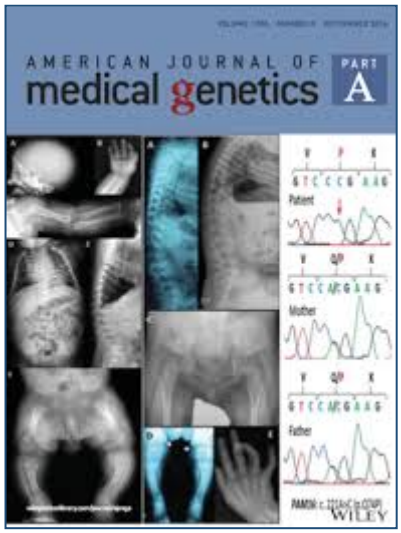
1q21.3 deletion involving GATAD2B: An emerging recurrent microdeletion syndrome



Tim-Aroon T, Jinawath N, Thammachote W, Sinpitak P, Limrungsikul A, Khongkhatithum C, ***Wattanasirichaigoon D.**

***Correspondence**: Duangrurdee Wattanasirichaigoon
Division of Genetics

Am J Med Genet A. 2017;173(3):766-770.

Impact Factor = 2.082

GATAD2B gene is involved in chromatin modification and transcription activity. Loss-of-function mutations of GATAD2B have recently been defined to cause a recognizable syndrome with intellectual disability (ID). Human TPM3 gene encoding thin filament protein is associated with myopathies. Both genes are located on chromosome 1q21.3. We herein report an infant with feeding difficulty, developmental delay, hypotonia, and dysmorphic features including small palpebral fissures, telecanthus, sparse hair and eyebrow, cup-shaped ears, and clinodactyly. Karyotype was normal. Single nucleotide polymorphism array revealed a 1.06 Mb deletion of chromosome 1q21.3, which was confirmed to be de novo. The deleted region encompassed 35 genes, including three known disease-associated genes, namely GATAD2B, TPM3, and HAX1. We further identify and summarize seven additional patients with 1q21.3 microdeletion from literature review and clinical databases (DECIPHER, ISCA/ClinGen). Genomic location analysis of all eight patients revealed different breakpoints and no segmental duplication, indicating that non-homologous end joining is a likely mechanism underlying this particular microdeletion. This data suggests that 1q21.3 microdeletion is a recurrent microdeletion syndrome with distinguishable phenotypes, and loss of function of GATAD2B is the major contributor of the characteristic facies and ID. Additionally, the deletion of TPM3 warrants a risk of concomitant muscle disease in our patient.

Bacteremia during neutropenic episodes in children undergoing hematopoietic stem cell transplantation with ciprofloxacin and penicillin prophylaxis

Choeyprasert W, Hongeng S, Anurathapan U,
***Pakakasama S.**

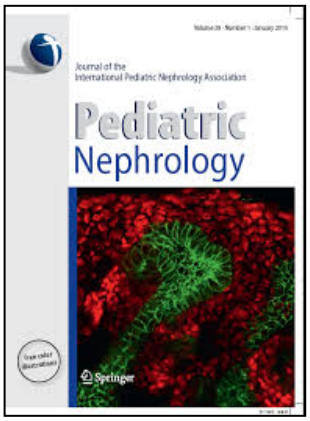
***Correspondence:** Samart Pakakasama
Division of Hematology and Oncology

Int J Hematol. 2017;105(2):213-220.

Impact Factor = 1.846

Bacteremia during neutropenic episodes is a cause of morbidity and mortality in patients undergoing hematopoietic stem cell transplantation (HSCT).We have used oral ciprofloxacin and penicillin V, from the start of the conditioning regimen until engraftment, for the prophylaxis of bacterial infection. The objective of this study was to retrospectively analyze the prevalence of and risk factors for breakthrough bacteremia during neutropenic episodes in autologous and allogeneic HSCT patients. There were 215 patients enrolled, with a median age of 8.32 years (range 0.51-21.64 years) between 2002 and 2014. The common underlying diseases were thalassemia and acute leukemia. Bacteremia was documented in 33 patients (15.3 %), with 39 microorganisms isolated. Escherichia coli (28.2 %) and Streptococcus viridans (12.8 %) were the most commonly isolated Gram-negative and Gram-positive bacteria, respectively. Multidrug resistant strains were found in 32 and 14.3 % of Gram-negative and Gram-positive bacteria, respectively. Risk factors for bacteremia were receiving anti-thymocyte globulin (ATG) [odds ratio (OR) 2.44, 95 % confidence interval (CI) 1.06-5.65, P = 0.037] and umbilical cord blood as a stem cell graft (OR 6.60, 95 % CI 1.04-41.83, P = 0.045). In conclusion, the prevalence of bacteremia was 15.3 % and the use of ATG and cord blood were risk factors for bacteremia during neutropenic episodes.

Blood pressure and heart rate during stress in children born small for gestational age



***Pirojsakul K**, Thanapinyo A, Nuntnarumit P.

***Correspondence:** Kwanchai Pirojsakul

Division of Nephrology

Pediatr Nephrol. 2017 Feb 11. [Epub ahead of print]

Impact Factor = 2.338

BACKGROUND: Increased sympathetic nervous system activity has been proposed as a potential mechanism for the blood pressure (BP) elevation seen in individuals born small for gestational age (SGA). This study was carried out to detect the changes in BP and heart rate (HR) in children born SGA during exposure to stress and to assess for changes in urinary catecholamine excretion.

METHODS: Nineteen children aged 6-14 years born SGA and 17 age- and gender-matched healthy controls were included in the study. The stress test included a mathematical test and venipuncture. BP and HR were monitored during the test. Spot urine samples were collected at baseline and after the stress test to determine dopamine, epinephrine and norepinephrine levels.

RESULTS: At baseline, there was no difference in BP and HR between the SGA and control groups, but mean urinary norepinephrine levels were slightly higher in the SGA group (55.7 ± 16.1 vs. 43.4 ± 3.8 mcg/gCr; P = 0.10). Compared to the control group, mean maximal HR increase was higher in the SGA group (31.3 ± 3.1 vs. 19.2 ± 3.8%; P = 0.008), and mean duration of maximal HR to baseline HR was longer (186 ± 23 vs. 97 ± 13 s, respectively; P = 0.003). There was a significant negative correlation between birth weight and maximal HR increase (r = -0.497, P = 0.003).

CONCLUSION: Children born SGA showed significantly greater increases in HR and significantly longer periods of tachycardia during exposure to stress than did healthy controls. The rise in HR was inversely correlated with birth weight. These findings suggest that children born SGA have a greater increase in sympathetic response when exposed to stress than do healthy individuals.

Breaking the ice: urine proteomics of medullary sponge kidney disease

***Chutipongtanate S.**

***Correspondence:** Somchai Chutipongtanate

Kidney Int. 2017;91(2):281-283.

Impact Factor = 8.563

Urinary proteomics is a promising tool for biomarker investigation, particularly in complex kidney diseases. Fabris and colleagues report that urinary laminin subunit alpha-2 is a potential diagnostic marker of medullary sponge kidney (MSK) disease by using a label-free quantitative proteomics platform and a clinically compatible enzyme-linked immunosorbent assay. The neglected issue of stone pathogenesis was also evidenced. This commentary discusses several considerations in biomarker validation, and how urinary proteomics breaks new ground in MSK research.

Comparisons of the outcomes between early and late tocilizumab treatment in systemic juvenile idiopathic arthritis

Pacharapakornpong T, Vallibhakara SA, Lerkvaleekul B,

***Vilaiyuk S.**

***Correspondence:** Soamarat Vilaiyuk

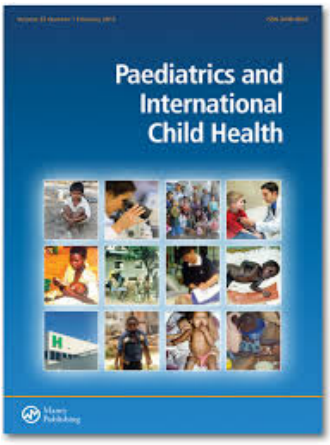
Division of Rheumatology

Rheumatol Int. 2017;37(2):251-255.

Impact Factor = 1.702

Around 40% of systemic juvenile idiopathic arthritis (SJIA) in Thailand is steroid dependent or fails to respond to conventional therapy; therefore, tocilizumab (TCZ), a humanized anti-IL-6 receptor antibody, was indicated in these patients. Due to financial problems, some patients cannot receive TCZ treatment immediately following failure of the conventional treatment occurs, leading to disability and poor quality of life. Therefore, this study focused on the outcomes between early and late TCZ treatment in SJIA patients. This was an observational study. Baseline characteristics and disease severity were collected. Patients were divided into the early TCZ treatment group and the late TCZ treatment group. The outcomes of this study were the remission rates by the end of the study and treatment response using the American College of Rheumatology Pediatric (ACR Pedi) 30, 50, 70 criteria at 3, 6, 9, and 12 months after TCZ initiation. Descriptive analyses were conducted to determine the outcomes. Twenty-three SJIA patients were included in this study. At the end of this study, patients in the early TCZ treatment had a remission rate of 54.5%, whereas none in the late TCZ treatment achieved remission. At the 12-month follow-up, 10 patients (91%) in the early TCZ treatment group and 6 patients (50%) in the late TCZ achieved ACR Pedi 70. The outcomes of TCZ treatment in SJIA patients depend on the time to start TCZ treatment. In the early TCZ treatment, SJIA patients had a higher remission rate and better treatment response than patients who received TCZ treatment late.

Detection of β-lactoglobulin in human breast-milk 7 days after cow milk ingestion



Matangkasombut P, Padungpak S, Thaloengsok S, Kamchaisatian W, Sasisakulporn C, Jotikasthira W, Benjaponpitak S, ***Manuyakorn W**.

***Correspondence:** Wiparat Manuyakorn

Division of Allergy and Immunology

Paediatr Int Child Health. 2017 Feb 22:1-5.[Epub ahead of print]

Impact Factor = 1.103

BACKGROUND: β-lactoglobulin (BLG), a major allergen in cow milk (CM) can be detected in human breast-milk (BM) and is associated with exacerbation of symptoms in breastfed infants with cow milk protein allergy (CMPA). Currently, it is not known how long lactating mothers who consume dairy products need to withhold breastfeeding.

OBJECTIVE: To elucidate the kinetics of BLG in BM after maternal ingestion of a single dose of CM.

METHODS: Nineteen lactating mothers, four of whom had infants with CMPA, were instructed to avoid CM for 7 days before ingesting a single dose of CM and to continue to withhold CM thereafter throughout the study period. BLG was measured by ELISA in BM from 15 mothers of healthy infants before and at 3, 6 and 24 h, and 3 and 7 days after CM ingestion. Four pairs of mothers and CMPA infants were enrolled for BM challenge after the mothers had ingested CM.

RESULTS: After CM ingestion, the level of BLG in BM increased significantly from 0.58 ng/ml (0.58 g/L) (IQR 0.38-0.88) to a peak level of 1.23 ng/ml (IQR 1.03-2.29), $p < 0.001$. The BLG level on day 3 (1.15 ng/ml, IQR 0.89-1.45) and day 7 (1.08 ng/ml (IQR 0.86-1.25) after CM ingestion was significantly higher than baseline ($p = 0.01$ and $p = 0.001$, respectively). BLG was detected in all BM samples from the four mothers of CMPA infants after CM ingestion, and the level was not different from that in the mothers of the 15 healthy infants. Three of the four CMPA infants developed symptoms such as maculopapular rash and hypersecretion in the airways after BM challenge.

CONCLUSIONS: BLG can be detected in BM up to 7 days after CM ingestion. Lactating mothers should suspend breastfeeding to CMPA infants more than 7 days after CM ingestion.

Effect of iron chelation therapy on glucose metabolism in non-transfusion-dependent thalassaemia

***Chuansumrit A,** Pengpis P, Mahachoklertwattana P, Sirachainan N, Poomthavorn P, Sungkarat W, Kadegasem P, Khlairit P, Wongwerawattanakoon P.

***Correspondence:** Ampaiwan Chuansumrit

Division of Hematology and Oncology

Acta Haematol. 2017;137(1):20-26.

Impact Factor = 1.053

AIMS: To compare insulin sensitivity, β-cell function and iron status biomarkers in non-transfusion-dependent thalassaemia (NTDT) with iron excess during pre- and post-iron chelation.

METHODS: Subjects with NTDT, aged older than 10 years, with serum ferritin >300 ng/ml, were included. Iron chelation with deferasirox (10 mg/kg/day) was prescribed daily for 6 months.

RESULTS: Ten patients with a median age of 17.4 years were enrolled. The comparison between pre- and post-chelation demonstrated significantly lower iron load: median serum ferritin (551.4 vs. 486.2 ng/ml, p = 0.047), median TIBC (211.5 vs. 233.5 µg/dl, p = 0.009) and median non-transferrin binding iron (5.5 vs. 1.4 µM, p = 0.005). All patients had a normal oral glucose tolerance test (OGTT) both pre- and post-chelation. However, fasting plasma glucose was significantly reduced after iron chelation (85.0 vs.79.5 mg/dl, p = 0.047). MRI revealed no significant changes of iron accumulation in the heart and liver after chelation, but there was a significantly lower iron load in the pancreas, assessed by higher T2* at post-chelation compared with pre-chelation (41.9 vs. 36.7 ms, p = 0.047). No adverse events were detected.

CONCLUSIONS: A trend towards improving insulin sensitivity and β-cell function as well as a reduced pancreatic iron load was observed following 6 months of iron chelation (TCTR20160523003).

Enhancement of β-globin gene expression in thalassemic IVS2-654 induced pluripotent stem cell-derived erythroid cells by modified U7 snRNA

Phanthong P, *Borwornpinyo S, Kitiyanant N, Jearawiriyapaisarn N, Nuntakarn L, Saetan J, Nualkaew T, Sa-Ngiamsuntorn K, Anurathapan U, Dinnyes A, Kitiyanant Y, ***Hongeng S.**

***Correspondence:** Suradej Hongeng and Suparerk Borwornpinyo
Division of Hematology and Oncology

Stem Cells Transl Med. 2017 Feb 18. [Epub ahead of print]

Impact Factor = 4.247

The therapeutic use of patient-specific induced pluripotent stem cells (iPSCs) is emerging as a potential treatment of β-thalassemia. Ideally, patient-specific iPSCs would be genetically corrected by various approaches to treat β-thalassemia including lentiviral gene transfer, lentivirus-delivered shRNA, and gene editing. These corrected iPSCs would be subsequently differentiated into hematopoietic stem cells and transplanted back into the same patient. In this article, we present a proof of principle study for disease modeling and screening using iPSCs to test the potential use of the modified U7 small nuclear (sn) RNA to correct a splice defect in IVS2-654 β-thalassemia. In this case, the aberration results from a mutation in the human β-globin intron 2 causing an aberrant splicing of β-globin pre-mRNA and preventing synthesis of functional β-globin protein. The iPSCs (derived from mesenchymal stromal cells from a patient with IVS2-654 β-thalassemia/hemoglobin (Hb) E) were transduced with a lentivirus carrying a modified U7 snRNA targeting an IVS2-654 β-globin pre-mRNA in order to restore the correct splicing. Erythroblasts differentiated from the transduced iPSCs expressed high level of correctly spliced β-globin mRNA suggesting that the modified U7 snRNA was expressed and mediated splicing correction of IVS2-654 β-globin pre-mRNA in these cells. Moreover, a less active apoptosis cascade process was observed in the corrected cells at transcription level. This study demonstrated the potential use of a genetically modified U7 snRNA with patient-specific iPSCs for the partial restoration of the aberrant splicing process of β-thalassemia.

Heated humidified high-flow nasal cannula for prevention of extubation failure in preterm infants

Soonsawad S, Swatesutipun B, Limrungsikul A,

***Nuntnarumit P.**

***Correspondence:** Pracha Nuntnarumit

Division of Neonatology

Indian J Pediatr. 2017;84(4):262-266.

Impact Factor = 0.808

OBJECTIVES: To compare extubation failure rate between the heated humidified high-flow nasal cannula (HHHFNC) and continuous positive airway pressure (CPAP) groups.

METHODS: Intubated infants with gestational age (GA) <32 wk, who were ready to extubate, were randomized to receive respiratory support with either CPAP or HHHFNC after extubation. In CPAP group, nasal mask CPAP with preset pressure and fraction of inspired oxygen (FiO_2) equal to positive end-expiratory pressure (PEEP) and FiO_2 of ventilator before extubation was applied. In the HHHFNC group, predefined flow rate according to the protocol was applied. Primary outcome was extubation failure within 72 h after endotracheal tube removal.

RESULTS: Forty-nine infants were enrolled; 24 in the HHHFNC and 25 in the CPAP group. Baseline demographic and respiratory conditions before extubation were similar. There was no difference in infants who met failed extubation criteria between the two groups [8 (33%) in HHHFNC vs. 6 (24%) in CPAP group ($p = 0.47$)]. However, 6 infants (75%) in HHHFNC and 4 infants (66%) in CPAP group who met failed extubation criteria could be rescued by bilevel CPAP. Therefore, the reintubation rate was comparable [2 infants (8.3%) in HHHFNC vs. 2 infants (8%) in CPAP group]. Morbidities or related complications were not different but infants in the HHHFNC group had significantly less nasal trauma (16.7% vs. 44%; $p = 0.03$).

CONCLUSIONS: In the index study, the extubation failure rate was not statistically different between infants who were on HHHFNC or CPAP support.

Incidences, risk factors and outcomes of neonatal thromboembolism

***Sirachainan N**, Limrungsikul A, Chuansumrit A, Nuntnarumit P, Thampratankul L, Wangruangsathit S, Sasanakul W, Kadegasem P.

***Correspondence:** Nongnuch Sirachainan

Division of Hematology and Oncology

J Matern Fetal Neonatal Med. 2017 Feb 9:1-5. [Epub ahead of print]

Impact Factor = 1.674

BACKGROUND: The incidences of thromboembolism (TE) in neonates were reported to be around 0.51 per 10,000 live births per year for overall TE and 24 per 10,000 NICU admissions per year. As the incidences of TE in children and adults are lower in Asian populations, the incidences, risk factors, and outcomes of neonatal TE may be different to those reports from other countries.

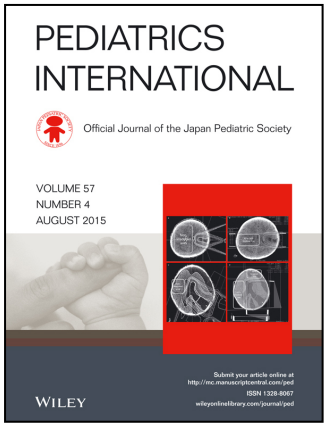
OBJECTIVES: To determine the incidences, risk factors, and outcomes of neonatal TE in a tertiary care hospital in Thailand.

MATERIALS AND METHODS: A retrospective study between the years 1998 and 2015.

RESULTS: From a total of 2463 neonatal admissions, 28 patients were diagnosed with TE. The female/male ratio was 1:1.2. The breakdown of diagnoses of neonatal TE were arterial ischemic stroke (AIS; 36%), arterial TE (ATE; 29%), deep vein thrombosis (DVT; 14%), cerebral venous sinus thrombosis (CVST; 11%), renal vein thrombosis (RVT; 3%), and purpura fulminans (2%). Underlying diseases were identified 57.1% of patients. The most common thrombophilic risk factor was protein C (PC) deficiency (14.3%). The overall mortality rate was 14.3%.

CONCLUSION: The most common TE was AIS. PC deficiency was the most prevalent inherited risk factor, especially in neonates without precipitating factors.

Ischemic stroke in Kawasaki disease



Prangwatanagul W, ***Limsuwan A.**

***Correspondence:** Alisa Limsuwan

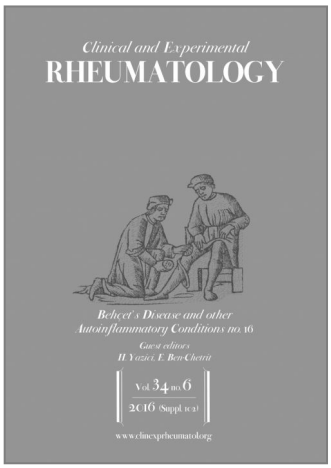
Division of Cardiology

Pediatr Int. 2017;59(1):92-96.

Impact Factor = 0.868

Pediatric stroke is considered to be rare. Stroke resulting from cerebral vasculitis is also uncommon in young children. With the increasing prevalence of Kawasaki disease (KD) diagnosis, this acquired vasculitis has been reported with various clinical presentations including neurological symptoms. Herein we describe the case of a KD patient presenting with stroke. A 15-month-old boy was referred due to stroke that occurred on the fifth day of febrile illness. He was initially admitted to another hospital due to fever and diarrhea. He was discharged and re-admitted 2 days afterward due to left hemiplegia. During the 10 days of the second hospitalization, he had a presumptive diagnosis of encephalomeningitis. Upon referral to the present hospital, he was found to have right middle cerebral artery branch stenosis and fusiform aneurysms of the coronary arteries. Retrospectively, the patient had the full clinical criteria for KD diagnosis. Therefore, stroke could be considered as one of the uncommon clinical manifestations of KD.

Myocardial performance index in active and inactive paediatric systemic lupus erythematosus



***Khositseth A,** Prangwatanagul W,
Tangnararatchakit K, Vilaiyuk S, Su-Angka N.
***Correspondence:** Anant Khositseth
Division of Cardiology and Critical Care Medicine

Clin Exp Rheumatol. 2017;35(2):344-500.
Impact Factor = 2.495

OBJECTIVES: To evaluate cardiac structure and function in paediatric SLE patients without clinical evidence of cardiovascular disease in active and inactive diseases.

METHODS: Patients aged ≤20 years who fulfilled the diagnostic criteria of active SLE underwent transthoracic echocardiography to evaluate cardiac structure and function, and were then followed up echocardiographically every 3-4 months until SLE disease was inactive. Patients with heart failure, myocarditis, pericarditis, endocarditis, coronary artery disease, or abnormal structural heart disease were excluded.

RESULTS: Twenty-six active SLE patients, mean age 13.2±3.3 years, of whom 20 were female (77%), were enrolled. Most patients had cardiac abnormalities especially LV global dysfunction assessed by left ventricular myocardial performance index (LV MPI). LV MPI by conventional method, by tissue Doppler imaging (TDI) at medial and lateral mitral valve annulus were significantly decreased when compared to LV MPI in patients with inactive disease (0.44±0.14 vs. 0.30±0.05, 0.52±0.09 vs. 0.36±0.04, and 0.51±0.09 vs. 0.35±0.05, $p<0.001$). Using receiver operating characteristic, LV MPI cut-off at 0.37, 0.40, and 0.40 by conventional, medial TDI, lateral TDI had sensitivity and specificity of 90% and 84%, 90% and 96%, 90% and 100%, respectively.

CONCLUSIONS: Left ventricular global dysfunction was found to be common in paediatric patients with active SLE. LV MPI by TDI might be useful to diagnose active SLE in paediatric patients.

Nebulized fluticasone for preventing postextubation stridor in intubated children: A randomized, double-blind placebo-controlled trial

Prasertsan P, Nakju D, Lertbunrian R, Chantra M,

***Anantasit N**.

***Correspondence:** Nattachai Anantasit

Division of Critical Care Medicine

Pediatr Crit Care Med. 2017 Mar 7. [Epub ahead of print]

Impact Factor = 2.659

OBJECTIVES: To evaluate the efficacy of nebulized fluticasone propionate in the prevention of postextubation stridor in children.

DESIGN: Double-blind, placebo-controlled randomized clinical trial.

SETTING: PICU in a tertiary referral center.

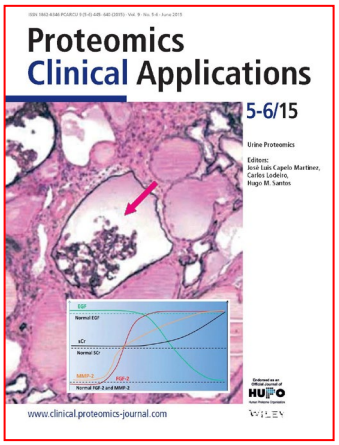
PATIENTS: Children 1 month to 15 years old who underwent mechanical ventilation.

INTERVENTIONS: Patients were randomly assigned into two groups after stratification based on age group receiving nebulized fluticasone 1,000 µg or normal saline solution, immediately after extubation. Vital signs and modified Westley score were evaluated for 6 hours after extubation. The primary outcome was the prevalence of postextubation stridor.

MEASUREMENTS AND MAIN RESULTS: One hundred forty-seven intubated children were enrolled into this study. Baseline characteristics between two groups were not different. There was no significant difference in the incidence of postextubation stridor (12/74 [16%] vs 13/73 [18%]; p = 0.797). However, when analyzing the subgroup of emergently intubated children, the fluticasone group had a longer delay median time for the initiation of noninvasive ventilation than the control group (380 [90-585] vs 60 [42-116] min; p = 0.044). The modified Westley scores at 30 and 60 minutes in the control group were significantly higher than the fluticasone group (4 vs 2, p = 0.04; 4.5 vs 0.5, p = 0.02, respectively).

CONCLUSIONS: The single dose of 1,000-µg nebulized fluticasone did not decrease the prevalence of postextubation stridor. However, it might be beneficial in emergently intubated children**.**

Plasma prefractionation methods for proteomic analysis and perspectives in clinical applications



***Chutipongtanate S,** Chatchen S, Svasti J.

***Correspondence:** Somchai Chutipongtanate

Proteomics Clin Appl. 2017 Feb 14.

[Epub ahead of print]

Impact Factor = 2.959

Plasma is a rich source of biomarkers with clinical relevance. However, the wide dynamic range of protein concentration hinders the detection of low abundance proteins. Plasma prefractionation methods serve as indispensable tools to reduce plasma complexity, allowing the opportunity to explore tissue-derived proteins which leak into the circulation. This review summarizes common approaches in plasma prefractionation methods for proteomic analysis and then discusses some considerations in plasma prefractionation for clinical applications, reviewing some examples of its use in clinical situations.

Randomised controlled trial of sustained lung inflation for resuscitation of preterm infants in the delivery room

Jiravisitkul P, Rattanasiri S,***Nuntnarumit P.**

***Correspondence:** Pracha Nuntnarumit

Division of Neonatology

Resuscitation. 2017 Feb;111:68-73.

Impact Factor = 5.414

AIM: To compare the effects of sustained lung inflation (SLI) vs. standard resuscitation on physiologic responses of preterm infants during resuscitation.

METHODS: Preterm infants (25-32 weeks gestational age) requiring positive-pressure ventilation or continuous positive airway pressure were randomly assigned to either the SLI group (SLI at 25cmH2O for 15s) or Non-SLI group (standard resuscitation alone). The heart rate (HR), oxygen saturation (SpO2), oxygen requirement, and intubation rate in the delivery room were evaluated.

RESULTS: Eighty-one infants were enrolled (SLI group, 43; Non-SLI group, 38). The use of SLI effectively reduced the oxygen requirement. The mean fraction of inspired oxygen 10min after birth was 0.28 (95% CI, 0.26-0.30) in the SLI group and 0.47 (95% CI, 0.43-0.52) in the Non-SLI group ($p<0.001$). During the first 5min, infants in the SLI group trended towards a higher HR and SpO2 than those in the Non-SLI group. The intubation rate in the delivery room was not different between the two groups; however, among infants ≤28 weeks gestational age, the intubation rate was lower in the SLI than Non-SLI group (5 of 17 [29%] vs. 10 of 16 [63%], respectively; $p=0.05$). The duration of respiratory support, survival without bronchopulmonary dysplasia, and the occurrence of pneumothorax were not different between the groups.

CONCLUSION: SLI in infants who require respiratory support appears to be effective in facilitating postnatal transition as determined by HR and SpO2 responses, resulting in less oxygen supplementation. Further studies are needed to confirm the benefits of SLI.

Successful HLA haploidentical HSCT with post-transplant cyclophosphamide in Wiskott-Aldrich syndrome

Kreetapirom P, ***Hongeng S**, ***Manuyakorn W,**
Anurathapan U, Pakakasama S, Sirachainan N,
Andersson BS.

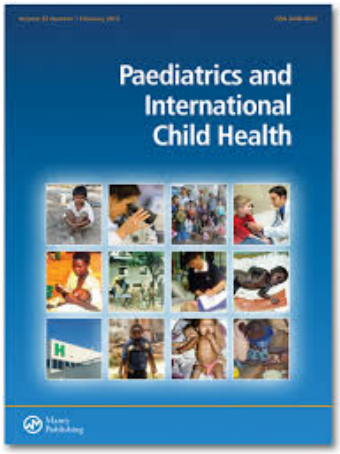
***Correspondence:** Suradej Hongeng and
Wiparat Manuyakorn
Divisions of Hematology and Oncology, and Allergy and Immunology

Bone Marrow Transplant. 2017 Mar 13 [Epub ahead of print]

Impact Factor = 3.636

Allogeneic hematopoietic stem cell transplantation (HSCT) is the only curative treatment for Wiskott–Aldrich syndrome (WAS). The survival rates of transplanted WAS patients using a matched related (MRD) or unrelated donor (MUD) range from 80 to 90%. Without HSCT, the median life expectancy is 15 years. However, the probability of finding a MRD or MUD is only 30 to 60%. Therefore, an alternative related, haploidentical donor should be considered for the patient who lacks a suitable matched donor. From the previous report, haploidentical HSCT in WAS patients yielded an unfavorable outcome. Currently, a haploidentical HSCT (haplo-HSCT) protocol using post-transplant cyclophosphamide (PT-Cy) for GvHD prophylaxis provides a more favorable outcome in patients with malignant and nonmalignant diseases. To the best of our knowledge, we herein report the first successful haplo-HSCT using PT-Cy in a WAS patient.

Testicular enlargement in a pre-pubertal boy with adrenocortical tumour



Korwutthikulrangsri M, Wattanatranon D, Teeraratkul S, Wijarn P, Mahachoklertwattana P, ***Poomthavorn P**.

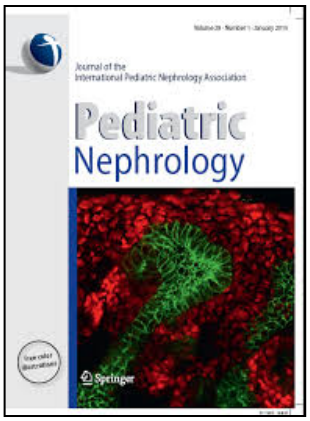
***Correspondence:** Preamrudee Poomthavorn
Division of Endocrinology and Metabolism

Paediatr Int Child Health. 2017 Jan 23:1-3. [Epub ahead of print]

Impact Factor = 1.103

Adrenocortical tumours are rare in children. Virilisation caused by overproduction of adrenal androgens is the most common presentation. The testes of pre-pubertal boys with this tumour are usually small or of pre-pubertal size. A 4.8-year-old boy with an adrenocortical tumour and symmetrical pubertal-sized testes is reported. The serum testosterone level was 204 nmol/L (<0.7), dehydro-epiandrosterone-sulphate 56.7 µmol/L (<1.5) and luteinizing and follicle-stimulating hormones were at suppressed levels. Histology demonstrated a diffusely increased mean tubular diameter of 90 µm (the size in a 12-year-old boy) and hyperplasia of Sertoli cells. There were no Leydig cells in the interstitial area. Prolonged exposure to an extraordinarily high testosterone level could have had stimulating effects on the seminiferous tubules and Sertoli cell growth and thus contributed to testicular enlargement.

Unusual cause of anemia in a child with end-stage renal disease: Answers



Chantarogh S, Kongkhanin U, Thanapinyo A, Saisawat P, Tangnararatchakit K, Chongviriyapan N, Lertthammakiat S, Praditpornsilpa K, ***Pirojsakul K.**

***Correspondence:** Kwanchai Pirojsakul

Division of Nephrology

Pediatr Nephrol. 2017 Mar 2. [Epub ahead of print]

Impact Factor = 2.338

Answers

1. Differential diagnosis for anemia with erythropoietin hyporesponsiveness includes: (1) inadequate dialysis, (2) chronic infection, (3) secondary hyperparathyroidism, (4) bone marrow disorder (e.g., myelodysplastic syndrome), (5) iron deficiency, (6) vitamin and mineral deficiencies (e.g., vitamin B12, folate, copper), (7) lead, zinc, and aluminium toxicity, (8) anti-erythropoietin antibody-associated pure red cell aplasia, and (9) Parvovirus B19 infection

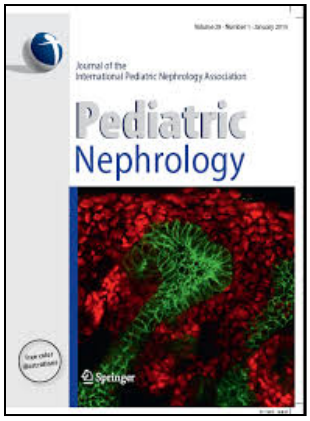
2. Bone marrow biopsy demonstrates erythroid hyperplasia with vacuolization of erythrocytes.

3. As the bone marrow findings were consistent with hypocupremia and serum copper was low at 8 mcg/dl (normal 20–70 mcg/dl), but serum zinc level was elevated at 224 mcg/dl (normal 70–120 mcg/dl), hypocupremia secondary to hyperzincemia was the most likely cause of anemia.

Discussion

Our patient developed anemia with erythropoietin hyporesponsiveness and neutropenia. The investigation showed low plasma levels of copper and ceruloplasmin. Plasma zinc concentration was found to be high. The result of these blood chemistries could confirm the diagnosis of hypocupremia (low plasma copper level) due to hyperzincemia, as other causes were excluded……

Unusual cause of anemia in a child with end-stage renal disease: Questions



Chantarogh S, Kongkhanin U, Thanapinyo A, Saisawat P, Tangnararatchakit K, Chongviriyapan N, Lertthammakiat S, Praditpornsilpa K, ***Pirojsakul K.**

***Correspondence:** Kwanchai Pirojsakul
Division of Nephrology

Pediatr Nephrol. 2017 Mar 2. [Epub ahead of print]
Impact Factor = 2.338

This is the case of a 5-year-old girl diagnosed with end-stage renal disease due to bilateral renal hypoplasia who developed anemia of unknown cause.

The comparisons between thermography and ultrasonography with physical examination for wrist joint assessment in juvenile idiopathic arthritis

Lerkvaleekul B, Jaovisidha S, Sungkarat W,

Chitrapazt N, Fuangfa P, Ruangchaijatuporn T,

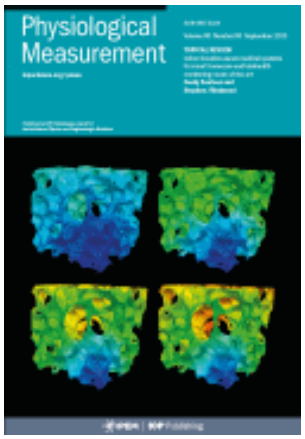
***Vilaiyuk S.**

***Correspondence:** Soamarat Vilaiyuk

Division of Rheumatology

Physiol Meas. 2017 Mar 1. [Epub ahead of print]

Impact Factor = 1.576



This study aimed to assess infrared thermography (IRT) and ultrasonography (US) for detecting wrist arthritis in juvenile idiopathic arthritis (JIA) patients. Although IRT could help us detecting joint inflammation, IRT studies in JIA patients with wrist arthritis are still limited. Currently, no validated US criteria exist for detecting arthritis, and the most useful parameters between Gray-scale ultrasound (GSUS) or Power Doppler ultrasound (PDUS) remain unclear. Therefore, this study focused on detecting wrist arthritis in varying degrees using IRT and US compared with physical examination. Of 46 JIA patients, 16 had previous wrist arthritis but currently inactive, 30 still had wrist arthritis, and the median ages (IQR) were 7.7 (4.3) and 10.2 (4.8) years respectively. Fifteen healthy participants were included, with a median age (IQR) of 9.2 (2.0) years. Using IRT, mean temperature (Tmean) and maximum temperature (Tmax) at skin surface in the region of interest (ROI) in the arthritis group were higher than in the inactive group and the healthy controls with $p < 0.05$. When patients with arthritis were subgroup analyzed by disease severity based on physical examination, the moderate to severe arthritis had Tmean and Tmax higher than the mild arthritis group with statistical significance. The Heat Distribution Index (HDI), two standard deviations of all pixel temperature values in the ROI, in the moderate to severe arthritis group was higher than in the healthy controls ($p = 0.027$). The receiver operating characteristic analysis in arthritis detection revealed diagnostic sensitivity of 85.7% and 71.4% and specificity of 80.0% and 93.3% at a cut-off points of Tmean ≥ 31.0 $^{\circ}$C and Tmax ≥ 32.3 $^{\circ}$C respectively. For US, GSUS and PDUS are useful in detecting arthritis, providing high sensitivity (83.3%) and specificity (81.3%). Our study demonstrated that both IRT and US were applicable tools for detecting wrist arthritis.

An international registry of survivors with Hb Bart's hydrops fetalis syndrome

Songdej D, Babbs C, Higgs DR; BHFS International Consortium

***Correspondence**: Higgs DR

Blood. 2017;129(10):1251-9.

Impact Factor = 13.164

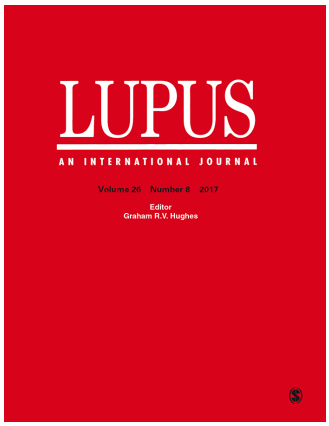
Hemoglobin (Hb) Bart's hydrops fetalis syndrome (BHFS) resulting from α0-thalassemia is considered a universally fatal disorder. However, over the last 3 decades, improvements in intrauterine interventions and perinatal intensive care have resulted in increasing numbers of BHFS survivors. We have initiated an international registry containing information on 69 patients, of which 31 are previously unpublished. In this perspective, we analyze the available clinical information to document the natural history of BHFS. In the future, once we have accrued sufficient cases, we aim to build on this study and provide information to allow counseling of at-risk couples. To date, 39 patients have survived beyond the age of 5 years, 18 of whom are now older than 10 years. Based on the available cases, we find evidence to suggest that intrauterine therapy provides benefits during the perinatal and neonatal period; however,

it may not provide additional benefits to long-term growth and neurodevelopmental outcomes. Growth retardation is a major adverse long-term outcome among BHFS patients with ~40% being severely affected in terms of weight and ~50% in terms of height. There is also an increased risk of neurodevelopmental delay as we find 20% (11/55) of BHFS survivors suffer from a serious delay of ≥6 months. Most patients in the registry require lifelong transfusion and often have associated congenital abnormalities and comorbidities. This perspective is a first step in gathering information to allow provision of informed counseling on the predicted outcomes of affected babies.

Carotid intima-media thickness and arterial stiffness in pediatric systemic lupus erythematosus

Su-Angka N, ***Khositseth A**, Vilaiyuk S, Tangnararatchakit K, Prangwatanagul W.

***Correspondence:** Anant Khositseth
Division of Critical Care Medicine and Cardiology

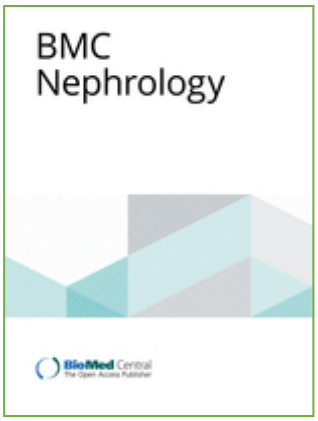


Lupus. 2017;26(9):989-995

Impact Factor = 2.454

Objectives The carotid intima-media thickness (CIMT) and carotid arterial stiffness index (CASI) act as the surrogate markers of atherosclerosis. We aim to assess CIMT and CASI in pediatric systemic lupus erythematosus (SLE). Methods Patients ≤ 20 years old fulfilling diagnostic criteria for SLE were enrolled. Patients with active smoking, coronary heart disease, cerebrovascular disease, arterial thrombosis, family history of hypercholesterolemia, chronic liver disease, or other chronic severe diseases were excluded. The patients were categorized into four groups: active SLE, age- and sex-matched control (control A), inactive SLE, and age- and sex-matched control (control I), according to the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI). All subjects underwent ultrasound of carotid arteries to evaluate CIMT and CASI. Results One hundred and two SLE patients (26 active and 76 inactive) and one hundred and three healthy controls (26 control A and 77 control I) were enrolled. The median CIMT in all groups were not significantly different (0.43, 0.41-0.44; 0.43, 0.41-0.44; 0.42, 0.41-0.43; and 0.42, 0.41-0.43 mm, respectively).The CASI in active SLE (13.5, 11.4-17.3) was significantly higher than in control A (8.2, 7.2-9.2) ($p < 0.0001$), whereas CASI in inactive SLE (12.7, 10.9-15.7) was significantly higher than in control I (8.9, 7.6-9.8). However, the CASI in active and inactive SLE was not significantly different. Conclusions The higher CASI in active and inactive pediatric SLE, implying functional change of carotid arteries, may be early evidence of increased atherosclerosis in pediatric SLE. This functional dysfunction has been found both in inactive and active SLE.

Clinical improvement of renal amyloidosis in a patient with systemic-onset juvenile idiopathic arthritis who received tocilizumab treatment: a case report and literature review



***Chantarogh S**, Vilaiyuk S, Tim-Aroon T, Worawichawong S.

***Correspondence:** Songkiat Chantarogh

Division of Nephrology

BMC Nephrol. 2017 May 12;18(1):159.
Impact Factor = 2.289

BACKGROUND: Juvenile idiopathic arthritis (JIA) is a common rheumatic disease in children and adolescents. Although JIA may cause secondary amyloidosis, this is a rare complication in patients with JIA and other rheumatic diseases. Many previous studies have revealed that common heterozygous or homozygous mutations in the MEFV gene are associated with systemic-onset JIA (SJIA).

CASE PRESENTATION: We herein report a case involving a 19-year-old female patient with difficult-to-control SJIA. She developed progressive proteinuria without clinical signs or symptoms of edema. Renal amyloidosis was diagnosed by renal pathologic examination, which demonstrated deposition of eosinophilic amorphous material in the interlobular arteries, arterioles, and interstitium. Electron microscopy showed fibrillary material deposits with a diameter of 8 to 10 nm. A heterozygous E148Q mutation in the MEFV gene was identified. Conventional disease-modifying anti-rheumatic drugs and etanercept had been used to treat the SJIA, but the disease could not be controlled. Therefore, we decided to start tocilizumab to control the disease activity. However, the patient was unable to receive a standard dose of tocilizumab in the early period of treatment because of socioeconomic limitations. Her disease course was still active, and proteinuria was found. Therefore, tocilizumab was increased to a dose of 8 mg/kg every 2 weeks (standard dose of SJIA), and the patient exhibited a clinical response within 3 months.

CONCLUSION: Refractory SJIA associated with renal amyloidosis is an uncommon cause of proteinuria in adolescents. Tocilizumab may be a beneficial treatment for renal amyloidosis in patients with SJIA.

Clinical outcomes in pediatric renal transplant recipients who received steroid-based immunosuppressive regimen

Chantarogh S, ***Tangnararatchakit K**, Tirapanich W, Viseshsindh W, Saisawat P, Pirojsakul K.

***Correspondence:** Kanchana Tangnararatchakit

Division of Nephrology

Transplant Proc. 2017;49(5):971-6.

Impact Factor = 0.908

BACKGROUND: Although the clinical outcomes of pediatric renal transplantation (RT) in developed countries have improved significantly, the data on clinical outcomes in developing countries are wildly different.

METHODS: Children and adolescents who had undergone RT at Ramathibodi Hospital between March 2001 and August 2014 were included.

RESULTS: Patients were divided into 2 groups: living related donor (LRD) group (n = 13) and deceased donor (DD) group (n = 30). Prolonged cold ischemic time over 13 hours was significantly associated with delayed graft function (P = .029). The prevalence of infection was 90.7%, in which urinary tract infection (UTI) was the most common infection. Although almost none of the patients in the LRD group received induction therapy, the prevalence of rejection was not significantly different between the 2 groups (P = .817). The comparison of graft survivals between LRD and DD groups were 100% vs 100%, 92.3% vs 100%, and 85.7% vs 81.8% at 1, 3, and 5 years, respectively (P = .938). Recurrent UTI and cytomegalovirus (CMV) infection had a negative effect on graft function at 1-year follow-up (P < .05). Rejections, bladder dysfunction, and donors aged ≥50 years were associated with graft deterioration at 3 years after RT (P < .01). None of these patients died with functioning graft.

CONCLUSION: This study demonstrated good graft and patient survival in Thai pediatric RT recipients. Although recurrent UTI and CMV infection were related to graft dysfunction at 1-year follow-up, infections had no effect on graft and patient survival in long-term follow-up.

Emergent literacy in Thai preschoolers: A preliminary study

Yampratoom R, Aroonyadech N, ***Ruangdaraganon N**, Roongpraiwan R, Kositprapa J.
***Correspondence:** Nichara Ruangdaraganon
Division of Developmental and Behavioral Pediatrics

J Dev Behav Pediatr. 2017;38(6):395-400.

Impact Factor = 2.393

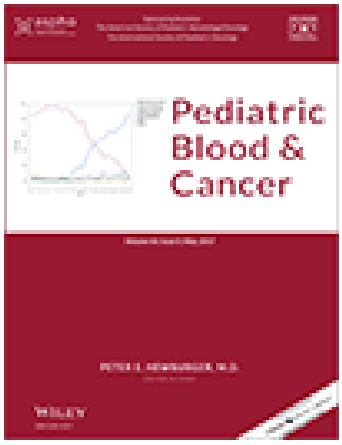
OBJECTIVE: To investigate emergent literacy skills, including phonological awareness when presented with an initial phoneme-matching task and letter knowledge when presented with a letter-naming task in Thai preschoolers, and to identify key factors associated with those skills.

METHODS: Four hundred twelve typically developing children in their final kindergarten year were enrolled in this study. Their emergent reading skills were measured by initial phoneme-matching and letter-naming tasks. Determinant variables, such as parents' education and teachers' perception, were collected by self-report questionnaires.

RESULTS: The mean score of the initial phoneme-matching task was 4.5 (45% of a total of 10 scores). The mean score of the letter-naming task without a picture representing the target letter name was 30.2 (68.6% of a total of 44 scores), which increased to 38.8 (88.2% of a total of 44 scores) in the letter-naming task when a picture representing the target letter name was provided. Both initial phoneme-matching and letter-naming abilities were associated with the mother's education and household income. Letter-naming ability was also influenced by home reading activities and gender.

CONCLUSION: This was a preliminary study into emergent literacy skills of Thai preschoolers. The findings supported the importance of focusing on phonological awareness and phonics, especially in the socioeconomic disadvantaged group.

Genotypes and phenotypes of protein S deficiency in Thai children with thromboembolism



Komwilaisak P, Sasanakul W, Chuansumrit A, Kanjanapongkul S, Wangruangsathit S, Lusawat A, Charoenkwan P, ***Sirachainan N**.

***Correspondence:** Nongnuch Sirachainan

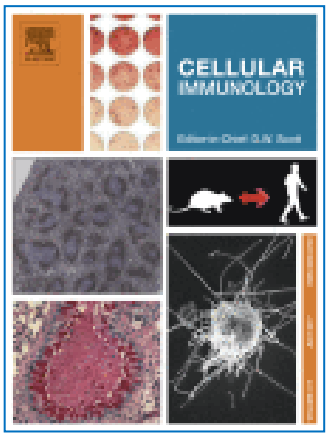
Division of Hematology and Oncology

Pediatr Blood Cancer. 2017;64(5).

Impact Factor = 2.513

The prevalence of protein S (PS) deficiency in Asian patients with venous thromboembolism is around 8-30%, higher than that in Caucasian populations. The present study reports the genotypes (including one novel mutation) and phenotypes of children with PS deficiency at a tertiary care institute. A total of six patients were included, three with arterial ischemic stroke, two with cerebral venous sinus thrombosis, and one with deep vein thrombosis. PS mutations were identified in four patients: p.R355C, p.G336D, p.E67A, and p.N188KfsX9. p.N188KfsX9 is a novel mutation with less than 20% PS activity noted in heterozygotes.

Immune responses to fungal aeroallergen in Heligmosomoides polygyrus-infected mice vary by age



Apiwattanakul N, Palipane M, *Samarasinghe AE

***Correspondence:** Samarasinghe AE

Cell Immunol. 2017;317:26-36.

Impact Factor = 3.172

Parasite infections in the developing world have been considered to promote resistance to immune-mediated diseases such as asthma. Mouse studies have shown that helminths and their products reduce the development of allergic asthma. Since epidemiologic studies that show similar protection are in relation to geohelminth infections that occur in early life, we hypothesized that the parasite-mediated protection against asthma may differ by age. Mice infected with Heligmosomoides polygyrus at 3-weeks of age had similar asthma phenotype compared to mice infected at 28-weeks of age wherein airway eosinophilia was unaltered but tissue inflammation and GC metaplasia were reduced. In contrast, mice infected at 18-weeks of age had elevated macrophagic airway inflammation with accompanying tissue pathology. The presence of γδ T cells and Treg cells in the airways was also regulated by age at worm infection. Our findings demonstrate the importance of age in immune responses that may regulate gut and lung diseases.

Low plasma FVII:C and activated FVII as predictive markers for overt disseminated intravascular coagulation

Lertthammakiat S, Anantasit N, Anurathapan U, Sirachainan N, Kadegasem P, ***Chuansumrit A.**

***Correspondence:** Ampaiwan Chuansumrit

Division of Hematology and Oncology

Thromb Haemost. 2017 May 11. [Epub ahead of print]

Impact Factor = 5.627

In sepsis, binding of factor VII (FVII:C) and activated factor VII (FVIIa) with tissue factor is the key step of coagulation resulting in disseminated intravascular coagulation (DIC). We conducted a prospective cohort study among 47 septic patients, aged 8 months to 18.8 years. They were initially divided into three groups of no DIC (n=27), non-overt DIC (n=14) and overt DIC (n=6). Blood samples were collected at 0, 24 and 48 hours (h) after the onset of sepsis. At the onset of sepsis, FVII:C tended to be lower in the non-overt DIC [median 57 % (interquartile range [IQR] 41-80)] and overt DIC groups [33 % (23-52)] than that in the no DIC group [65 % (44-87)]. Whereas FVIIa tended to be lower in the overt DIC group [1.29 % (0.50-4.19)] than those in the non-overt DIC [3.01 % (1.01-5.24)] and no DIC groups [2.49 % (1.14-3.13)]. At 24 h, FVII:C was significantly lower in the non-overt DIC [57 % (41-101)] and overt DIC groups [31 % (28-49)] than that in the no DIC group [83 % (70-102)]. While FVIIa was significantly lower in the overt DIC group [2.15 % (0.86-3.96)] than that in the no DIC group [3.83 % (2.90-5.46)]. Using FVII:C <65 % or FVIIa <3 % at 24 h among patients without hepatic dysfunction to determine overt DIC at 24 h, the sensitivity was 83.9 % and 77.4 %, respectively, and the specificity was both 83.3 %. Patients with low FVII:C and low FVIIa at 24 h after the onset of sepsis had a 20.8-fold (95 % confidence interval [CI], 2.0-213.0, p=0.010) and 14.4-fold (95 %CI, 1.5-142.4, p=0.023) chance of overt DIC

Mosquito Allergy in Children: Clinical features and limitation of commercially-available diagnostic tests

***Manuyakorn W**, Itsaradisaikul S, Benjaponpitak S, Kamchaisatian W, Sasisakulporn C, Jotikasthira W, Matangkasombut P.

***Correspondence:** Wiparat Manuyakorn

Division of Allergy and Immunology

Asian Pac J Allergy Immunol. 2017 Apr 1. [Epub ahead of print]

Impact Factor = 1.011

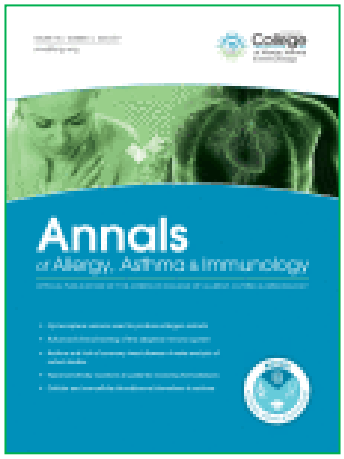
OBJECTIVE: To determine the clinical features of mosquito allergy in children and the ability of commercially available mosquito allergy tests to detect children with mosquito allergy in Thailand.

METHODS: Patients with mosquito allergy aged 1 month to 18 years were recruited. Demographic data, history of mosquito allergy (onset of the reaction, reaction type) and clinical features were recorded. A skin prick test using a commercially available whole body allergen extract from Culex pipiens was performed, and serum was tested for specific IgE antibodies to Aedes communis whole body extract.

RESULTS: A total of 50 patients with mosquito allergy were enrolled. The median age of enrolled children was 6.2 years with an average age of onset of 2 years [interquartile range (IQR) 1-6]. Half of the children were female. The most common skin lesion from mosquito allergy was erythematous papules (n = 45, 76.3%). The majority of children (58%) were in stage 3 (immediate and delayed type of reactions). One child (2%) was in the desensitization stage after 4.6 years of symptoms. The causative mosquito species could be identified only in 26 (52%) children: 16 (32%) childrenwere positive for Aedes communis, 17 (34%) children were positive for Culex pipiens and 7 (14%) children were positive for both Aedes communis and Culex pipiens. Having positive IgE antibodies against Aedes communis was significantly more common in boys (n = 13, 48.1%) than girls (n = 3, 13%) ($p < 0.01$).

CONCLUSIONS: Immediate and delayed skin reaction is the most common manifestation in mosquito allergy children. Commercially available tests for mosquito allergy can detect only 30-50% of children with mosquito allergy.

Optimal needle length for epinephrine prefilled syringe in children



*** Manuyakorn W,** Bamrungchaowkasem B, Ruangwattanapaisarn N, Kamchaisatian W, Benjaponpitak S.

***Correspondence:** Wiparat Manuyakorn

Division of Allergy and Immunology

Ann Allergy Asthma Immunol. 2017;118(6):740-1

Impact Factor = 3.728

Patients with a history of anaphylaxis have a strong indication for the use of epinephrine auto-injectors (EAIs).[1] However, EAIs are available in only 59.1% of countries according to a recent report from the World Allergy Organization. Appropriate EAI needle length for delivering epinephrine into intramuscular tissues has been studied in adults and children. EAIs also require a pressure trigger for delivery of epinephrine, which compresses the subcutaneous tissues and thus shortens the skin-to-muscle depth (STMD). In consequence, studies on needle length for EAIs might not apply to the needle length for syringes prefilled with epinephrine. We recently reported on the stability and sterility of syringes prefilled with epinephrine as alternative treatment for those patients with anaphylaxis. To determine the optimal needle length for a syringe prefilled with epinephrine, we performed a cross-sectional study evaluating STMD and skin-to-bone depth (STBD), which represent the minimum and maximum acceptable needle lengths, respectively.

R147W in PROC gene is a risk factor of thromboembolism in Thai children

***Sirachainan N**, Chuansumrit A, Sasanakul W, Yudhasompop N, Mahaklan L, Vaewpanich J, Charoenkwan P, Kanjanapongkul S, Visudtibhan A, Wongwerawattanakoon P.

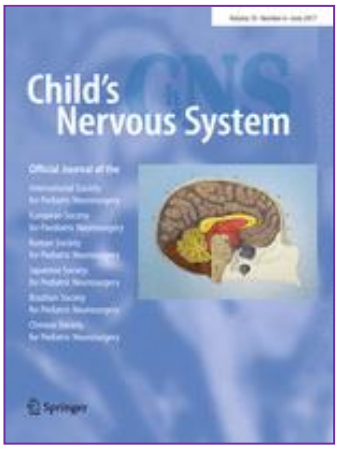
***Correspondence:** Nongnuch Sirachainan
Division of Hematology and Oncology

Clin Appl Thromb Hemost. 2017 Jan 1: [Epub ahead of print]

Impact Factor = 2.096

The p.R147W mutation, the c.C6152T in exon 7, causing a change in amino acid from arginine to tryptophan of the PROC gene has been reported as a common mutation in Taiwanese populations with venous thromboembolism (VTE). The present study aimed to identify the prevalence of p.R147W in the Thai population and children with TE and the risk of developing TE. Patients aged ≤18 years diagnosed with TE were enrolled. The PROC gene was amplified by polymerase chain reaction using a specific primer in exon 7. The restriction fragment length polymorphism was designed using MwoI restriction enzyme. A total of 184 patients and 690 controls were enrolled. The most common diagnosis of TE was arterial ischemic stroke (AIS), at 100 (54.3%), followed by VTE, at 38 (20.6%), and cerebral venous sinus thrombosis (CVST), at 23 (12.5%). The prevalence of heterozygous and homozygous p.R147W in patients and controls was 9.5% versus 5.8% and 2.7% versus 0.1%, respectively. Heterozygous p.R147W had odds ratios (ORs) of 1.8 (95% confidence interval [CI]: 1.0-3.2, P = .04), 3.2 (95% CI: 1.2-8.2, P = .009), and 4.5 (95% CI: 1.6-12.8, P = .002) of developing overall TE, VTE, and CVST, respectively. Homozygous p.R147W had ORs of 20.2 (95% CI: 2.3-173.7, P < .001), 21.4 (95% CI: 2.2-207.9, P < .001), and 43.3 (95% CI: 3.8-490.6, P < .001) of developing overall TE, AIS, and CVST, respectively. This study suggested that p.R147W is a common mutation and increased risk of TE in Thai children.

Reported outcomes of children with newly diagnosed high-grade gliomas treated with nimotuzumab and irinotecan



Sirachainan N, Boongird A, Swangsilpa T, Klaisuban W, Lusawat A, ***Hongeng S.**

***Correspondence:** Suradej Hongeng

Division of Hematology and Oncology

Childs Nerv Syst. 2017;33(6):893-7.

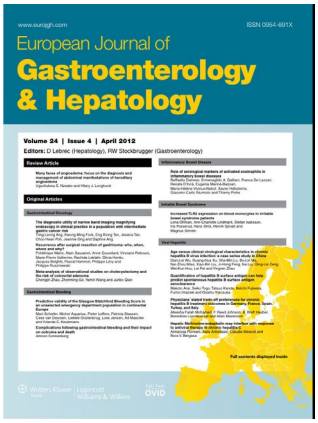
Impact Factor = 1.081

PURPOSE: The outcome of children with high-grade gliomas (HGGs) treated with radiation and adjuvant chemotherapy remains poor. The expression of epidermal growth factor receptor (EGFR) has been established in children with HGGs. This report demonstrated the outcomes of adjuvant nimotuzumab, an EGFR inhibitor, with irinotecan in pediatric HGGs.

METHODS: Children with newly diagnosed HGGs were enrolled. Two weeks after surgery, nimotuzumab with a dose of 150 mg/m2 was given every week during radiation. After completion of radiation, a 4-week cycle of nimotuzumab (150 mg/m2) at week 1 and 3 and irinotecan (125 mg/m2) at week 1, 2, and 3 was given.

RESULTS: Sixteen patients (5 females, 11 males), with a mean ± SD age of 8.2 ± 3.5 years were included. Tumors were located at the supratentorial region (50.0%), infratentorial region (43.8%), and both locations (6.2%). The 5-year PFS and OS were 19.9 ± 11.6 and 31.5 ± 13.0%, respectively. Median times of PFS and OS were 1.8 and 1.9 years, respectively. Prognostic factors related to good outcome were the location of tumor at the supratentorial region or outside brainstem and the extension of surgery. Side effects were minimal, with grade 1 anemia in three patients and diarrhea in one patient. Although, the adjuvant regimen of nimotuzumab and irinotecan slightly increases the overall outcome when compared to the historical study, the advantages of this protocol were minimal side effect, short period of hospitalization, and improved OS in patients who received extensive surgery.

Stool cytomegalovirus polymerase chain reaction for the diagnosis of cytomegalovirus-related gastrointestinal disease



Prachasitthisak N, ***Tanpowpong P,** Lertudomphonwanit C, Treepongkaruna S,
Boonsathorn S, Angkathunyakul N, Sornmayura P, Chantratita W.

***Correspondence:** Pornthep Tanpowpong

Division of Gastroenterology

Eur J Gastroenterol Hepatol. 2017 May 15. [Epub ahead of print]
Impact Factor = 2.093

OBJECTIVES: The diagnosis of cytomegalovirus-related gastrointestinal disease (CMV-GI disease) still requires histopathology, but biopsy is considered invasive. Stool CMV PCR has been reported in adults as an alternative method to diagnose this condition; hence, the results between studies are discrepant. Moreover, no pediatric studies on stool CMV real-time PCR in CMV-GI disease have been carried out. Here, we evaluate the value of stool CMV real-time PCR in detecting CMV-GI disease among immunocompromised children.

METHODS: We enrolled immunocompromised patients aged younger than 20 years who presented with gastrointestinal symptoms at a teaching hospital during January 2015-March 2016. Stool samples were analyzed for CMV real-time PCR. All patients underwent esophagogastroduodenoscopy and colonoscopy with mucosal biopsy.

RESULTS: We performed stool CMV real-time PCR in 31 patients, but two could not undergo endoscopy. Therefore, 29 patients were analyzed. Two additional stool samples showed inhibitors that interfere with the PCR testing and were precluded from the final analysis. Among 27 patients, we found CMV-GI disease in seven (26%) patients. The sensitivity, specificity, and accuracy of stool CMV real-time PCR were 71, 85, and 82%, respectively. We also found that all patients with CMV-GI disease had positive plasma CMV real-time PCR (>150 copies/ml). A significant association between stool and plasma CMV real-time PCR was also noted ($P<0.001$).

CONCLUSION: Stool CMV real-time PCR may be used as a noninvasive tool in the diagnosis of CMV-GI disease. Plasma CMV real-time PCR shows a significant correlation with stool CMV real-time PCR and also represents high diagnostic values.

Efficacy of nasal cellulose powder in the symptomatic treatment of allergic rhinitis: A randomized, double-blind, placebo-controlled trial

***Manuyakorn W**, Klangkalya N, Kamchaisatian W, Benjaponpita S, Sasisakulporn C, Jotikasthira W.

***Correspondence:** Wiparat Manuyakorn

Division of Allergy and Immunology

Allergy Asthma Immunol Res. 2017;9(5):446-52.

Impact Factor = 2.957

PURPOSE: Nasal Cellulose Powder (NCP), which can prevent from binding an allergen to nasal mucosa, may reduce allergic rhinitis (AR) symptoms in dust mite-sensitized children. This study was conducted to assess the efficacy of NCP in improving clinical symptoms of a nasal airflow limitation and the response of nasal inflammatory cells.

METHODS: Children with dust mite-sensitized AR aged 6-18 years were recruited. After a 4-week run-in period, NCP or a placebo was administered, 1 puff per nostril 3 times daily for 4 weeks. The nasal provocation test (NPT) with Dermatophagoides pteronyssinus (Der p) was performed before and after treatment. The daily symptom scores (DSS), daily medication scores (DMS), the peak nasal inspiratory flows (PNIF), nasal airway resistance (NAR), as well as the maximum tolerated dose of NPT and eosinophil counts in nasal scraping, were evaluated.

RESULTS: Sixty children (30 NCP and 30 placebos) were enrolled. Before treatment, there were no significant differences in age, dust mite control measures, DSS, DMS, PNIF, NAR, the maximum tolerated dose of NPT, or nasal eosinophil scores between children receiving NCP and placebos. After treatment, there were no significant differences between the NCP and placebo groups in the median (range) of the outcomes-DSS: 2.06 (0.18-3.77) vs 1.79 (0.08-7.79), P=0.756; DMS: 1.60 (0-5.13) vs 0.56 (0-4.84), P=0.239; PNIF (L/min): 110 (60-160) vs 100 (50-180), P=0.870; NAR (Pa/cm^3/s): 0.40 (0.20-0.97) vs 0.39 (0.24-1.32), P=0.690; the maximum tolerated dose of NPT and the nasal eosinophil scores: 1 (0-4) vs 1 (0-4), P=0.861.

CONCLUSIONS: NCP treatment may not be more effective than placebo treatment in dust mite-sensitized AR children.

Ultrasound versus traditional palpation to guide radial artery cannulation in critically Ill children: A randomized trial

*Anantasit N, Cheeptinnakorntaworn P, Khositseth A, Lertbunrian R, Chantra M.

*Correspondence: Nattachai Anantasit

Division of Critical Care Medicine

J Ultrasound Med. 2017 Jul 8. [Epub ahead of print]

Impact Factor = 1.547

OBJECTIVES: To identify success rates for radial artery cannulation in a pediatric critical care unit using either palpation or ultrasound guidance to cannulate the radial artery.

METHODS: A prospective randomized comparative study of critically ill children who required invasive monitoring in a tertiary referral center was conducted. All patients were randomized by a stratified block of 4 to either ultrasound-guided or traditional palpation radial artery cannulation. The primary outcomes were the first attempt and total success rates.

RESULTS: Eighty-four children were enrolled, with 43 randomized to the palpation technique and 41 to the ultrasound-guided technique. Demographic data between the groups were not significantly different. The total success and first attempt rates for the ultrasound-guided group were significantly higher than those for the palpation group (success ratio, 2.03; 95% confidence interval, 1.13-3.64; P = .018; and success ratio, 4.18; 95% confidence interval, 1.57-11.14; P = .004, respectively). The median time to success for the ultrasound-guided group was significantly shorter than that for the palpation group (3.3 versus 10.4 minutes; P < .001). Cannulation complications were lower in the ultrasound-guided group than the palpation group (12.5% versus 53.3%; P < .001).

CONCLUSIONS: The ultrasound-guided technique could improve the success rate and allow for faster cannulation of radial artery catheterization in critically ill children.

Validation of different pediatric triage systems in the emergency department

Aeimchanbanjong K, ***Pandee U.**

***Correspondence:** Uthen Pandee

Division of Ambulatory Pediatrics

World J Emerg Med. 2017;8(3):223-7.

Impact Factor = N/A

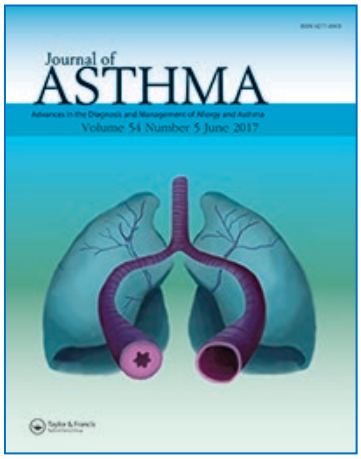
BACKGROUND: Triage system in children seems to be more challenging compared to adults because of their different response to physiological and psychosocial stressors. This study aimed to determine the best triage system in the pediatric emergency department.

METHODS: This was a prospective observational study. This study was divided into two phases. The first phase determined the inter-rater reliability of five triage systems: Manchester Triage System (MTS), Emergency Severity Index (ESI) version 4, Pediatric Canadian Triage and Acuity Scale (CTAS), Australasian Triage Scale (ATS), and Ramathibodi Triage System (RTS) by triage nurses and pediatric residents. In the second phase, to analyze the validity of each triage system, patients were categorized as two groups, i.e., high acuity patients (triage level 1, 2) and low acuity patients (triage level 3, 4, and 5). Then we compared the triage acuity with actual admission.

RESULTS: In phase I, RTS illustrated almost perfect inter-rater reliability with kappa of 1.0 ($P<0.01$). ESI and CTAS illustrated good inter-rater reliability with kappa of 0.8-0.9 ($P<0.01$). Meanwhile, ATS and MTS illustrated moderate to good inter-rater reliability with kappa of 0.5-0.7 ($P<0.01$). In phase II, we included 1 041 participants with average age of 4.7±4.2 years, of which 55% were male and 45% were female. In addition 32% of the participants had underlying diseases, and 123 (11.8%) patients were admitted. We found that ESI illustrated the most appropriate predicting ability for admission with sensitivity of 52%, specificity of 81%, and AUC 0.78 (95%CI 0.74-0.81).

CONCLUSION: RTS illustrated almost perfect inter-rater reliability. Meanwhile, ESI and CTAS illustrated good inter-rater reliability. Finally, ESI illustrated the appropriate validity for triage system

Thai pediatricians' current practice toward childhood asthma



***Kamalaporn H**, Chawalitdamrong P, Preutthipan A.

***Correspondence:** Harutai Kamalaporn

Division of Pulmonology

J Asthma. 2017 Jul 11:1-14.[Epub ahead of print]

Impact Factor = 1.746

BACKGROUND: Childhood asthma is a substantial health burden in Thailand. Due to a lack of pediatric respiratory specialists (pediatric pulmonologists and allergists; RS), most Thai children are cared for by general pediatricians (pediatric primary care providers (PCP)).

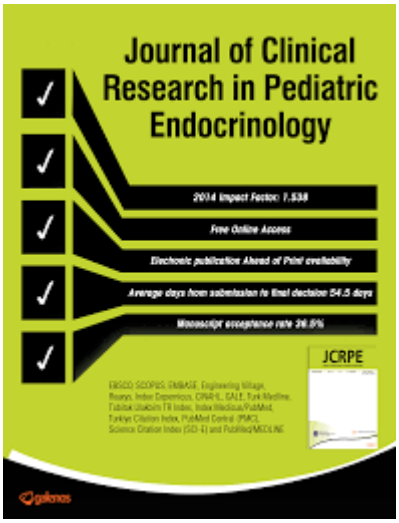
OBJECTIVES: We investigated whether current practices of Thai pediatricians complied with asthma guidelines and compared practices (diagnosis and treatments) provided by PCP and RS.

METHODS: A cross-sectional study was conducted using electronic surveys including four case scenarios of different asthma phenotypes distributed to Thai pediatricians. Asthma diagnosis and management were evaluated for compliance with standard guidelines. The practices of PCP and RS were compared.

RESULTS: From 800 surveys distributed, there were 405 respondents (51%). Most respondents (81%) were PCP, who preferred to use clinical diagnosis rather than laboratory investigations to diagnose asthma. For acute asthmatic attacks, 58% of the pediatricians prescribed a systemic corticosteroid. For uncontrolled asthma, 89% of the pediatricians prescribed at least one controller. For exercise-induced bronchospasm, 55% of the pediatricians chose an inhaled bronchodilator, while 38% chose a leukotriene receptor antagonist (LTRA). For virus-induced wheeze, 40% of the respondents chose an LTRA, while 15% chose inhaled corticosteroids (ICS). PCP prescribed more oral bronchodilators (31% vs. 18%, $p = 0.02$), antibiotics (20% vs. 6%, $p < 0.001$), and antihistamines (13% vs. 0%, $p = 0.02$) than RS for the management of an acute asthmatic attack.

CONCLUSIONS: Most of the Thai pediatricians' practices toward diagnosis and treatment of acute asthmatic attack and uncontrolled asthma conform to the guidelines. PCP prescribed more oral bronchodilators, antibiotics, and antihistamines than RS.

Acute effects of blood transfusion on insulin sensitivity and beta-cell function in children with beta-thalassemia / HbE disease



Wankanit S, Chuansumrit A, Poomthavorn P, Khlairit P, Pongratanakul S, ***Mahachoklertwattana P.**

***Correspondence**: Pat Mahachoklertwattana

Division of Endocrinology and Metabolism

J Clin Res Pediatr Endocrinol. 2017 Jul 24.

[Epub ahead of print]

Impact Factor = 1.118

OBJECTIVE: To assess acute effects of blood transfusion on insulin sensitivity and β-cell function in thalassemia patients.

METHODS: Fifty children and adolescents with β-thalassemia/HbE disease were enrolled in a prospective cohort study. Hemoglobin, serum ferritin and oral glucose tolerance test (OGTT) were performed prior to, and one week after blood transfusion. Insulin sensitivity indices [Homeostatic Model Assessment (HOMA) of Insulin Resistance (HOMA-IR), Whole Body Insulin Sensitivity Index (WBISI)] and β-cell function indices [HOMA of β-cell function (HOMA-β), Insulingenic Index (IGI), and Disposition Index (DI)] were calculated from glucose and insulin levels obtained during the OGTT.

RESULTS: Following blood transfusion, hemoglobin and serum ferritin were significantly increased [8.5 to 10.1 g/dL ($p < 0.001$) and 1764 to 2160 ng/mL ($p < 0.001$), respectively]. β-Cell function indices were significantly increased [median HOMA-β: 74.3 vs. 82.7 ($p = 0.033$); median IGI: 59.6 vs. 79.3 ($p = 0.003$); median DI: 658 vs. 794 ($p = 0.01$)]. However, insulin sensitivity index (WBISI) tended to decrease and insulin resistance index (HOMA-IR) tended to increase, but with no significance. Multivariate analysis showed that pre-transfused serum ferritin was the major factor negatively associated with WBISI and positively associated with HOMA-IR, but pre-transfused hemoglobin had no significant association with insulin sensitivity indices after transfusion.

CONCLUSION: This study demonstrated that acute increases in serum ferritin and hemoglobin following blood transfusion in patients with thalassemia may contribute to an increase in insulin secretion and a trend towards increasing insulin resistance.

Exome sequencing for simultaneous mutation screening in children with hemophagocytic lymphohistiocytosis

Mukda E, Trachoo O, Pasomsub E, Tiyasirichokchai R, Iemwimangsa N, Sosothikul D, Chantratita W,
***Pakakasama S.**

***Correspondence:** Samart Pakakasama

Division of Hematology and Oncology

Int J Hematol. 2017;106(2):282-90.

Impact Factor = 1.610

In the present study, we used exome sequencing to analyze PRF1, UNC13D, STX11, and STXBP2, as well as genes associated with primary immunodeficiency disease (RAB27A, LYST, AP3B1, SH2D1A, ITK, CD27, XIAP, and MAGT1) in Thai children with hemophagocytic lymphohistiocytosis (HLH). We performed mutation analysis of HLH-associated genes in 25 Thai children using an exome sequencing method. Genetic variations found within these target genes were compared to exome sequencing data from 133 healthy individuals. Variants identified with minor allele frequencies <5% and novel mutations were confirmed using Sanger sequencing. Exome sequencing data revealed 101 non-synonymous single nucleotide polymorphisms (SNPs) in all subjects. These SNPs were classified as pathogenic (n = 1), likely pathogenic (n = 16), variant of unknown significance (n = 12), or benign variant (n = 72). Homozygous, compound heterozygous, and double-gene heterozygous variants, involving mutations in PRF1 (n = 3), UNC13D (n = 2), STXBP2 (n = 3), LYST (n = 3), XIAP (n = 2), AP3B1 (n = 1), RAB27A (n = 1), and MAGT1 (n = 1), were demonstrated in 12 patients. Novel mutations were found in most patients in this study. In conclusion, exome sequencing demonstrated the ability to identify rare genetic variants in HLH patients. This method is useful in the detection of mutations in multi-gene associated diseases.

Serum glypican 4 level in obese children and its relation to degree of obesity

Leelalertlauw C, Korwutthikulrangsri M, Mahachoklertwattana P, Chanprasertyothin S, Khlairit P, Pongratanakul S, ***Poomthavorn P.**

***Correspondence:** Preamrudee Poomthavorn

Division of Endocrinology and Metabolism

Clin Endocrinol (Oxf). 2017 Jul 26. [Epub ahead of print]

Impact Factor = 3.327

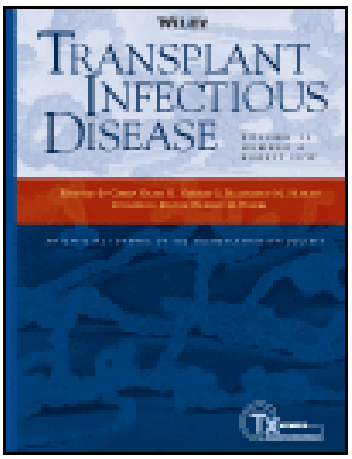
OBJECTIVE: Previous adult studies have demonstrated associations of serum glypican 4 (Gpc4) and obesityparameters and insulin sensitivity. However, an association of serum Gpc4 and glucose metabolism remains contradictory. Study of serum Gpc4 in obese children has not been conducted. We aimed to determine serum Gpc4 levels in obese children with various degrees of obesity.

DESIGN, PATIENTS AND MEASUREMENTS: Up to 370 overweight and obese children, aged 6-18 years were enrolled in this cross-sectional study. Oral glucose tolerance test (OGTT) was performed with fasting serum Gpc4, lipid profiles, aspartate aminotransferase (AST) and alanine aminotransferase (ALT) measured. Insulin sensitivity and β-cell function indices were calculated from plasma glucose and serum insulin levels derived from the OGTT. Bioelectrical impedance analysis was performed for body fat determination. Comparisons of serum Gpc4 levels among the groups of childrenwith various degrees of obesity were performed.

RESULTS: Serum Gpc4 levels progressively increased in children with increasing body mass index standard deviation score (BMI SDS) tertiles [median (interquartile range, IQR): 2.3 (1.8, 3.2), 2.6 (1.9, 3.4) and 3.2 (2.4, 3.8) μg/L, P<.001]. There were no differences in serum Gpc4 levels among children in the different glucose metabolism categories. Log serum Gpc4 levels were positively correlated with SDSs of weight and BMI, cholesterol, AST and ALT. No associations of log serum Gpc4 and insulin sensitivity and β-cell function indices were demonstrated.

CONCLUSIONS: Serum Gpc4 levels were increased with increasing degrees of obesity. There were no differences in serum Gpc4 levels among glucose metabolism categories.

Viral-specific T-cell response in hemorrhagic cystitis after haploidentical donor stem cell transplantation.



Apiwattanakul N, *Hongeng S, Anurathapan U, Pakakasama S, Srisala S, Techasaensiri C, Andersson BS.

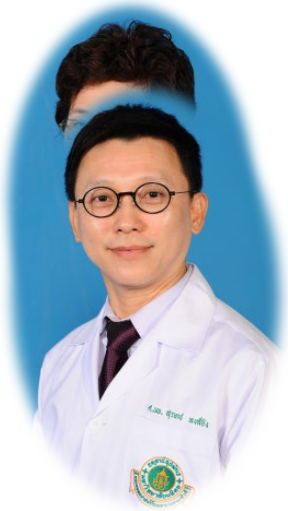
***Correspondence:** Suradej Hongeng

Divisions of Hematology and Oncology

Transpl Infect Dis. 2017 Sep 2. [Epub ahead of print]

Impact Factor = 1.719

Viral hemorrhagic cystitis (HC) after hematopoietic stem cell transplantation (HSCT) can be devastating. Standard treatment modalities have not been well established, but immune reconstitution may be necessary for sustained viral clearance. We studied five pediatric patients who developed viral HC after haplo-identical HSCT. All patients developed virus-specific CD4- and CD8-positive T cells, and the emergence of these viral-specific T cells was temporally associated with successful viral clearance.



Association of factor VIII and factor IX mutations, HLA Class II, tumour necrosis factor-α and interleukin-10 on inhibitor development among Thai haemophilia A and B patients

***Chuansumrit A**, Sasanakul W, Sirachainan N, Kadegasem P, Wongwerawattanakoon P, Mahaklan L, Nathalang O.

***Correspondence:** Ampaiwan Chuansumrit

Division of Hematology and Oncology

Haemophilia. 2017 Sep 11. [Epub ahead of print]

Impact Factor = 3.569

One of the most problematic complications found in patients with haemophilia is the occurrence of inhibitor against the epitopes of the factor VIII and IX genes. The prevalence of factor VIII inhibitor is found among 25%- 30% of patients with severe haemophilia A, while factor IX inhibitor is found among only 2%- 3% of patients with haemophilia B.1 The occurrence of inhibitor creates difficulty in effective bleeding control resulting in higher treatment costs and increased morbidity and mortality. Both genetic and environmental factors have been shown to be involved in inhibitor development.4-7 This study aimed to investigate the association of factor VIII and IX mutations and poly-morphisms of HLA- DRB1, TNF- α and IL- 10 influencing the risk of in-hibitor development among Thai patients with haemophilia A and B.

Development of hepatocyte-like cell derived from human induced pluripotent stem cell as a host for clinically isolated hepatitis C virus

Sa-Ngiamsuntorn K, ***Hongeng S**, Wongkajornsilp A.
***Correspondence**: Suradej Hongeng
Division of Hematology and Oncology

Curr Protoc Stem Cell Biol. 2017;42:4A.13.1-4A.13.34.
Impact Factor = 1.480

This unit describes protocols to develop hepatocyte-like cells (HLCs) starting from mesenchymal stem cells (MSCs) as a natural host for hepatitis C virus (HCV). These include the preparation of MSCs from bone marrow, the reprogramming of MSCs into induced pluripotent stem cells (iPSCs), and the differentiation of iPSCs into HLCs. This unit also incorporates the characterization of the resulting cells at each stage. Another section entails the preparations of HCV. The sources of HCV are either the clinically isolated HCV (HCVser) and the conventional JFH-1 genotype. The last section is the infection protocol coupled with the measurement of viral titer.

Pulse oximetry s ritical congenital heart diseases at hospital settings

***Nuntnarumit P**, Thanomsingh P, Limrungsikul A, Wanitkun S, Sirisopikun T, Ausayapao P.

***Correspondence:** Pracha Nuntnarumit

Division of Neonatology

J Perinatol. 2017 Oct 19. [Epub ahead of print]

Impact Factor = 2.313

OBJECTIVE: To evaluate the predictive abilities of pulse oximetry screening

(POS) for critical congenital heart disease (CRIT.CHD) at two different hospital

settings in Thailand.

STUDY DESIGN: The study was conducted in healthy newborns at Ramathibodi Hospital (RH), a university hospital and Maharat Nakhon Ratchasima Hospital (MH), a regional hospital. Positive POS was defined as oxygen saturation (SpO2) <95% or difference between pre- and postductal SpO2 >3%.

RESULTS: Of 11 407 live births, 10 603 (92.9%) newborns were enrolled with a follow-up rate at 1 month of 78.3%. Incidence of CRIT.CHD (per 1000 live births) at RH and MH were 5.7 and 2.7, respectively. POS could detect three newborns who would have had a missed diagnosis. Sensitivity of POS for CRIT.CHD at RH was 82.3% vs 100% at MH. Overall specificity was 99.9% and false-positive rate was 0.009%. Combination of POS and physical examination (PE) enhanced detection ability to 100% at both hospitals.

CONCLUSION: POS combined with PE improved detection of CRIT.CHD. Routine POS is useful in personnel-limited settings.

Megestrol acetate-caused adrenal insufficiency and delayed puberty in a male adolescent with spinal tumour

Leelalertlauw C, Mahachoklertwattana P, Hongeng S,

***Poomthavorn P.**

***Correspondence:** Preamrudee Poomthavorn

Division of Endocrinology and Metabolism

J Paediatr Child Health. 2017 Oct 12. [Epub ahead of print]

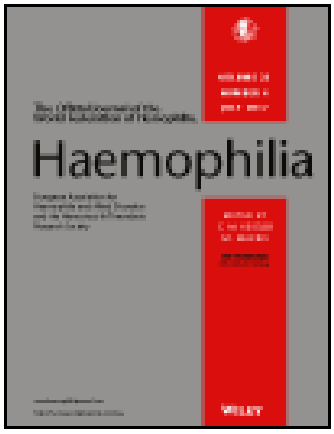
Impact Factor = 1.572

Megestrol acetate (MA), a progestin analogue, has been pre-scribed to promote appetite and weight gain in patients with cancer and acquired immunodeficiency syndrome. MA has been reported to cause endocrine-related problems, including adrenal insufficiency and hypogonadism in adult patients. There were only a few reports of MA-caused adrenal insufficiency and even fewer of MA-caused hypogonadism in children with cancer. We report a case of MA-caused adrenal insufficiency and delayed puberty in a male adolescent with spinal tumour.

Case Report

A 19-year-old male adolescent with epithelioid sarcoma of lum-bar vertebrae presented with fever and shock. The spinal sarcoma had been diagnosed since he was 8 years of age and was treated with surgical resection; chemotherapy, including vincristine, eto-poside, adriamycin, cyclophosphamide, ifosfamide, actinomycin D and melphalan; and radiotherapy. Recurrence of the tumour with metastases had occurred since he was 12 years of age. Palli-ative treatment was commenced because of incurable cancer. Multiple medications, including pregabalin, morphine sulphate (20 mg/day), duloxetine, clonazepam and MA, had been pre-scribed since then. MA at a dose of 160 mg daily was adminis-tered to treat cachexia and promote his appetite. The cumulative dose up until the presentation was approximately 13 440 mg. His past history revealed three previous episodes of shock with ‘uni-dentified cause’ during the past 1.5 years. Shock was successfully treated with fluid resuscitation and inotropic drugs.

Development of a Thai version of the paediatric bleeding assessment tool (Thai paediatric-BAT) suitable for use in children with inherited mucocutaneous bleeding disorders



Pakdeeto S, Natesirinilkul R, Komwilaisak P, Rand ML, Blanchette VS, Vallibhakara SA, ***Sirachainan N**

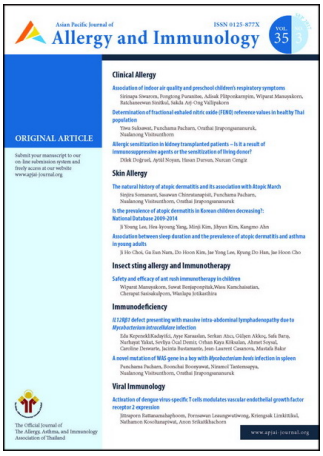
***Correspondence:** Nongnuch Sirachainan
Division of Hematology and Oncology

Haemophilia. 2017;23(6):e539-e542.

Impact Factor = 3.569

Inherited mucocutaneous bleeding disorders consist of various defects of haemostasis such as von Willebrand disease (VWD), and platelet and connective tissue disorders. Due to the subjective nature of re-ported bleeding symptoms, standardized bleeding questionnaires with scores that evaluate the severity of bleeding have been developed, for example the Vicenza and the Molecular and Clinical Markers for the Diagnosis and Management of Type 1 VWD (MCMDM- 1VWD) bleeding questionnaires for the diagnosis of VWD. In children, the Paediatric Bleeding Questionnaire (PBQ) and International Society on Thrombosis and Haemostasis Bleeding Assessment Tool (ISTH-BAT) have been developed and include bleeding symptoms unique to the neonatal period such as cephalohaematoma and umbilical stump bleeding. Both questionnaires have been used to assess bleeding in children with VWD and platelet function disorders.

Needle length for epinephrine prefilled syringes in children and adolescents: Is it one inch?



***Manuyakorn W**, Bamrungchaowkasem B, Ruangwattanapaisarn N, Kamchaisatian W, Benjaponpitak S.

***Correspondence:** Wiparat Manuyakorn
Division of Allergy and Immunology

Asian Pac J Allergy Immunol. 2017 Sep 17. [Epub ahead of print]
Impact Factor = 1.011

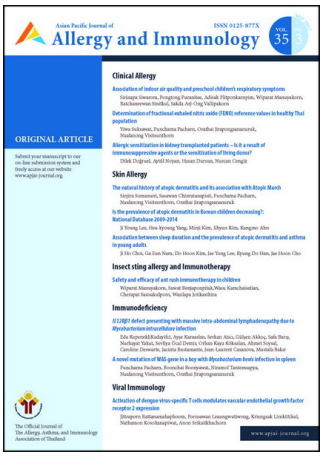
BACKGROUND: Intramuscular epinephrine is the first line drug in the treatment of anaphylaxis. This study was to identify the appropriateness of 1 inch needle length for epinephrine prefilled syringes in children.

METHODS: Children aged 1 month to 18 years were enrolled. Skin to muscle depth (STMD) and skin to bone depth (STBD) were measured using an ultrasonography at the mid-anterolateral thigh. A 1 inch needle was considered as being appropriate if the STBD was more than 1 inch and the STMD was less than 1 inch.

RESULTS: Seventy five infants, 75 pre-school aged children, 75 school aged children and 147 adolescent were enrolled: 196 (52.7%) children were male. A 1 inch needle length was appropriate for 61% of the infants, for 88% of the preschool children, for 99% of the school aged children and for 95% of the adolescents. Thigh circumference ≥23 cm, BMI ≥16 kg/m² and BW ≥6 kg in infants provided the sensitivity of 74%-96% in predicting the appropriateness of 1 inchneedle. In preschool group, thigh circumference ≥25 cm, BMI ≥13.5 kg/m² and BW ≥10 kg provided the sensitivity of 98.5-100% in predicting the appropriateness of 1 inch needle. Thigh circumference ≥49 cm in adolescents provided the sensitivity of 75% in predicting that a 1 inch needle was too short.

CONCLUSION: One inch needle length may not be appropriated for intramuscular injection at thigh in all children. Thigh circumference, BMI and body weight are useful for predictor for using the 1 inch needle.

The effectiveness of newly developed written asthma action plan in improvement of asthma outcome in children



Lakupoch K, Manuyakorn W, Preutthipan A,

***Kamalaporn H.**

***Correspondence:** Harutai Kamalaporn

Division of Pulmonology

Asian Pac J Allergy Immunol. 2017 Sep 17. [Epub ahead of print]
Impact Factor = 1.011

BACKGROUND: Providing asthma education about controller medication use and appropriate management of asthma exacerbation are the keys to improving the disease outcome. Many asthma guidelines recommend that physicians provide written asthma action plan (WAAP) to all of their asthmatic patients. However, the benefit of WAAP is unclear. Thus, we have created a new WAAP which is simplified in Thai and more user friendly.

OBJECTIVE: To determine the effectiveness of the newly developed asthma action plan in management of children with asthma.

METHODS: Asthmatic children who meet inclusion criteria all received the WAAP and they were followed up for 6 months with measurement of outcome variables, such as asthma exacerbation that required emergency room visit, unscheduled OPD visit, admission and school absence in order to compare with the past 6 months before receiving the WAAP.

RESULTS: The analyzed outcomes of forty-nine children show significantly reduced emergency room visit (P-value 0.005), unscheduled OPD visit (P-value 0.046), admission days (P-value 0.026) and school absence days (P-value 0.022). Well controlled group and mild severity group were not the factors that contribute to decreased emergency room visit but step up therapy may be the co-factor to decreased ER visit.

CONCLUSIONS: The results of this study suggest that the provision of newly developed WAAP is useful for improving self-care of asthma patients and reducing asthma exacerbation.

The newly developed screening tool for detection of delayed language development in Thai children: A cross sectional study

***Chuthapisith J**, Wantanakorn P, Roongpraiwan R, Ruangdaraganon N.

***Correspondence:** Jariya Chuthapisith

Division of Developmental and Behavioral Pediatrics

J Med Assoc Thai.2017; 100(10):1081-7

Impact Factor = N/A

Objective: To examine the diagnostic accuracy and validity of the Ramathibodi Language Development (RLD) questionnaire, a new Thai language development screening tool for young children.

Material and Method: The RLD questionnaire was completed by parents of 319 typically developing children, aged 18 to 30 months old. All children were referred to developmental and behavioral pediatricians. The Mullen Scales of Early Learning (Mullen, 1995) was administered to confirm the diagnosis of typically developing children and children with delayed language development.

Results: The cut-off score that best distinguished children with delayed language development and typically developing children was 8, with a sensitivity and specificity of 72.1% and 92%, respectively. The discrimination ability was good, with an area under the curve of 0.82 (95% CI 0.74 to 0.90).

Conclusion: The RLD questionnaire is a promising screening instrument for use to detect children with language development delay early during well-child care visits or at primary care centers.

Investigation of FoxO3 dynamics during erythroblast development in β-thalassemia major

Thanuthanakhun N, Nuntakarn L, Sampattavanich S, Anurathapan U, Phuphanitcharoenkun S, Pornpaiboonstid S, Borwornpinyo S, ***Hongeng S.**

***Correspondence**: Suradej Hongeng

Division of Hematology and Oncology

PLoS One. 2017;12(11):e0187610.

Impact Factor = 2.806

The FoxO3 transcription factor is a key regulator of oxidative stress and erythroid maturation during erythropoiesis. In this study, we explored the involvement of FoxO3 in severe β-thalassemia. Using primary CD34+ hematopoietic progenitor cells from patients with β-thalassemia major, we successfully developed an in vitro model of ineffective erythropoiesis. Based on this model, FoxO3 activity was quantified in single cells using high throughput imaging flow cytometry. This study revealed a significant reduction of FoxO3 activity during the late stage of erythroblast differentiation in β-thalassemia, in contrast to erythropoiesis in normal cells that maintain persistent activation of FoxO3. In agreement with the decreased FoxO3 activity in β-thalassemia, the expression of FoxO3 target genes was also found to decrease, concurrent with elevated phosphorylation of AKT, most clearly at the late stage of erythroid differentiation. Our findings provide further evidence for the involvement of FoxO3 during terminal erythropoiesis and confirm the modulation of the PI3K/AKT pathway as a potential therapeutic strategy for β-thalassemia.

Decreased oxygen exposure during transportation of newborns

***Ruangkit C**, Soonsawad S, Tutchamnong T, Swatesutipun B.*

Correspondence: Chayatat Ruangkit
Division of Neonatology

Arch Dis Child. 2017 Nov 25. [Epub ahead of print]

Impact Factor = 3.265

Oxygen is the most common treatment for newborns in need of respiratory support. However, oxygen can cause tissue injury through reactive oxygen species formation, especially in premature infants with reduced antioxidant defences, and may result in short-term and long-term toxic effects in multiple organ systems. Although most hospitals have the capability to tightly control oxygen delivery to hospitalised neonates, in many circumstances, the need is overlooked during infant transport. Lack of awareness of harm or appropriate medical equipment invariably results in excessive oxygen exposure. We developed a quality improvement programme to decrease oxygen exposure to newborns during their transportation, thus improving patient safety and quality of care

Large-scale proteomics identifies MMP-7 as a sentinel of epithelial injury and of biliary atresia

***Lertudomphonwanit C,** Mourya R, Fei L, Zhang Y, Gutta S, Yang L, Bove KE, Shivakumar P, Bezerra JA.

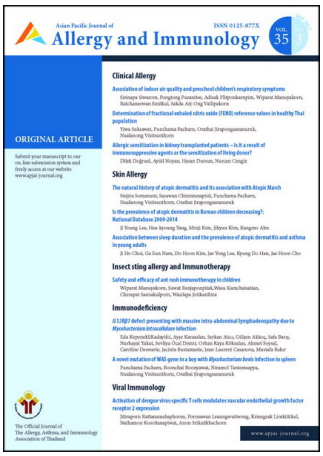
Correspondence: Chatmanee Lertudomphonwanit
Division of Gastroenterology

Sci Transl Med. 2017;9(417). pii: eaan8462

Impact Factor = 16.761

Biliary atresia is a progressive infantile cholangiopathy of complex pathogenesis. Although early diagnosis and surgery are the best predictors of treatment response, current diagnostic approaches are imprecise and time-consuming. We used large-scale, quantitative serum proteomics at the time of diagnosis of biliary atresia and other cholestatic syndromes (serving as disease controls) to identify biomarkers of disease. In a discovery cohort of 70 subjects, the lead biomarker was matrix metalloproteinase-7 (MMP-7), which retained high distinguishing features for biliary atresia in two validation cohorts. Notably, the diagnostic performance reached 95% when MMP-7 was combined with γ-glutamyltranspeptidase (GGT), a marker of cholestasis. Using human tissue and an experimental model of biliary atresia, we found that MMP-7 is primarily expressed by cholangiocytes, released upon epithelial injury, and promotes the experimental disease phenotype. Thus, we propose that serum MMP-7 (alone or in combination with GGT) is a diagnostic biomarker for biliary atresia and may serve as a therapeutic target.

De novo food allergy in pediatric liver transplantation recipients



Sinitkul R, Manuyakorn W, Kamchaisatian W, Vilaiyuk S, Benjaponpitak S, Lertudompholwanit C, ***Treepongkaruna S.**

Correspondence: Suporn Treepongkaruna

Division of Gastroenterology

Asian Pac J Allergy Immunol. 2017 Nov 22. [Epub ahead of print]

Impact Factor = 1.011

BACKGROUND: Food allergy (FA) prevalence is increasing in pediatric liver transplantation (LT). However, the clinical course is still limited.

OBJECTIVE: This retrospective cohort study aimed to identify the prevalence, risk factors, and the natural history of de novo FA in children post LT.

METHODS: Medical records of pediatric LT recipients from Jan 2001 - Dec 2014 were reviewed. De novo FA was diagnosed by symptoms after exposure to culprit food occurring after LT, and improvement after diet elimination. FA was confirmed if reproduced symptoms after re-challenge or documented sensitization or indicated gastrointestinal eosinophilia.

RESULTS: Among 46 post LT children, 54.3% developed de novo FA at a median time of 12.2 months [Interquartile range (IQR) 6.2, 21.3 months] post LT. The confirmed FA was 39.1%. Gastrointestinal symptom was the most common manifestation followed by skin, anaphylaxis, and others. Culprit foods were cow's milk, shellfish, egg, wheat, soybean, peanut, coconut, fish and monosodium glutamate. The risk factors of FA were transplantation during age below 2 years [hazard ratio (HR), 2.62; 95% confidence interval (CI), 1.04 - 6.59; p = 0.03), atopic history in family (HR, 5.67; 95% CI, 1.33 - 24.12; p = 0.01), and Epstein-Barr (EBV) viremia (HR, 2.39; 95% CI, 1.02 - 5.63; p = 0.04).

CONCLUSIONS: de novo FA in pediatric LT is not uncommon. Age at LT younger than 2 years, family history of atopy, and EBV viremia are associated with developing FA. Development of tolerance after elimination culprit diets for 3 years is similar to general population.

Association of indoor air quality and preschool children's respiratory symptoms

***Puranitee** P, Siwarom S, Plitponkarnpim A, Manuyakorn W, Sinitkul R, Arj-Ong Vallipakorn S.

***Correspondence:** Pongtong Puranitee

Division of Ambulatory Pediatrics

Asian Pac J Allergy Immunol. 2017;35(3):119-26.

Impact Factor = 1.011

BACKGROUND: This study aimed to assess the association between exposure to indoor air pollution in DCCs (Child Day Care Centers) and the respiratory symptoms of children under 6 years old.

METHODS: Air quality data were collected three times regarding seasonal variation. Pollutants measured in 11 DCCs included PM10, CO, NO2, SO2, O3, benzene, bacteria, fungi, and dust mite. The frequency of respiratory symptoms including coughing, rhinitis, and dyspnea were recorded via teacher and parent-report questionnaires. Fractional exhaled nitric oxide (FENO) levels were measured to assess airway inflammation.

RESULTS: In total, 436 children participated in the study, with 83% completing data collection in all 3 seasons. The frequency of rhinitis correlated with PM10 (IRR=70.3, 95%CI=12.4-399.7, p< 0.001), CO (IRR=3.2, 95%CI=2.4-4.2, p< 0.001), benzene (IRR=2.3, 95%CI=1.8-3.2, p< 0.001) and D. pteronyssinus level (IRR=2.1 95%CI=1.7-2.7, p< 0.001). The frequency of coughing correlated with levels of PM10 (IRR=15.2, 95%CI=3.0-78.2, p< 0.001), CO (IRR=2.8, 95%CI=2.1-3.7, p< 0.001), and benzene (IRR=1.4, 95%CI=1.1-1.9, p=0.02). The frequency of dyspnea correlated with D. pteronyssinus level (IRR=3.9, 95%CI=1.7-9.1, p=0.001). FENO levels associated with high benzene levels (OR=5.9, 95%CI=1.5-22.9, p=0.01).The majority of DCCs had at least one PM10 measurement above the standard level, which were noted in all 3 seasons. Three DCCs had PM10levels above the standard level in all seasons. Overall, 64% of the DCCs had indoor bacterial counts above the standard level in all seasons.

CONCLUSIONS: PM10 and bacterial count is a significant problem in Bangkok metropolitan DCCs. The respiratory symptoms of children positively associated with PM10, CO, benzene and dust mite levels.

Safety and efficacy of ant rush immunotherapy in children

***Manuyakorn W,** Benjaponpitak S, Kamchaisatian W, Sasisakulporn C, Jotikasthira W.

***Correspondence:** Wiparat Manuyakorn

Division of Allergy and Immunology

Asian Pac J Allergy Immunol. 2017;35(3):156-60.
Impact Factor = 1.011

BACKGROUND: The Rush Immunotherapy (RIT) protocol is a valid alternative in order to reach the maintenance phase early. However, there are scarce studies in the literature that have evaluated the safety and the efficacy of an ant RIT process in children.

OBJECTIVE: To evaluate the safety and the efficacy of an ant RIT protocol and to identify the risk factors for systemic reactions (SRs) during an RIT procedure in children.

METHOD: A retrospective review was conducted for those children who were receiving an ant RIT procedure. The 3-day RIT protocol consisted of hourly subcutaneous injections in order to achieve a 0.5 ml maintenance dose of a 1:100 weight/volume (wt/vol) of the Solenopsis invicta whole body extract. The safety for an RIT procedure was monitored by using the World Allergy Organization Subcutaneous Immunology Systemic Reaction Grading System. The efficacy was assessed by the reactions after a field ant re-sting.

RESULT: A total of 20 children who were receiving an ant RIT therapy were reviewed. The mean age was 9.5+3.07 years. There were 6 systemic reactions (SRs) from 324 injections during the RIT procedure (1.85%). All of the systemic reactions were Grade 1-2. There were no associations of SRs regarding age, gender, an atopic history, or the levels of immunoglobulin E (IgE) sensitization to the ants. Among the 14 patients who experienced a field ant re-sting, 4 (28.5%) patients developed Grade 3 SRs. These Grade 3 reactions were resolved after an increase of the maintenance dose to 0.5 ml of a 1:50 wt/vol. There was a significant difference in the mean age of those children who had ant re-sting systemic reactions and those who had no reactions (6.75+0.95 year vs. 10.8+3.29, p=0.036).

CONCLUSION: Rush immunotherapy with ant in children is safe and it has a low occurrence of severe systemic reactions. It is an alternative treatment for those patients requiring a rapid protection.