

RESEARCH ACTIVITIES REPORT CRU ASSOCIATE MEMBERS (CRAMS) AND CLINICIAN SCIENTIST COTERIE (CSC) FOR SERIE 5/2023 SHARING FROM CRAMS AND CSC MEMBERS! 5/2023



By Salwana Ahmad

CRAMs Online Meeting was held every 2 months among CRAMs Members, Clinician Scientist Coterie (CSC) Members, and staff among Hospital Sultan Abdul Aziz Shah (HSAAS), UPM, and Faculty of Medicines and Health Sciences, UPM. This session was intended for the CRAMs members to share their research activities in the department and how they are coping with all the coming challenges and striving to keep moving forward. During the session, the members will have to present their research activities report comprising remarkable research activities and outputs, promoting positive perceptions and motivation for facing challenges, improving clinical research, and cultivating research & networking. In light of cultivating the spirit of research and knowledge sharing, here are the summaries of the presentation shared for all of us to get to learn how is everyone is doing in proceeding with the quality research in UPM.



DEPARTMENT OF PEADIATRIC

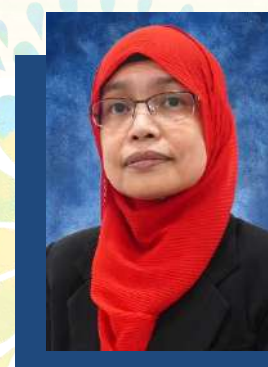
Background:

The Department of Paediatrics was established on 1st June 2006 for clinical teaching and curriculum for year 1 to year 5 Medical Students. Apart from clinical services, this department is also responsible for producing postgraduates from the Master of Medicine (Paediatrics) course to trained qualified graduates as general pediatricians. This program offers full-time course work that aligns with high national standards to produce highly qualified graduates. Apart from the teaching activities mentioned above, the excellence in research and professional consultation is also taken part.

Department Specialist and Lecturers:

The department consists of 17 Specialists in total:

- ❖ 13 Academic members- 8 active academic staff
- ❖ 4 Lecturers on subspecialty training, 1 ongoing PhD.



CRAMs Member:
Dr. Putri Binti Yubbu

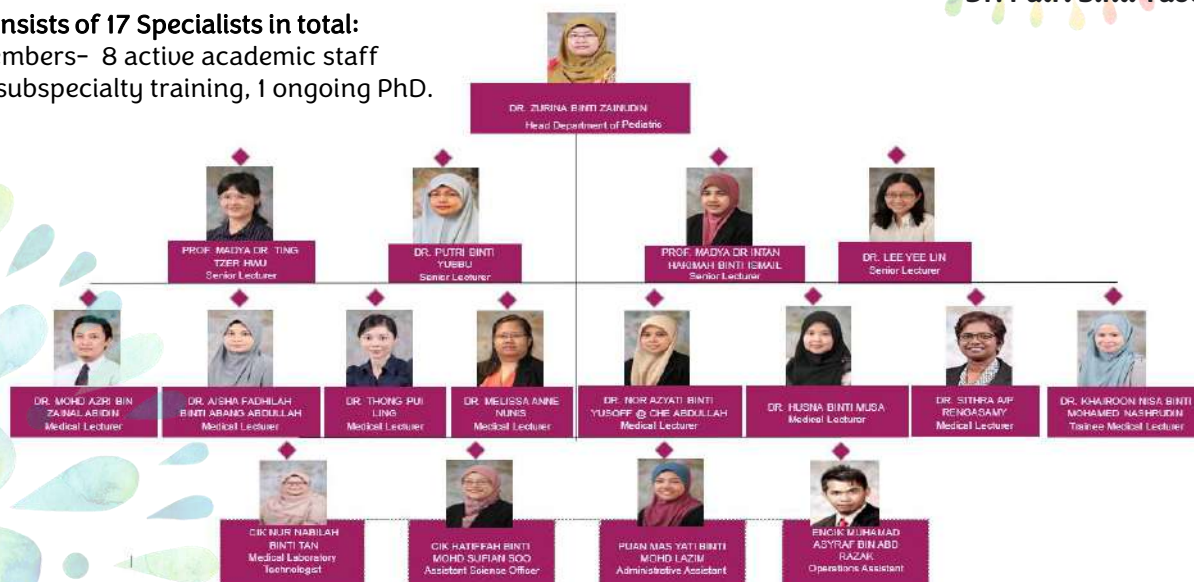


Figure 1 Organization Chart for the Department of Paediatric

Establishment of Clinical Immunology, Dermatology, & Allergy (IDEAL) Research Center

1. Advanced Medical Research in Allergology and Clinical Immunology (AMRAC) is a center for improving the diagnosis and treatment of PID and allergic diseases through clinical activities, research education, and advocacy.
2. It has been well known as Malaysia's referral center for pediatric allergy (food) and clinical immunology (primarily PID).
3. In HSAAS, Allergy and Clinical Immunology Centre (AMRAC):
 - ❖ Provides advanced and comprehensive care for allergy and PID patients
 - ❖ Consist 2 interrelated units to cater to patients and research needs:
 - Allergy & Clinical Immunology Specialist (ACIS) Centre
 - Primary Immunodeficiency and Allergy Diagnostic & Research Laboratory (PEARL)

REMARKABLE RESEARCH ACTIVITIES AND OUTPUTS

Research Highlights and Achievements:

Year	Number of Publication/Year
2020	15
2021	10
2022	12
2023	10 (Ongoing)

Table 1 Number of publications based on past research projects.

Publications Highlight: Top 10% and Q1 and Q2 Journal (Allergy and Immunology)

Journal of Allergy and Clinical Immunology
Volume 146, Issue 5, November 2020, Pages 1005-1007

Paradigms and perspectives

Dietary patterns in childhood and their effect on gut microbiota—an Asian perspective on atopy risk

Intan Hekimah Ismail MMed, PhD¹, Christophe Lay PhD^{2,3}, Noorizan H.A. Majid MMed⁴, Way Seah Lee MD⁵, Bee Wah Lee MD⁶, Amir Hamzah Abdul Latiff MMed, MRCP⁷, Hern Tze Tina Tan PhD⁸, Ian Knol PhD⁹, Yeong Yeh Lee MD, PhD¹⁰

frontiers in immunology

Targeted Gene Sanger Sequencing Should Remain the First-Tier Genetic Test for Children Suspected to Have the Five Common X-Linked Inborn Errors of Immunity

Koon-Wing Chan¹, Chung-Yin Wang¹, Daniel Loung¹, Xinyuan Yang¹, Susanna F. S. Fok¹, Priscilla H. S. Mak¹, Lei Yao¹, Yuan Ma¹, Huazhen Mao², Xiaodong Zhao³, Weiling Leng⁴, Sunit Singh⁵, Menamed-Raha Barboiuhe⁶, Jianxin He⁷, Le-Ping Jiang⁸, Wood-Kang Lee⁹, Minh-Huong Thi Le⁹, Dina Mukhtari¹⁰, Fatma Johanna Santos-Oliveira¹¹, Reza Djighe¹², Prathim Reiset¹³, Intan Hekimah Ismail¹⁴, Amir Hamzah Abdul Latiff¹⁵

frontiers in immunology

Transition practice for primary immunodeficiency diseases in Southeast Asia: a regional survey

Chee Min Chan¹, Amir Hamzah Abdul Latiff², Lukman Muzni Mohd³, Intan Hekimah Ismail⁴, Intan Susana Sidi Hamid⁵, Wee Keng Liew⁶, Yousif Zhora⁷, Narasara Subramanian⁸, Dorena Narisana⁹, Faema Johanna Santos-Oliveira¹⁰, Mary Anne R. Cassio¹¹, Le Nguyen Ngoc-Gwynh¹², Anh Thi Van Nguyen¹³, Nguyen Thanh Trung¹⁴, Nguyen Minh Tuan¹⁵

frontiers in Pediatrics

Impact of Primary Immunodeficiency Diseases on the Life Experiences of Patients in Malaysia From the Caregivers' Perspective: A Qualitative Study

Rayyanah Ahmed Moolali¹, Intan Juliana Abd Hamid², Iliq Fadzilah Hashimi³, Zaitun Thasneem Zamrudin⁴, Priscilla Parveen Abu Bakar⁵, Fathimath Taib⁶, Noorwanry Mohamad⁷, Enock Manganyi⁸, Intan Hekimah Ismail⁹, Amir Hamzah Abdul Latiff¹⁰ and Lukman Mohd Noh¹¹

genes

A Novel De Novo NFKBIA Missense Mutation Associated to Ectodermal Dysplasia with Dysgammaglobulinemia

Chai Teng Chear^{1,2}, Bader Abdul Kader El Farra^{3,4}, Marina Sham⁵, Kavertha Ramalingam⁶, Lukman Mohd Noh⁷, Intan Hekimah Ismail⁸, Mei Yee Chiew⁹, Mohd Farid Baharin⁹, Adirata Mal Ripen¹⁰ and Saharudin Bin Mohamad^{11,12}

frontiers in Cellular and Infection Microbiology

Lactobacillus for the treatment and prevention of atopic dermatitis: Clinical and experimental evidence

Anni Xie¹, Ailing Chen², Yunguo Chen³, Dichen Liu⁴, Shenyu Wang⁵, Daochen Chen^{6,7} and Huiwang Yu⁸

frontiers in Immunology

Editorial: Creating Awareness for Primary Immunodeficiencies in the Southeast and East Asia Regions

Intan Hekimah Ismail¹, Hirokazu Kanegane^{2,3} and Xiaodong Zhao⁴

Allergy

Pharmacological Management of Allergic Rhinitis: A Consensus Statement from the Malaysian Society of Allergy and Immunology

Baharudin Abdullah¹, Amir Hamzah Abdul Latiff², Anura Mithellee Manivel³, Fauziah Mohamed Jamil⁴, Harvinder Singh Dalip Singh⁵, Intan Hekimah Ismail⁶, Jeevanan Jathendran⁷, Jeyasathy Santhasaya⁷, Kent Chee Keen Woo⁸, Phaik Choo Khoo⁹, Kulljit Singh⁹, Nurashikin Mohammad¹⁰, Sakinah Mohamad¹¹, Salina Hussain¹¹ and Rajini Moses^{12,13}

Publications Highlight: Top 10% and Q1 Journal (Paediatric Cardiology)

The International Journal of Cardiovascular Imaging (2022) 38:1505–1516
https://doi.org/10.1007/s10554-022-02587-y

ORIGINAL PAPER

Peak apical recoil rate is a simplified index of left ventricular untwist: validation and application for assessment of diastolic function in children

Putri Yubbu^{1,2} · Hunter Kauffman¹ · Renzo Calderon-Anyosa¹ · Andrea E. Montero¹ · Tomoyuki Sato¹ · Daisuke Matsubara¹ · Anirban Banerjee¹

Received: 30 November 2021 / Accepted: 25 February 2022 / Published online: 15 March 2022
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Abstract
The use of untwisting rate as a novel index of LV diastolic function in clinical practice has been limited due to its tedious and time-consuming analysis. Therefore, we simplify the untwist measurement by only measuring the LV apex's recoil rate and validating and applying peak apical recoil rate (PARR) as an index of diastolic dysfunction (DD) in pediatric subjects during increased and decreased lusitropic states. We recruited 153 healthy subjects (mean age 13.8 ± 2.9 years), of whom 48 performed straight leg raising exercise and an additional 46 patients (mean 8.4 ± 5.6 years) with documented pulmonary capillary wedge pressures (PCWP) (validation cohort). In addition, we studied 16 dilated cardiomyopathy patients (mean age 9.5 ± 6.3 years) (application cohort). PARR and isovolumic relaxation time (IVRT) were compared to PCWP. Both PARR and PARR normalized by heart rate (nPARR) were excellent in detecting patients with PCWP ≥ 12 mmHg and greatly superior to IVRT in this respect (AUC: 0.98, 95% CI [0.96, 1.0] vs. AUC: 0.795%CI [0.54, 0.86]). In DCM patients, PARR and nPARR were greatly decreased compared to controls (–38.6 ± 18.6% vs –63.1 ± 16.3%, p < 0.001) and (–0.43 ± 0.20 %

Journal of the American College of Cardiology
Volume 76, Issue 17, 27 October 2020, Pages 1947–1961

Original Investigation

Echocardiographic Findings in Pediatric Multisystem Inflammatory Syndrome Associated With COVID-19 in the United States

Daisuke Matsubara MD, PhD, Hunter L. Kauffman BS, Yan Wang MD, Renzo Calderon-Anyosa MD, MSc, Sumeekha Nadaral MD, Matthew D. Elias MD, Travis J. White MD, Deborah L. Tarowicz CRNP, Putri Yubbu MBBS, Therese M. Giglio MD, Alexa N. Hoarty MD, Joseph W. Rossano MD, Michael D. Quartermain MD, Anirban Banerjee MD

Cardiomyopathies

Left Ventricular Rotational Mechanics in Children After Heart Transplantation

Hythem M. Nawaytou, MBChB; Putri Yubbu, MD; Andrea E. Montero, MD; Deiparjan Nandi, MD; Matthew J. O'Connor, MD; Robert E. Shaddy, MD; Anirban Banerjee, MD

Background—Left ventricular (LV) dysfunction after orthotopic heart transplantation (OHT) is multifactorial and can be an indicator of graft rejection or coronary artery vasculopathy. Analysis of rotational mechanics may help in the early diagnosis of ventricular dysfunction. Studies describing the left ventricular rotational strain in children after OHT are lacking. It is important to establish the baseline rotational mechanics in pediatric OHT to pursue further studies in this population.

Methods and Results—Rotational strain measured by speckle tracking was compared in 52 children after OHT, with no evidence of active rejection or coronary artery vasculopathy with 35 age-matched normal controls. Twelve OHT patients and 13 controls underwent moderate exercise with pre- and postexercise echocardiography. Torsion, slope of the systolic limb of the torsion–radial displacement loop, and the untwist rate were significantly higher in OHT patients (torsion: median 2.7°/mm [Q1–Q3, 2.3–3.2] versus 2.3°/mm [Q1–Q3, 1.8–2.7]; P=0.03, torsion–radial displacement loop: 2.7°/mm [Q1–Q3, 2.1–3.6] versus 2.0°/mm [Q1–Q3, 1.6–2.7]; P=0.008, indexed peak untwist rate: –21.6°/s/cm [Q1–Q3, –24.3 to –15.7] versus –17.1°/s/cm [Q1–Q3, –19.6 to –13.3]; P=0.01). Contrary to controls, OHT recipients were unable to increase torsion with exercise (OHT: 2.8°/cm [2.3–3.2] versus 3°/cm [2.4–3.5]; P=0.81, controls: 2.2°/cm [2–2.6] versus 3°/cm [2.4–3.7]; P=0.01, pre and post exercise, respectively). The systolic slope of the torsion–radial displacement loop relationship decreased with exercise in most OHT patients.

Conclusions—Baseline rotational strain in OHT patients is higher than normal with a blunted response to exercise. The slope

Journal of the American Society of Echocardiography
Volume 31, Issue 8, August 2018, Pages 951–961

Clinical Investigations
Pediatric Cardiomyopathies

A Preliminary Study of Left Ventricular Rotational Mechanics in Children with Noncompaction Cardiomyopathy: Do They Influence Ventricular Function?

Hythem M. Nawaytou MBChB^{a,b}, Andrea E. Montero MD^a, Putri Yubbu MD^a, Renzo J.C. Calderon-Anyosa MD^a, Tomoyuki Sato MD^a, Matthew J. O'Connor MD^a, Kelley D. Miller CRNP^a, Philip C. Ursell MD^a, Julien I.E. Hoffman MD^a, Anirban Banerjee MD, FACC^a

The International Journal of Cardiovascular Imaging
https://doi.org/10.1007/s10554-018-1367-4

ORIGINAL PAPER

Diagnostic value of myocardial deformation pattern in children with noncompaction cardiomyopathy

Putri Yubbu^{1,2} · Hythem M. Nawaytou^{1,3} · Renzo Calderon-Anyosa¹ · Anirban Banerjee¹

Received: 16 February 2018 / Accepted: 14 May 2018
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Abstract
The current echocardiographic diagnostic criteria for noncompaction cardiomyopathy (NCC) have variable sensitivity and low specificity. Moreover, there are limited data on the use of myocardial deformation imaging for early detection of myocardial dysfunction in children with NCC. We describe left ventricular (LV) deformation patterns in children with NCC, with the goal of identifying a potential diagnostic pattern. We prospectively enrolled 30 children with NCC (47% male; mean age 7.2 years) and 30 age- and gender-matched controls. Extent and severity of non compaction in each segment were evaluated in LV 16-segment model. Regional (base, mid and apex) and segmental (16 segments) longitudinal strain (LS), circumferential strain (CS) and radial strain (RS) were measured using speckle tracking echocardiography. In all patients with NCC, regional and segmental CS and RS at the apex were significantly decreased compared to controls (CS apex: 10.2 ± 5.1% vs 20.2 ± 6.0%, p = 0.001; RS apex: 23.5 ± 6.7% vs 44.1 ± 14.5%, p = 0.001). The circumferential strain

Cardiology in the Young
cambridge.org/cy

Original Article

Cardiovascular causes of tracheobronchial compression: a decade experience in a Paediatric Congenital Heart Centre

Putri Yubbu¹, Haifa Abdul Latiff¹, Husna Musa^{1,2}, Navin Kumar Devaraj¹, Nurul Adha Mohd Razif¹, Sivakumar Sivalingam³ and Harri Samion⁴

Abstract
Background: Vascular compression of the airway often complicates CHD management. This study evaluated the use of CT in determining cardiovascular causes, clinical manifestations, and outcome of tracheobronchial compression among children with CHD. Methods: A retrospective review of clinical records of all patients with CT scan evidence of tracheobronchial compression from January 2007 to December 2017 at National Heart Institute. Cardiovascular causes of tracheobronchial compression were divided into three groups: group I: vascular ring/pulmonary artery sling; group II: abnormally enlarged or malposition cardiovascular structure due to CHD; group III: post-CHD surgery. Results: Vascular tracheobronchial compression was found in 81 out of 810 (10%) patients who underwent CT scan. Group I lesions were the leading causes of vascular tracheobronchial compression (55.9%), followed by group II (24.0%) and group III (19.9%). The median age of diagnosis in groups I, II, and III were 10.8 months, 3 months, and 15.6 months, respectively. Half of group I patients are manifested with stridor

Publications Without Research Grant

Progress in Pediatric Cardiology
Journal homepage: www.sciencedirect.com/journal/progress-in-pediatric-cardiology

Review

Vascular compression of the airways: Issues on management in children with congenital heart disease

Putri Yubbu¹, Navin Kumar Devaraj², Dg. Zuraini Sahadan³, Haifa Abdul Latiff⁴

Abstract
Vascular compression of the airway is an uncommon condition that tends to be diagnosed and treated early, in many cases involving disease with congenital heart disease (CHD), vascular compression of the airway is a significant cause of morbidity. The most common congenital anomalies associated with airway compression are vascular ring (VR). Other causes could include enlargement of the ductus, pulmonary artery, and nodules characterised as a result of underlying cardiac lesions or following cardiac intervention. This diagnostic remains challenging as the clinical presentation of vascular compression of the airway can be nonspecific. Therefore, in children with CHD, a high index of suspicion of possible vascular compression of the airway is essential to any patient experiencing recurrent respiratory symptoms or feeding difficulty in usual variable delay in diagnosis. On the other hand, prenatally detection of the vascular ring has increased significantly over the years that almost eliminate route to symptomatic cases. The latest step away from the prenatal detection of symptomatic patients that may lead to non-invasive and operative or minimally-invasive options, including bronchoscopy and angiography. Magnetic resonance imaging (MRI) and cardiac computed tomography (CT) angiography are a commonly used tool for confirmation of diagnosis and pre-operative planning. The main component of vascular compression of the airway in children with CHD can be equally challenging as complication might arise and affect CHD and subsequent airway disease as a consequence. The non-invasive diagnosis in symptomatic patients with VR or pulmonary artery (PA) sling is essential. Careful pre-operative planning with a good pre-operative and post-operative airway management is essential to ensure successful treatment.

Cardiology in the Young
cambridge.org/cy

Original Article

Cardiovascular causes of tracheobronchial compression: a decade experience in a Paediatric Congenital Heart Centre

Putri Yubbu¹, Haifa Abdul Latiff¹, Husna Musa^{1,2}, Navin Kumar Devaraj¹, Nurul Adha Mohd Razif¹, Sivakumar Sivalingam³ and Harri Samion⁴

Abstract
Background: Vascular compression of the airway often complicates CHD management. This study evaluated the use of CT in determining cardiovascular causes, clinical manifestations, and outcome of tracheobronchial compression among children with CHD. Methods: A retrospective review of clinical records of all patients with CT scan evidence of tracheobronchial compression from January 2007 to December 2017 at National Heart Institute. Cardiovascular causes of tracheobronchial compression were divided into three groups: group I: vascular ring/pulmonary artery sling; group II: abnormally enlarged or malposition cardiovascular structure due to CHD; group III: post-CHD surgery. Results: Vascular tracheobronchial compression was found in 81 out of 810 (10%) patients who underwent CT scan. Group I lesions were the leading causes of vascular tracheobronchial compression (55.9%), followed by group II (24.0%) and group III (19.9%). The median age of diagnosis in groups I, II, and III were 10.8 months, 3 months, and 15.6 months, respectively. Half of group I patients are manifested with stridor

Publications Highlight: Top 10% and Q1 Journal (Paediatric Endocrinology)

CASE REPORT

Nutritional Rickets in Three Toddlers during Covid-19 Pandemic Lockdown

Lee YL^a, Yusoff NA^b, Ting TH^a

^aPaediatrics Endocrine Unit, Department of Paediatrics, Hospital Sultan Abdul Aziz Shah, Universiti Putra Malaysia, Serdang, Selangor, Malaysia
^bDepartment of Paediatrics, Universiti Putra Malaysia, Serdang, Selangor, Malaysia

ABSTRACT

Nutritional rickets is a worldwide problem which has been increasingly reported globally. Three toddlers aged 1-2 years presented in March to April 2021 with bony deformities one year of national Covid-19 pandemic lockdown since March 2020. All three were exclusively breastfed till presentation without formula milk supplementation. Wearing

Received: 4 June 2019 | Revised: 12 December 2019 | Accepted: 13 January 2020
 DOI: 10.1111/pecl.12985

ORIGINAL ARTICLE

Molecular diagnosis of maturity-onset diabetes of the young in a cohort of Chinese children

Aijing Xu¹ | Yunting Lin¹ | Huiying Sheng¹ | Jing Cheng¹ | Huifen Mei¹ | Tzer Hwu Ting² | Chunhua Zeng¹ | Cuili Liang¹ | Wen Zhang¹ | Cuiling Li¹ | Xiuzhen Li¹ | Li Liu¹

¹Department of Genetics and Endocrinology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangzhou, China
²Department of Paediatrics, Faculty of Medicine & Health Sciences, University Putra Malaysia, Serdang, Malaysia

Abstract

Objective: The purpose of this study was to identify the genetic mutations in maturity-onset diabetes of the young and estimate the frequency and distribution of these mutations in southern China.

Journal of Paediatrics and Child Health

ORIGINAL ARTICLE

Thyroid autoimmunity and autoimmune thyroid disease in Malaysian girls with Turner syndrome: An understudied population

Yee L Lee¹, Azriyanti A Zaini², Arini N Idris³, Raja A Abdullah⁴, Jeanne SL Wong^{5,6}, Joyce SS Hong⁷, Suhaimi Hussain⁸, Poi G Lim³, Song H Lim⁹, Noor SM Nor^{10,11}, Loo L Wu¹² and Muhammad Y Jalaludin²

¹Paediatric Endocrine Unit, Department of Paediatrics, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang, ²Paediatric Endocrine Unit, Department of Paediatrics, Faculty of Medicine, University Malaya, ³Paediatric Endocrine Unit, Department of Paediatrics, Hospital Tunku Azizah, ⁴Mal Pakar Kanak-kanak, Universiti Kebangsaan Malaysia, Kuala Lumpur, ⁵Paediatric Endocrine Unit, Department of Paediatrics, Hospital Universiti Sains Malaysia, Kota Bharu, ⁶Paediatric Endocrine Unit, Department of Paediatrics, Putrajaya Hospital, Putrajaya, ⁷Women and Children's Hospital, Kota Kinabalu, ⁸Department of Paediatrics, Faculty of Medicine (I-PPerforM), Universiti Teknologi MARA (UiTM), Shah Alam and ¹²Subang Jaya

Tavana et al. *Italian Journal of Pediatrics* (2022) 48:193
<https://doi.org/10.1186/s13052-022-01385-5>

Italian Journal of Pediatrics

RESEARCH

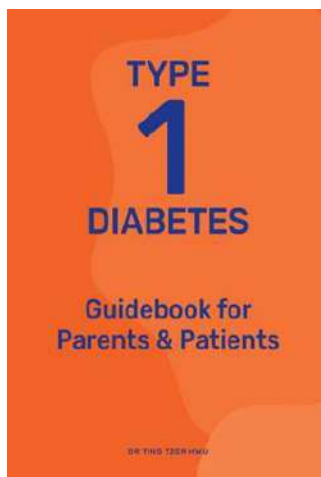
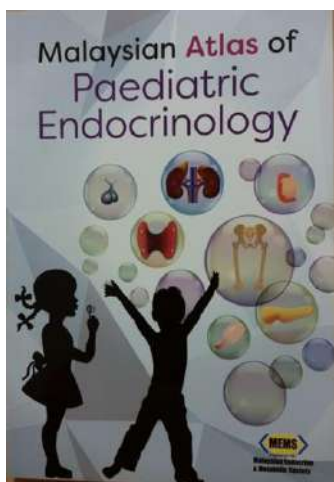
Open Access

Whole exome sequencing identifies two novel variants in *PHEX* and *DMP1* in Malaysian children with hypophosphatemic rickets

Nahid Tavana¹, Tzer Hwu Ting^{2*}, Kaitao Lai^{3,4}, Marina L. Kennerson^{3,5} and Karuppiah Thiakavathy^{1,6*}

Abstract

Background: Hypophosphatemic rickets (HR) is a genetic disease of phosphate wasting that is characterized by defective bone mineralization. The most common cause of the disease is mutations in the phosphate regulating gene with homologies to endopeptidases on the X chromosome (*PHEX*) gene. The aims of this study were to identify the gene variants responsible for HR in three cases of Malaysian origin from three independent families and to describe their clinical, biochemical, and radiological features.



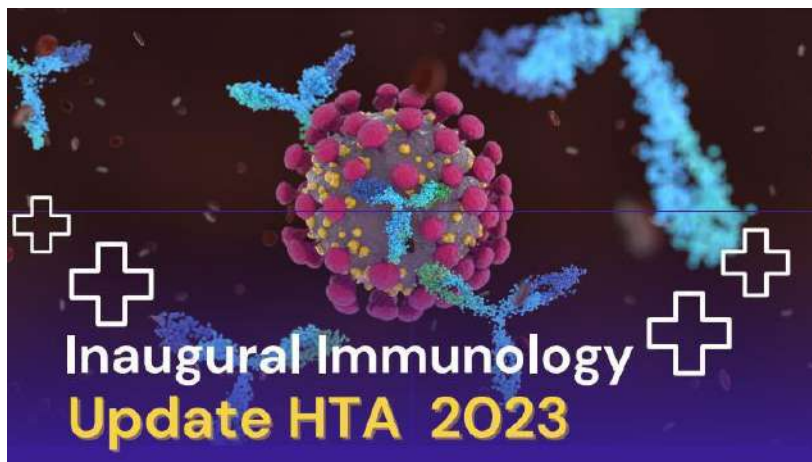
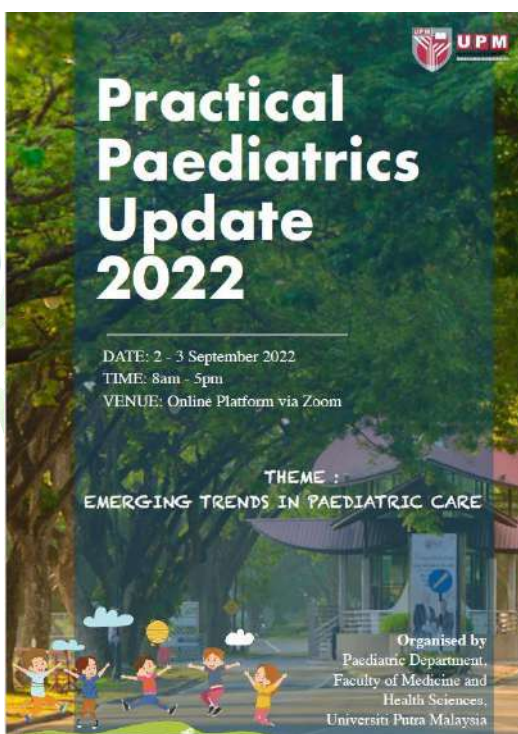
REMARKABLE RESEARCH ACTIVITIES AND OUTPUTS

Research Highlights and Achievements:

Year	Number of Publication/Year
2020	15
2021	10
2022	12
2023	10 (Ongoing)

Table 1 Number of publications based on past research projects.

PRACTICAL PAEDIATRIC UPDATE



Outcome Of Superior Vena Cava Obstruction Following Congenital Heart Disease Intervention in Serdang Hospital
Guruzhanni Vivekanandan, Putri Yubhi, Nur Azrya Yusoff

Abstract
Background: Superior vena cava (SVC) obstruction is a rare but serious complication following congenital heart disease (CHD) interventions in children. It may result in significant clinical sequelae if left untreated. There is limited data on the management approach and outcome of SVC obstruction related to cardiac intervention in children. Therefore, the study aims to investigate the clinical manifestation, management strategy and outcome of SVC obstruction following CHD interventions.
Methods: A retrospective review of the clinical records of all patients with clinical and echocardiographic or radiological findings of SVC obstruction under 18 years from January 2016 to December 2021 at Serdang Hospital was performed.
Results: There were 12 cases of SVC obstruction out of 1200 patients who underwent cardiac interventions. Of the 12 patients, 9 were males (75%), median age 7.2 [3.2-18 months]. SVC obstruction was related to valvular injuries due to central line and pacemaker insertion (58.3%), cannulation during bypass surgery (15.3%) and following Warden procedure (25%). Facial congestion and oedema were the most common presentation (42%), followed by recurrent pleural effusion (42%), dyspnea (25%), hydrocephalus (17%), and 2 (16%) of patients were asymptomatic. About nine patients (75%) underwent central line placement over SVC tributaries. Revascularization with the endoscopic technique was done in 33% of them, where two patients required re-stenting due to in-stent stenosis. Only one patient underwent surgery for SVC repair and thrombus evacuation. About 17% in our study cohort underwent thrombolysis as a primary intervention in acute presentation. Two deaths (16.7%) were reported from patients who underwent no intervention because of severe pneumonia secondary to recurrent pleural effusion and prolonged hospital stay.
Conclusion: Facial congestion, oedema and recurrent pleural effusion were the most common presentation. Endovascular stenting is a safe treatment modality but has a high risk of re-intervention. SVC obstructions following CHD should be managed aggressively due to high morbidity and mortality.

HEALTH RELATED QUALITY OF LIFE (HRQOL) FOLLOWING FONTAN OPERATION AT THE NATIONAL HEART INSTITUTE (NH)
Mohamed Ajmal Ha Mohamed¹, Putri Yubhi², Ming Chen Leong³

¹ Department of Paediatric, Universiti Putra Malaysia
² Department of Paediatric, Universiti Putra Malaysia
³ Paediatric and Congenital Heart Centre, Institut Jantung Negara (National Heart Institute of Malaysia)

Background: Patients with Fontan circulation suffer from morbidity and physical limitations that affects their quality of lives. Therefore, the study examined the clinical characteristics and health-related quality of life (HRQOL) of patients at the National Heart Institute (NH) following the Fontan operation.
Methods: This comparative cross-sectional study was conducted between January 2021 and December 2021 at National Heart Institute, Kuala Lumpur, among patient who are 8 years and older who had undergone Fontan palliation for at least 1 year. Patients were assessed using electronically distributed age-based Pediatric Quality of Life Inventory™ (PedsQL) Generic and Cardiac modules. The scores from the patients were compared with those of age-match students in the Klang Valley and patient's parents.
Results: A total of 113 patients and 120 normal subjects with their parents participated in the study. Almost half of the Fontan procedure patients were in the age group of 13-18 years, female dominant (51.3%), and congenital Atrial was the primary underlying cardiac anomaly (52%). The most common Fontan complication was valve regurgitation (52.4%), followed by arrhythmias (12.4%). Fontan patients have reduced HRQOL scores compared to their healthy peers in almost all domains. The younger patients (aged 8-12 years), low income family and fathers with no formal education, reported significantly low scores in social, cognitive and physical scores, respectively. Parents with SpO2 < 95% and taking more medications showed low HRQOL scores in a social and physical scores. Those with many complications demonstrated significantly low HRQOL scores in all domains. Patients with NYHA class I and those who underwent Cardiac MRI showed good HRQOL scores in almost all domains. There was poor agreement between patients' and parents' perceived HRQOL in most of the domains in the generic module but the difference was not as explicit in the cardiac module.

Title: Covid-19 Related Stress and Fear Level among Parents with and without Primary Immunodeficiency Children in Malaysia

Summary of abstract
COVID-19 pandemic has affected multiple aspects of life all around the world which trigger physical health and mental health concern among the communities and parents with chronic condition children. Primary immunodeficiency disease (PID) is one of the chronic diseases of concern as they are immunodeficient, and hence higher risk for infection and serious complications. This study was aiming to determine the psychological impact of COVID-19 on the parents living with PID children.
This was a comparative cross-sectional study conducted at the referral centre of PID patients in HELM Malaysia. Data were collected by online and manual form survey from 14th January to 7th February 2022 involving total of 202 parents (81 from PID group and 101 from healthy group).
Majority of the respondents were female, Malay, age ranged were between 33-64 years old, from West Malaysia, educated and among the B40 income group. The socio-demographic and socioeconomic characteristics between both groups of parents were comparable. Most of the respondents were having moderate level, followed by severe level of fear of COVID-19. For the COVID-19 Stress Scale, more than half of the respondents had severe level, followed by high stress level, average stress level and mild stress level. None of them showed low stress level. The stress score for FCV-19S was 21.02 ± 8.81, range 7-35, and the CSS stress score was 75.15 ± 27.41, (range 9-134). The group of parents with PID children showed a higher fear and stress level towards COVID-19, however, was statistically not significant with p value 0.261 and 0.790 respectively.
COVID-19 pandemic has introduced unprecedented levels of stress and fear among all parents regardless of the status of the children's clinical health. Mental health aspect of the COVID-19 pandemic are expected to continue for months and years and hence interventions directly targeting parental burn-out and families with children are warranted.

Title: The Risk Factors of Nosocomial Infection after Paediatric Cardiac Surgery in Serdang Hospital, Malaysia.

Presenter: Dr. Khairoun Nisa binti Mohamed Nohradin

Background: Paediatric cardiac surgery outcomes have improved tremendously over the last 20 years and despite the advances in the survival of congenital heart disease (CHD) throughout the years, nosocomial infection (NI) after paediatric cardiac surgery remains a significant cause of morbidity and mortality in children with CHD.
Methodology: A retrospective case-control study was conducted in Serdang Hospital involving patients aged from newborn to 13-years-old who had undergone cardiac surgery over a 3-year study period.
Results: There were 181 patients who were diagnosed with NI after paediatric cardiac surgery and were identified as cases. One hundred eighty one patients with no evidence of NI were drawn via random sampling and were selected as control. There were 230 episodes of NI occurred in 151 patients out of 1150 patients who had undergone cardiac surgery, yielding a NI rate of 18.3%. The most frequent site of infections was ventilator associated pneumonia (VAP), followed by central line blood stream infection (CLABSI), pneumonia and surgical site infections (SSI). The most abundant organisms yielded in rank of frequency order were *Pseudomonas aeruginosa*, followed by *Acinetobacter baumannii*, *Stenotrophomonas maltophilia* and *Enterobacter coli*. Multivariate analysis revealed the following independent risk factors for NI: patients with underlying Devereux syndrome and other syndromes, pre-operative hospitalisation days of more than 7 days, lymphoplasma, aortic cross clamp time of more than 60 days, post-operative ventilation days of more than 7 days and intensive care unit (ICU) stay of more than 17 days. The case fatality rate of NI was 21.5%.
Conclusion: The results of the present study draw attention on the preventive measures to improve the outcome of cardiac surgery. Continuous surveillance, as well as better infection control practices, must be enforced to reduce the incidence of NI, and its morbidity and mortality.

ABSTRACT

Safety and Efficacy of Pharmacological Treatments Available for Multisystem Inflammatory Syndrome in Children (MIS-C): A Systematic Review

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Introduction: In early April 2023, rare cases associated with SARS-CoV-2 were reported in children known as Multisystem Inflammatory Syndrome in Children (MIS-C). However, this emergent resulted in lack of studies and evidence-based suggestions established in pharmacological approaches for MIS-C. **Objective:** This study aims to describe the existing pharmacological management for MIS-C. **Methods:** A systematic search via EBSCOhost and Scopus databases was conducted on August 18, 2023, using the terminologies children, MIS-C, PIMS, and SARS-CoV-2. A PRISMA flow diagram was used to report the study selection process. Newcastle-Ottawa Scale (NOS) and GRADE tools were used for quality analysis process. Data synthesis was done by extracting the interventions on treatments used, efficacy and side effects. **Results:** From the included 28 articles, 2128 children with MIS-C were studied. The main pharmacological approaches were immunomodulatory therapy; intravenous immunoglobulin

Research Highlights and Achievements:

No.	STAFF NAME	RESEARCH TITLE	Grant	Submitted/ Approved/ Rejected	Amount (RM)
1.	Assoc. Prof. Dr Intan Hakimah Ismail	Characterisation of Immunoglobulins (IgG, IgA, IgM), IgG Subclasses and Specific Antibody Responses Among Healthy At Hospital Pengajar Universiti Putra Malaysia and Hospital Serdang	IPS	Approved	20,000.00
2.	Dr Zurina Zainudin	Investigating The Cause Of Spontaneou Preterm Births Through Whole Exome DNA Mutation And Quantitative Gene And Protein Expression Analyses	FRGS	Ongoing (2020 – 2023)	185,900
3.	Dr. Putri Yubbu	Gene Mutations In Heterotaxy Syndrome With Congenital Heart Disease Using Next Generation Sequencing Technology	Heartlink	Approved-MoA	29,273.99
		Outcomes Following Prenatal Diagnosis Of Fetal Rhabdomyomas: A Fetal Heart Society Collaborative Study	Fetal Heart Society	Approved-MoA	40,000.00
		Clinical Profile And Genetic Analysis Of Children With Heterotaxy Syndrome Using Whole Exome Sequencing	FRGS	Rejected	190,000.00
		Improving Clinical Skill That Leverage Audio-visual Aids In Detecting Abnormal Heart Sounds Among Medical Students	GIPP	Rejected	20,000.00
4.	Dr. Melissa Anne Nunis	Factors Associated With Successful Peripheral Intravenous Cannulation Among Neonates In Hospital Pengajar Universiti Putra Malaysia	IPM	Ongoing	48,360
5.	Dr. Khairoun Nisa Mohamed Nashrudin	Title: Pemindahan Ilmu Mengenai Asas Pertolongan Cemas Kanak- Kanak ('Basic First Aid For Children') Kepada Komuniti Suri Rumah Bandar Baru Bangi.	(KTGS)	Submitted	9050

Current Research Activities:

YEAR	TITLE	PRINCIPAL INVESTIGATOR	CO-INVESTIGATORS	BUDGET (RM)	ETHICS APPROVAL
2021	Discovery of genetic aberrations in paediatric patients suspected of rare genetic disorders	Assoc. Prof. Dr.Ting Tzer Hwu, Jabatan Pediatrik	<ul style="list-style-type: none"> Prof. Madya Dr. Intan Hakimah Ismail Dr. Lee Yee Lin Dr. Mohd Azri Bin Zainal Abidin Dr. Zurina Zainudin Dr. Nor Azyati Yusoff Dr. Melissa Anne Nunis Dr. Sithra A/P Rengasami 		JKEUPM-2021-436
2021	Efficacy of vein translumination device among neonates admitted to HPUPM- a randomized control study	Dr. Melissa Anne Nunis	<ul style="list-style-type: none"> Dr. Zurina Zainuddin Dr. Lim Zi Han (master student) 	IPM Grant RM48300	JKEUPM-2021-039
2021	Covid 19 stress and fear levels among parents if primary immunodeficiency children in Malaysia	Prof. Madya Dr. Intan Hakimah Ismail	<ul style="list-style-type: none"> Dr. Mohd Azri Bin Zainal Abidin Dr. Saidatul Saadah Ramlah (master student) 		JKEUPM-2021-715
2021	Health related Quality of Life among primary Immunodeficiency patients in HPUPM	Prof. Madya Dr. Intan Hakimah Ismail	<ul style="list-style-type: none"> Dr. Mohd Azri Bin Zainal Abidin Dr.Theiva Rani (master student) 		JKEUPM-2021-353

PROMOTING POSITIVE PERCEPTIONS AND MOTIVATION FOR FACING CHALLENGES, IMPROVING CLINICAL RESEARCH, and CULTIVATING RESEARCH & NETWORKING.

Challenges

The department found that it was hard to do research for a few reasons:

1. Unsupportive environment to do research.
2. Long process for ethical clearance and approval.
3. Long process to get MoA agreement to complete.

Motivation

Moving forward by sharing knowledge and joining more conferences, programs, workshops etc.

Steps were taken to improve clinical research

Step forward for publications

- Adding the APC cost in Grant proposal.
- Using Tabung Amanah as Grant for the lecturer who do not have grant

Brainstorming ideas

- Participating in multiple workshops online including CRAM CRU online.

Find good opportunities for networking and collaboration

- Collaborating with private industry for clinical trials.

Clinical Profile And Outcome Of Superior Vena Cava Obstruction Following Congenital Heart Disease Intervention In A Single Cardiac Center

N. Yusoff, P. Yubbu, G. Vivekanandan, YK Ooi, A. Abdul Ghani, S. Mazlan, GT Koh;
Pediatric Department, Universiti Putra Malaysia, Serdang, MALAYSIA, Pediatric Cardiology, Hospital Sultan Ismail, Serdang, MALAYSIA, Cardiothoracic Department, Hospital Sultan Ismail, Serdang, MALAYSIA

INTRODUCTION	RESULTS	DISCUSSION
Superior vena cava (SVC) obstruction is a rare but serious complication following congenital heart disease (CHD) interventions in children. It may result in significant clinical sequelae if left untreated. There is limited data on the management approach and outcome of SVC obstruction related to cardiac intervention in children. Therefore, the aim is to investigate the clinical manifestation, management strategy, and outcome of SVC obstruction following cardiac interventions.	<p>Reconstructed CT-Three views reveal narrowing of SVC in PA-section</p> <p>Angiogram showing a narrowing of the SVC into irregularly dilated to posterior with the intervention over (white arrow)</p> <p>Angiogram showing SVC stenosis. Poorer flow across the stenosis and flow across the stenosis and good distal perfusion</p>	The prevalence of SVC obstruction following cardiac interventions in this cohort study is 0.8%, with male predominance and 2 mortality cases as result of complication related to SVC obstruction. Vascular injury secondary to central line insertions is the leading risk factor behind the obstruction. Poor antegrade blood flow has led to facial congestion and oedema, the commonly reported sign and symptoms. Although recurrent pleural effusions were only reported in 42% of the study, it has proven to be the most essential associated complication if no intervention is

8th World Congress of Pediatric Cardiology and Cardiac Surgery
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At the heart of the pediatric and cardiac GLOBAL COMMUNITY

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Ventricular Septal Defect Is A Risk for Sinus of Valsalva Rupture in Adults: Experience for 2 Main Cardiac Referral Centers in Malaysia

Paten Yubbu, Govindaraja Vivekanandan, Er Chee Yeh, Abdul Muz Jamil, Leong Ming Chern,
1. Department of Pediatric, Faculty of Health and Sciences, University Putra Malaysia, 2. Cardiothoracic Department, Sultan Ismail Shah Hospital, Serdang, 3. Pediatric Congenital Heart Center, National Heart Institute (NH) Malaysia

INTRODUCTION	RESULTS	DISCUSSION																										
Ruptured Sinus of Valsalva (RSOV) is uncommon but can be associated with significant morbidity and mortality. Despite being the most common type of congenital heart disease (CHD) with a good prognosis, ventricular septal defect (VSD) association with RSOV is rarely studied in detail. The	<table border="1"> <thead> <tr> <th>Clinical parameters</th> <th>n (%)</th> </tr> </thead> <tbody> <tr> <td>Number of patients</td> <td>29</td> </tr> <tr> <td>Age of presentation, median (IQR) year</td> <td>20 (17-30)</td> </tr> <tr> <td>Male %</td> <td>62</td> </tr> <tr> <td>BMI (range)</td> <td>20.2 (17.25-25.86)</td> </tr> <tr> <td>The interval between presentation and surgery, months</td> <td>2.8 (0.4-2.9)</td> </tr> <tr> <td>NYHA Classification</td> <td></td> </tr> <tr> <td>Class I</td> <td>12 (24%)</td> </tr> <tr> <td>Class II</td> <td>10 (34%)</td> </tr> <tr> <td>Class III</td> <td>4 (9%)</td> </tr> <tr> <td>Class IV</td> <td>3 (9%)</td> </tr> <tr> <td>Dynamic</td> <td>14 (48%)</td> </tr> <tr> <td>Pathology</td> <td>14 (48%)</td> </tr> </tbody> </table> <p>Surgical intervention</p> <p>RSOV and isolated aortic regurgitation</p> <p>RSOV repair with VSD/ASD repair</p> <p>RSOV repair with aortic regurgitation</p>	Clinical parameters	n (%)	Number of patients	29	Age of presentation, median (IQR) year	20 (17-30)	Male %	62	BMI (range)	20.2 (17.25-25.86)	The interval between presentation and surgery, months	2.8 (0.4-2.9)	NYHA Classification		Class I	12 (24%)	Class II	10 (34%)	Class III	4 (9%)	Class IV	3 (9%)	Dynamic	14 (48%)	Pathology	14 (48%)	A total of 20/230 heart surgeries were performed, from which 29 patients diagnosed to have RSOV and underwent surgical intervention giving an incidence rate of 1.4%. The overall outcome of the surgery was good but 1/3 (4%) mortality due to septic shock, reported in a case of RSOV with severe AR and TR complicated by infective endocarditis.
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We would like to thank Dr. Putri Yubbu for sharing. We hope that the sharing can transform tacit knowledge into explicit, written, and easily communicated knowledge for the right people to receive the right information at the right time. See you the next time!



Check out more information about our CRU Associate Members (CRAMs) for the Year 2022/2023 Member on HSAAS website at [CRAMs Members](#).

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