE-mediated responses, atopic dermatitis, and gastrointestinal symptoms. The severity of the reaction and the potential risk for anaphylaxis on reexposure should be assessed. Milk, soy, egg, wheat, and peanut allergies are common in children, whereas peanut, tree nut, fish, shell fish allergies, and allergies to fruits and vegetables are common in adults. Structural proteins are important determinants of the severity of the reactions and may often predict the natural history and cross reactivity. The evaluation of a child with suspected food allergy includes detailed medical history, physical examination, screening tests and response to elimination diet and to oral food challenge. None of the screening tests, alone or in combination, can definitely diagnose or exclude it. The main principle of food allergy management is avoidance of the offending antigen. An incorrect diagnosis is likely to result in unnecessary dietary restrictions, which, if prolonged, may adversely affect the child's nutritional status and growth. Majority of the patients outgrow their allergies to milk, soy, egg, and wheat, and some to peanut also, therefore, patients should be periodically reassessed.
THE DRIPPY NOSE: WHAT PAEDIATRICIANS SHOULD KNOW

Norrashidah Abdul Wahab

Department of Paediatrics, Serdang Hospital, Selangor

Millions of people continually suffer from a drippy or stuffy nose with no exception in children. They can have symptoms of nasal congestion as young as in neonatal period. Some of them, the doctors find no sign of colds or allergies that can explained the symptoms. Runny nose or drippy nose or congested nose refers to the same condition in which it is called rhinitis. It refers to the patients presented with persistent nasal discharge or secretion. The onset of such symptoms can be either acute or chronic in duration, intermittent or persistent. Several conditions can cause a drippy nose and most of the time, it is a self-limiting disease. However, further evaluation and treatment are required for recurrent and persistent symptoms of drippy and congested nose especially in children. The causes of drippy nose can be divided into two major components. For many, the cause is non-allergic chronic rhinitis, a little-understood condition that is only now coming under study. Non-allergic rhinitis is a catch-all diagnosis for a hypersensitive nose with no obvious explanation. The second component is allergic rhinitis. There might be a much higher incidence of non-allergic rhinitis than allergic rhinitis, in part because many people with allergies also have overlapping symptoms of a non-allergic nasal disease. The non-allergic rhinitis seems to be common in children as well as in adult. The non-allergic rhinitis causes can be due to exposure to irritants, certain drugs, upper respiratory infections (viral, bacterial), foreign body in the nose, congenital structural abnormality of the nose and gastroesophageal reflux disease. Allergic rhinitis can be divided into seasonal and perennial allergy. Vasomotor rhinitis in response to the breathing dry air is also a common cause of drippy nose in children. Even though there are overlapping symptoms, specific symptoms and signs will be able to differentiate between cold, flu, allergy and non-allergic causes of drippy nose. There is a significant difference in the mechanisms of allergic and non-allergic nasal disease causing a drippy nose by looking at the response towards steroid nasal sprays in eliminating the symptoms. Certain allergic tests will help in determining the possible causes of the symptoms. Parental history of allergic rhinitis in both parents increased the risk of suffering from allergic-like symptoms. There was significant association with concurrent atopy, blood eosinophilia and sensitization to house dust mite. Allergic rhinitis can begin as early as 18 months of life. Oral decongestants, antihistamines and topical steroid sprays may provide temporary relief for patients with severe congestion and who have eosinophils present in the nose. For other patients with very dry, blocked noses, frequent sprays of salt water can help. But for those who more often have a drippy nose, facial tissue, until recently, have been the only solution. Ipratropium bromide, a drug approved for asthmatics, has shown promise in drying up a runny nose. Doctors are also testing atropine, a drug that has long been used in the treatment of digestive and urinary tract disorders. Both drugs appear to inhibit mucous production, but they do not reduce sneezing, congestion or sinus pressure. Treat the GORD if it is the cause of drippy nose. Surgical intervention may be required in structural abnormalities of the nose.