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## Oral Presentation

### Ototoxicity in Children with Hepatoblastoma Receiving Platnum Chemotherapy: Audiologic and Speech Profile

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**Background:** Hepatoblastoma (HB) is the most common primary liver tumour in children. Platinum-based chemotherapy is widely applied in treatment of HB, however is associated with significant ototoxicity leading to permanent hearing loss that may impact speech sound perception and articulation.

**Purpose:** This study aimed to describe the platinum-induced hearing loss associated with the treatment of children with HB, its frequency and onset time. Speech perception function and articulation pattern were also investigated.

**Methods:** Retrospective review of medical records was done in paediatric patients with HB diagnosed between 2011 to 2020 and treated in Hong Kong public hospitals. Charting of hearing loss, severity and onset time was done by audiologist. Modified Brock Criteria were used to define the grade of hearing loss, if any. Speech therapy progress notes were analysed by speech therapist for describing speech perception function and any articulation error.

**Findings:** Records of 16 patients were complete and included in the analysis out of the 41 patients identified. High-frequency sensorineural hearing loss was noted in 37.5% of patients (6/16) and the average onset time was two months after the start of the first dose of chemotherapy. Further deterioration of hearing loss was noted in majority of patients (5/6) with severity ranged from grade 1B (20%), 2A (40%) and 3 (40%). No patient was referred for hearing aid while only one patient was referred to speech therapy for his speech problem – stopping error of high frequency sound /s/ was noted (/s/->[t]) despite patient was able to discriminate the two sounds at word level.

**Conclusions:** Prevalence rate of hearing loss in this population might not be accurately evaluated in this study given the poor integrity of data and inconsistent tests administered. However, as paediatric patients are experiencing improved survival, effects and implications of high-frequency hearing loss with regards to speech and language development should be considered in their treatment. Regular hearing monitoring along chemotherapy cycles as well as early referral to hearing aids prescription and speech therapy especially for young children are advocated.

### Incorporation of Tubular Dysfunction Staging Into the Diagnostic Criteria of Acute Kidney Injury for Assessment of Renal Insult in Paediatric Critical Care

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**Introduction:** Electrolyte disturbances due to tubular dysfunction is a prevalent condition in paediatric critical care. We evaluated the relationship between electrolyte disturbances and tubular dysfunction among critically ill children and determined the role of incorporating the severity of tubular dysfunction into the diagnostic criteria of acute kidney injury (AKI) for assessing the degree of renal insult.

**Methods:** A prospective study recruiting children aged 1 month to 18 years old from 6/2020 to 6/2021 admitted to the paediatric intensive care unit (PICU) of the Hong Kong Children's Hospital was conducted to study the epidemiology of AKI and electrolytes disturbances. Appropriate urinary investigations for tubular function assessment were also performed.

**Findings:** There were 254 episodes of admission (58.3% male) enrolled for final analysis with a mean age of 6.4±5.5 years old. The incidence of AKI was 41.3% and 94.9% of children developed electrolyte disturbances. The median number of electrolytes disturbance was 3 (2) types. A high proportion of urinary wasting of magnesium (70.6%), phosphate (67.4%) and potassium (28.6%) were observed among children with hypomagnesaemia, hypophosphataemia and hypokalaemia. 62.6% of children with ≥2 types of electrolyte disturbances showed elevated urinary beta-2-microglobulin level. The number of types of tubular dysfunction was associated with PICU stay (relative risk 1.6 [1.4, 1.8]). A staging system based on the number of types of tubular dysfunction was devised with distribution of nil: 17.7%, stage 1: 45.3%, stage 2: 29.5% and stage 3: 7.5%. Compared to using the AKI staging alone, incorporating the tubular dysfunction staging into the AKI staging system performed better for modelling the

PICU length of stay (akaike information criterion 1957.8 vs 2106.5) but less well for the prediction of PICU mortality (area under the curve of 0.893 vs 0.937).

**Conclusion:** Electrolyte disturbances and tubular dysfunction were commonly observed in PICU and were associated with both morbidity and mortality. The tubular dysfunction staging may provide information regarding electrolytes status and morbidity and could serve as an additional domain to capture a more comprehensive picture of acute renal insult.

### A Quality Improvement Project on Reducing Perioperative Hypothermia for Very Low Birth Weight Infants

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**Background:** Very low birth weight (VLBW) infants are at risk of hypothermia as they have inadequate subcutaneous fat and under-developed thermoregulatory mechanism. This risk is increased when the infant undergoes procedure in the operating theatre due to the cool environment and the general anaesthesia. Definition of hypothermia according to WHO, is axillary temperature <36.5℃. Hypothermia is also stratified as mild (36℃-36.4℃), moderate (32℃-35.9℃) and severe (≤32℃, and <35℃ in extreme low birth weight (ELBW) babies). In November 2021, two ELBW infants' temperature had recorded significant hypothermia with axillary temperature <35℃ upon arrival to NICU after operation. These had brought the attention to the clinical team. A quality improvement project aimed at reducing hypothermia in neonates throughout the process of pre-/ intra-/ post-operation and on arrival from OT to NICU was conducted.

**Objectives:** 1) To reduce perioperative hypothermia incidence in neonatal surgeries. 2) To implement improvement strategies to maintain the temperature within normal range in neonates requiring operation through cross-departmental collaborative efforts. 3) To standardise the workflow for enhancing the thermoregulation of neonates in both NICU and operating theatre.

**Methodology:** A Quasi-Experimental design study was conducted to evaluate the effectiveness of implementing the standardised workflow and checklist on reducing perioperative hypothermia. Retrospective chart reviews from electronic clinical information system in OT and NICU were performed during the pre-intervention review period (1/1/2019 till 8/11/2021) and post-intervention review period (16/11/2021 till 6/7/2022). Definition of hypothermia by WHO was adopted. Post-operative temperature measurement method was axillary. Multi-disciplinary team discussions were held to identify possible gaps leading to intra-operative hypothermia prior to interventions.

**Interventions:** Educational activities in enhancing OT nurses' awareness in neonatal thermoregulation, nurse initiatives in developing and implementing standardised workflow and checklist to guide thermal care in both NICU and OT.

**Outcome Measurement:** Demographics data was collected and recorded. Primary outcome was patient's post-operative axillary temperature within 30 minutes on arrival to NICU from OT. Secondary outcome included intraoperative temperature, relationship between patient or clinical characteristics and hypothermia. Data was input and analysed using IBM SPSS version 26. Descriptive analysis was performed, including means, standard deviations (SD) for continuous variables, and frequencies and percentage for categorical variables. Mean difference of the pre- and post-interventions temperatures between subjects and control were analysed with the unpaired samples t-test.

**Findings:** 81 episodes (pre-intervention) and 27 episodes (post-intervention) of neonatal surgeries were reviewed and analysed. Episodes of postoperative hypothermia were significant decreased from 57% to 11% ($p<0.0001$). Moreover, the mean temperature significantly increased from 36.3℃ (pre-intervention) to 36.9℃ (post-intervention). Body weight at operation was break down into 4 groups for calculation and significant relationship shown between body weight at OT and/or intra-operative blood transfusion and hypothermia. There was 5/27 episodes (19%) of intraoperative hypothermia captured during post-intervention period, and 2/27 episodes (7.4%) found post-operative hyperthermia (>37.5℃) the relationship between intra-operative hypothermia and post-op temperature was inconclusive. The hypothermia risk is increased in VLBW infants undergone major operation requiring intra-operative blood transfusion.

**Conclusion:** Hypothermia may have detrimental effect in neonates especially ELBW infants. However, perioperative hypothermia could be significantly reduced by implementing the improvement strategies through multidisciplinary collaboration.

### The Topoisomerase-I Inhibitor Gimatecan Potently Suppresses B-cell Precursor Acute Lymphoblastic Leukaemia Without Concurrent Cardiotoxicity

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**Background and Aims:** Topoisomerase-II inhibitors such as daunorubicin are standard chemotherapeutic agents for the treatment of leukaemia. However, severe complications could be resulted from their well-recognised myelosuppressive and cardiotoxic nature. This study aims at assessing the efficacy, safety and mechanism of gimatecan, a topoisomerase-I inhibitor currently being investigated in Phase II trials for advanced solid cancers, in a preclinical setting of acute leukaemia.

**Methods:** Gimatecan activities were screened *in vitro* on B-cell precursor acute lymphoblastic leukaemia (BCP-ALL) and acute myeloid leukaemia (AML) cell lines as well as cord blood haematopoietic stem/progenitor cells (HSPCs). Efficacy *in vivo* was evaluated in NOD/SCID or NSG mice grafted with BCP-ALL cell lines or B-lymphoblasts from children with refractory leukaemia. Impact on normal haematopoiesis was assessed in HSPC-reconstituted mice, and cardiotoxicity on human iPSC-derived cardiomyocytes. Mechanisms of action were mined by RNA sequencing followed by Western blotting validation.

**Results:** Gimatecan exhibited an exceptionally higher potency (median IC50: 1.2 nM) than standard chemotherapeutics (median IC50: 182-361 nM) against BCP-ALL with profound selectivity over AML (3.6-fold) or HSPCs (75-fold). Following single-agent gimatecan treatment, complete disease remission (>95% inhibition) and markedly extended survival (1.2-3.3-fold) were observed in animals grafted with BCP-ALL cell lines with distinct cytogenetic backgrounds, and in patient-derived xenografts of highly resistant BCP-ALL failing salvage chemotherapy or upfront immunotherapies. Recurrent sensitivity was observed at leukaemia relapse, suggesting its potential for reinduction. Gimatecan demonstrated a mild but transient suppression on normal haematopoiesis and did not induce myocardial damage as opposed to daunorubicin. Mediation of classical apoptosis, G2/M cell cycle arrest and activation of the tumour suppressive p53 pathway underlay its leukaemia inhibitory mechanisms.

**Conclusions:** Gimatecan possesses remarkable anti-leukaemia activities and tolerable toxicity profiles compared to counterpart topoisomerase-II inhibitors. This study therefore provides a strong scientific foundation to repurpose gimatecan for BCP-ALL and points towards randomised clinical trials.

### Iron Deficiency (ID) Among School-aged Adolescents in Hong Kong: Prevalence, Predictors and Effects on Quality of Life

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**Purpose:** ID is a common nutritional deficiency affecting children/adolescents worldwide. This study aimed to determine the (1) prevalence of ID and ID with anaemia (IDA) among school-aged adolescents, (2) identify the clinical and dietary predictors of iron status, and (3) its impact on HRQoL.

**Methods:** We recruited 183 boys and 340 girls (mean age=17.55, SD=1.05 years) from 16 schools through the Red Cross Blood donation campaigns. Their serum ferritin (Fe) levels and complete blood counts were measured. ID is defined as Fe <30 pmol/L. IDA is defined as ID with anaemia, which is defined as a haemoglobin level <12 g/dl (girls) and <13 g/dl (boys). They also reported their dietary and menstrual (girls only) patterns using structured questionnaires, and HRQoL using the PedsQL 4.0. Multivariable general linear modeling were conducted to identify predictors of Fe, adjusting for age and sex.

**Findings:** The overall prevalence of ID and IDA was 9.9% and 6.5%, respectively. None of the boys had ID or IDA. Among girls, the rate of ID was 15.2%, and IDA was 10.0%. One-third (36.3%) of the participants (boys 28.4%, girls 40.6%) reported having a regular habit of skipping ≥1 meal/day. Lower Fe was found in adolescents who skipped ≥1 meal/day (Est=-35.1, SE=14.7; P=0.017), or skipped breakfast (Est=-36.8, SE=15.6; P=0.019). Lower Fe was found in girls who reported higher average number of sanitary products per cycle (P=0.0072) and higher bleeding severity (P=0.043). The multivariable analyses showed that lower Fe (per 10 pmol/L) is correlated with poorer physical functioning (Est=0.78, SE=0.27; P=0.047) and school functioning (Est=0.81, SE=0.40; P=0.045). Skipping meals is associated with poorer physical

functioning (P=0.0017) and school functioning (P=0.027).

**Conclusions:** The rate of IDA is low among adolescents in Hong Kong. However, ID rate in girls (15.2%) is similar to the higher end of the prevalence rates in other industrialised countries (5.2%-16.6%). Medical professionals should identify anemic symptoms in girls before they manifest as functional impairments, and improve awareness on the potential health consequences of poor dietary habits on ID and the well-being of adolescents.

**Funding:** Health and Medical Research Fund, Food and Health Bureau, the HKSAR Government (Ref: 17180441). We would like to acknowledge the principals and staff of the participating schools.

### Genetic Diagnosis and Clinical Characterisation of Facioscapulohumeral Muscular Dystrophy in Hong Kong Chinese Patients Using Molecular Combing and Whole-Exome Sequencing

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**Background:** Facioscapulohumeral Muscular Dystrophy (FSHD) is the third most common hereditary muscular dystrophy with autosomal dominant inheritance. Most patients have asymmetrical onset of weakness of the face, shoulder, hip and leg muscles. FSHD1 (95% of cases) is due to the contraction of D4Z4 units. Currently, FSHD1 genetic testing is only available oversea or in local private genetic laboratory. Clinical trials on treatment are underway overseas.

**Purpose:** The study evaluates the genotypic-phenotypic profiles of FSHD1 patients in Hong Kong.

**Methods:** This prospective cohort study recruited patients with clinical symptoms suggestive of FSHD from QMH Adult and Paediatric Neurology clinics from March 2021 to July 2022. Molecular combing or Southern blotting determined the D4Z4 unit number. Patients confirmed FSHD1 enrolled the FSHD patient registry and completed the survey on symptoms evaluation. FSHD clinical score determined from examination assessed clinical severity.

**Findings:** Seventy-three patients joined the study. Fifty patients from 33 families (68.5%) were confirmed to have FSHD1 with male to female ratio 1.27:1. The mean age of symptoms onset 23.4y (SD:15.7y) and that of diagnosis was 50.0y (SD:15.8y). While facial and shoulder weakness was the earliest symptom recognised (mean age: 25y), shoulder and leg muscle weakness were the most common symptom reported (n=26/32, 81.3%). Patients with lower D4Z4 unit numbers were found to have earlier symptom onset (PCC=0.580, p=0.001) and higher clinical severity (PCC=0.476 p=0.006). For the ten elderly patients (≥60y), all patients required to use assistive aids for walking had D4Z4 unit ≤6 units (n=6). Compared to traditional southern blotting, molecular combing also gave more robust genomic data for diagnosis.

**Conclusion:** This study confirmed FSHD1 is a common hereditary muscular dystrophy in Hong Kong and the D4Z4 unit number has prognostic influence that helps guide the counselling, management and possible targeted treatment in the future.

### An Extended Asthma Service After Hospital Discharge with a Newly Asthma Application

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**Background:** Asthma education is an effective intervention for reducing readmission in asthmatic children. A newly developed asthma education app is an informative kit and two-way communication system for asthmatic children and their families, as they can retrieve asthma-relevant information and self-assess their asthma-controlled status from the asthma app after discharge.

**Objective:** To test the feasibility of the newly launched asthma app.

**Methods:** Parent-child dyads were invited if the child aged 2-17 was admitted, from August 2021 to July 2022, with an asthma diagnosis or had more than two episodes of wheezing attacks within a year. All the participants downloaded the asthma app on their smartphones during hospitalisation and had a phone follow-up at 4-week after discharge. The data was collected through the clinical management system, phone follow-up, and the asthma app.

**Findings:** Forty-two parent-child dyads were invited to this pilot study, but one dyad could not participate because of the mobile setting. A significant number of children were aged 2-7 (70%). Over half of the participants (56%)

have self-assess over two weeks, and not less than one-fifth have self-assess until the subsequent follow-up. Only one participating child (2.4%) without a prescribed preventer was readmitted within 30 days after being discharged. Three participating parents posted their questions regarding inhalers through the app.

**Conclusions:** The new asthma app has been smoothly launched and is effective in asthma control. The self-assess report helps adjust the medication dosage and treatment plan. A further study is to measure the effectiveness the asthma control in children.

### Modified Renal Angina Index for Prediction of Acute Kidney Injury Among Critically Ill Children

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**Introduction:** Renal angina index (RAI) is a useful clinical tool for determining the risk of acute kidney injury (AKI) among critically ill children. Recent studies have identified additional AKI risk factors that are not included in the RAI.

**Methods:** A prospective study on AKI epidemiology was conducted from 6/2020 to 6/2021 on children aged 1 month to 18 years old admitted to the Paediatric Intensive Care Unit (PICU) of the Hong Kong Children's Hospital. AKI was diagnosed according to the KDIGO criteria. Medication intensity was defined as the number of concomitant nephrotoxic medication exposure. We determined the relative risk (RR) of potential risk factors for AKI development and used the receiver operating characteristics (ROC) curves to evaluate if incorporating additional factors into the RAI model could improve its diagnostic performance.

**Findings:** Altogether 254 episodes of admission were enrolled for analysis. 58.3% of them were male with a mean age of 6.4±5.5 years old. The AKI incidence was 41.7%. Apart from factors that were already included in the RAI, additional factors including post-cardiac operation (RR: 1.36 [1.00, 1.85]), non-invasive ventilatory support (RR: 2.63 [1.36, 5.06]), PIM3 score (RR: 1.05 [1.02, 1.08]), total dose of nephrotoxic medication (RR: 1.01 [1.00, 1.01]) and maximal medication intensity (RR: 1.15 [1.04, 1.28]) were identified. The predictive performances of PIM3 score, the original RAI model, and two additional models incorporating different combinations of new factors were evaluated. The relative rise of serum creatinine level on PICU admission, instead of Day 1 of PICU stay was used. Model 3 yielded the best area under the curve (AUC) for predicting AKI during PICU stay (AUC: 0.82) with the optimal cut-off point being 8. It also performed the best for predicting AKI on Day 2 (AUC: 0.75) and Day 3 (AUC: 0.72) of admission.

**Conclusion:** The use of serum creatinine on PICU admission with the incorporation of post-cardiac surgery, requirement of non-invasive ventilation and nephrotoxic medication exposure into the RAI model could enhance its predictive performance for AKI development among critically ill children.

### Comparison of Flow Rate Between Bottle Teats Typically Used in Neonatal Unit and Commercially Available Teats in Hong Kong

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**Background:** Oral feeding can be physiologically challenging for medically fragile infants. Multiple factors contribute to swallowing safety during bottle feeding such as medical condition, maturity and rate of milk flow. The rate of milk flow can affect infants' coordination of fluid management with respiration; feeding an infant with a teat of inappropriate flow rate could affect feeding performance or increase the risk of aspiration. The flow rate varies with different teats, and thus a quantitative comparison of the flow rates of common locally available teats is crucial to medical professionals working with infants.

**Purpose:** This study aimed at determining the quantitative differences of milk flow rates of currently available teats used for bottle-feeding infants in neonatal unit and the commercially available teats.

**Methods:** Four disposable or reusable teats (H1-H4) used in wards of Hong Kong Children's Hospital, and six teats (C1-C6) of three brands available commercially suggested for preterm and 0-6 month-old babies – with two levels of flow rate from each brand were identified. Flow rates of teats were determined by measuring the amount of milk expressed in the first minute using a breast pump by Medela, simulating an infant's sucking. All teats were fitted onto a 60 ml bottle filled with 60 ml of Wyeth S-26 ultima stage 1 ready-to-feed infant formula at room temperature,

tilted with a standard angle for maintaining the same hydrostatic pressure. Each teat was tested three times for an averaged flow rate.

**Findings:** Flow rates of H1-H4 ranged from 10.6 ml/min to 37 ml/min (mean: 20.3 ml/min), while that of the C1-C6 ranged from 4.3 ml/min to 17.3 ml/min (mean: 10.0 ml/min). H4 (flat teat with wings) was noted to have a much faster flow (37.0 ml/min) as compared to that of the other hospital teats (H1-H3) with a range of 10.6 ml/min to 17.8 ml/min. The disposable hospital teat (H3) and commercially available teat (C6) having the same feed hole size under the same brand were found to have comparable flow rates. Variation of flow rates within a same teat was higher in disposable teats.

**Conclusions:** Flow rates of hospital teats are faster, of wider range and more variable compared with commercially available teats. Findings suggest inclusion of more teats for hospital use to provide options for slower milk flow and stable flow rate. They can also be used as a reference when identifying equipment to support feeding of infants who are hospitalised to maintain swallowing safety and facilitate feeding progression.

### Supraglottic Laryngoplasty for Moderate to Severe Laryngomalacia, Report from United Christian Hospital

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**Introduction and Aim:** Laryngomalacia is the most common cause of stridor in neonates and infants. While most can be managed conservatively, those with severe breathing or swallowing symptoms may require intervention. Non-invasive ventilation and feeding tubes are effective treatment, yet prolonging the hospital stay of these babies. Supraglottic laryngoplasty has been emerging as a surgical alternative, we are reviewing the results before our translocation of this service to the Hong Kong Children's Hospital.

**Methods:** 256 patients with laryngomalacia were evaluated as inpatient in the United Christian Hospital between 2010-2020, of which 51 received supraglottic laryngoplasty. The charts of these 51 patients were reviewed retrospectively. We reported on the patient's demography, comorbidities, symptoms, operative details and course of recovery. Factors influencing the outcomes of supraglottic laryngoplasty were calculated using statistical methods.

**Findings:** Supraglottic laryngoplasty was performed in 40 infants as sole surgery (single group) and as part of other airway surgery in 11 infants (combined group). Thirty-seven present babies were in the moderate laryngomalacia while the rest were suffering severe laryngomalacia. Fifty-eight percent patients reported synchronous airway lesion. This is significantly higher than those reported in literature. Within a month post-op, 92% of babies weaned from ventilation assistance, 72.5% weaned from feeding tubes, and 80.4% infants could be discharged home. Impact of individual outcome factors such as gestation, use of laser and medication use are calculated.

**Conclusion:** Supraglottic laryngoplasty is a safe and effective alternative for infants with moderate and severe laryngomalacia who were dependent on ventilation or feeding assistance.

### Behavioural and Emotional Outcomes in Children with Short Sleep Duration or Daytime Sleepiness

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**Background:** Sleep deprivation is common in local children. Many children also manifest with daytime sleepiness. However, how sleep duration and sleepiness relate to behavioural and emotional outcomes remains to be explored.

**Objective:** To investigate the relationship of short sleep duration and excessive daytime sleepiness (EDS) with neuro-behavioural outcomes in non-obese healthy children.

**Method:** This was a retrospective cross-sectional study. Children, aged 5-11 years old, were recruited. All subjects underwent evaluation including sleep history, overnight polysomnography, sleep diary and questionnaire. EDS was defined as a score of paediatrics daytime sleepiness scale (PDSS) >15. Short sleeper was defined as <9 hours of sleep per day based on the recommended sleep duration. Behavioural and emotional outcomes were measured with child behavioural checklist score (CBCL). Higher total score signifies poorer outcomes.

**Results:** 420 children (mean age of 8.7±1.6 years; 61% male) were analysed. One hundred and twenty-one were healthy controls, 100 were short sleepers, 103 were children with EDS and remaining 96 were combined group of shorter sleepers with EDS. There were significantly higher total scores in all sections of CBCL in the combined group of short sleepers (60.8±9.6) when compared to short sleepers (54.6±8.7) and EDS group (59.2±9.8) ($p<0.001$). Increased in PDSS scores was associated with increased in CBCL scores in a dose dependent relationship.

**Conclusion:** Adverse impact of daytime sleepiness on behavioural and emotional outcomes in children was observed, which highlights the importance of recognising daytime sleepiness and identifying the underlying causes to guide management.

### Surgical and Speech Outcomes of Surgical Treatment for Children with Velopharyngeal Dysfunction

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**Background:** Velopharyngeal dysfunction (VPD) refers to the inadequate separation of the oral and nasal cavities during speech and/or swallowing, and is commonly seen in patients with a repaired cleft palate or 22q11.2 deletion syndrome. VPD can give rise to significant speech impairments including hypernasality and audible nasal emission, while corrective surgery is the gold standard treatment.

**Purpose:** This study aimed to evaluate the surgical and speech outcomes in VPD corrective surgery done in the Hong Kong Children's Hospital – where the centralised and coordinated multidisciplinary care of children with complex medical conditions has been recently organised, for clinical audit and service planning.

**Methods:** A retrospective study was carried out to review the clinical presentations and the speech profile changes of patients who have undergone VPD surgery from January 2020 to February 2022. Patients' demographics and VPD corrective operation details including age at operation, surgical techniques, complications were charted from medical records. Findings of perceptual speech assessment using Cantonese Cleft Speech Assessment Tool by speech therapists were retrieved – ratings of different speech parameters before and 3-months after the surgery were compared.

**Findings:** Ten patients (3 male: 7 female) received VPD surgical treatment at the Hong Kong Children's Hospital in the study period. Their age at operation ranged from 3-year-old to 14-year-old. Seven (70%) of them suffered from velopharyngeal insufficiency related to the underlying cleft palate anomaly; while three (30%) of them had genetically confirmed 22q11.2 deletion syndrome and no cleft palate anomaly. Three surgical techniques were applied including posterior pharyngeal flap (60%), primary Furlow palatoplasty repair (20%) and revision Furlow palatoplasty repair (20%). There were no short and median term surgical complications such as surgical site dehiscence reported. As for speech outcomes, patients were found to have general improved ratings in speech parameters with significant decrease in hypernasality ($p<0.05$).

**Conclusions:** Our early data suggests that VPD surgical management in our centre is safe and effective in improving patient's speech profile, regardless of their different underlying pathophysiology. The isolated significant improvement in nasality could be explained by the majority of patients having mild to moderate velopharyngeal dysfunction at baseline. Future studies should expand the sample size, monitor the speech outcomes with a longer follow-up time and with respect to the later commencement of post-op speech therapy – to facilitate service planning.

### Evaluation of Intranasal Dexmedetomidine for Bedside Procedure Sedation at a Tertiary Paediatric Surgical Centre. And How Does it Compare to Oral Chloral Hydrate?

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**Background/Introduction:** Sedation is often required for paediatric bedside procedures. The traditional choice of chloral hydrate via oral route is often associated with side effects such as vomiting and desaturation. Intranasal dexmedetomidine is a new sedation agent shown in meta-analysis that has better side effect profile and tolerability.

**Purpose:** To assess the effectiveness of intranasal dexmedetomidine for paediatric surgical bedside procedures in a tertiary paediatric surgical centre.

**Methods:** Retrospective observational cohort study of consecutive patients with bedside procedure performed requiring sedation between February 2020 to September

2022 was done. Demographic data collected include age, body weight and gender. Primary outcome was sedation effectiveness, procedure completion rate and safety profile of intranasal dexmedetomidine. Secondary outcome was the difference in sedation effectiveness of intranasal dexmedetomidine compared to a historical cohort of patients that had oral chloral hydrate use. Statistical analysis was performed using SPSS (version 28.0), means were compared with independent t test and categorical data compared using chi-square and fisher-exact tests.

**Findings:** 145 patients were included (male 127, female 18), with mean age 3.6-year-old. 108 patients had intranasal dexmedetomidine and 37 had oral chloral hydrate. Procedures requiring sedation include penile dressing management (71%, n=103), incision and drainage of abscesses (9.7%, n=14), removal of sutures/drains (6.9%, n=10), scald wound debridement (4.8%, n=7), groin examination/hernia reduction (3.4%, n=5), steroid injection into keloid (2.1%, n=3) and suction rectal biopsy (2.1%, n=3). Dexmedetomidine was effective in 80.6% (n=87) (Group A) and unsuccessful in 19.4% (n=21) (Group B). No statistical difference was noted between Group A and B regarding to age groups and types of procedure. Procedure was successfully completed in 98.1% (n=106). Overall no complication was noted. The sedation effectiveness of intranasal dexmedetomidine is 80.6% vs 67.6% (p=0.116) compared to chloral hydrate.

**Conclusion:** Intranasal Dexmedetomidine is an effective and safe alternative sedation agent for different paediatric surgical bedside procedures.

## Poster Presentation

### Effects of Insomnia and Obstructive Sleep Apnoea on Blood Pressure in Children and Adolescents

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**Background:** Obstructive sleep apnoea (OSA) is associated with neurocognitive and metabolic disturbances in children. Chronic insomnia is associated with hypertension in adult.

**Purpose:** To investigate the associations of sleep-disordered breathing (SDB) and insomnia symptoms with ambulatory blood pressure (ABP) in non-obese children and adolescents.

**Methods:** This study was a retrospective analysis of a hospital-based cohort. Each participant completed a sleep questionnaire, underwent overnight polysomnography and 24-hour ABP monitoring.

**Findings:** 747 subjects were included (mean age 11.0 ± standard deviation (SD) 2.5 years; 62% male), with 29 healthy controls, 261 snorers, 36 subjects with insomnia symptoms, 379 OSA, and 42 with combined insomnia and OSA. There was significantly higher diastolic blood pressure (DBP) z-score in snoring (mean 0.71±SD 0.87), OSA (0.89±0.95), insomnia (0.99±0.92) and combined groups (1.04±0.96) compared to controls (0.34±0.87) ($p<0.05$). The linear means plot of the DBP z-score across the different groups had a p-value of <0.001. There was an increasing proportion of diastolic non-dippers (<10% BP dip from wakefulness to sleep), prehypertensives, and hypertensives from controls to the combined group ($p<0.05$).

**Conclusions:** A dose-dependent relationship with DBP z-score is demonstrated across the spectrum of sleep disorders. Both childhood insomnia and SDB are associated with a higher nocturnal DBP z-score with the highest score in the children who had combined insomnia and OSA. Tackling paediatric sleep problems may prevent the future development of cardiovascular diseases.

### The Effects of Type of Feeding, Mode of Delivery, and Antibiotics Use on the Preterm Gut Microbiota

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**Introduction:** Gut microbiota carries out multiple functions including metabolism, trophic activities, and protection. Preterm infants are particularly susceptible to dysbiosis, predisposing them to a myriad of conditions later in life.

**Purpose:** To explore the effects of type of feeding, mode of delivery, and antibiotics use on preterm gut microbiota.

**Methods:** 161 cases were included in this study. Samples taken at birth and at discharge were divided into groups for type of feeding (mainly breastfed, mixed, mainly formula fed), mode of delivery (vaginal delivery and caesarean section), and use of antibiotics (antibiotics use and no antibiotics use). Microbiome Analyst was used for data analysis.

**Findings:** At discharge, relative abundances showed greater diversity for groups with mixed feeding, vaginal delivery, and no antibiotics use. Relative abundances of Bifidobacterium were greater in groups with mainly breastfeeding, vaginal delivery, and no antibiotics use. Shannon index was statistically significant at birth for the mixed group compared to the mainly breastfed group, and the caesarean section group compared to the vaginal delivery group.

**Conclusions:** Mixed feeding, vaginal delivery and no antibiotics use showed to have contributions to gut microbiota diversity. Certain deviations from literature, such as an increased Shannon index at birth in the caesarean section group and the mixed feeding group, affirm the presence of inter-individual variability in preterm infants. As mixed feeding is common in preterm infants and has been shown here to increase gut microbiota diversity, the benefits of combining formula and breastmilk on the health of preterm infants should be further explored.

### Harvesting Stem Cells from Donors with Abnormal Red Blood Cells in the Paediatric Setting

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**Introduction:** The apheresis operator adjusts the level of cell collection based on the information provided by the automated interface management (AIM) system which reads the darkness of the cells of the apheresis machine. Therefore, the collection efficiency of the product could be affected and poor product yield can be obtained if the AIM system information to the operator is incorrect and misinterprets the darkness of cells. Two cases of stem cell harvest from donors who had aberrant red blood cells done in Hong Kong Children's Hospital will like to be reported.

**Results and Outcomes:** An adult donor of a 3-year-old child was found to have Haemoglobin H disease (HbH) with microcytic anaemia (mean corpuscular volume (MCV) = 54.8 fL, normal range: 80.9-93.9 fL) during donor workup. Studies revealed that donors with low MCV often had poorer CD34+ stem cell harvest yields. After consulting the medical team and the manufacturer, we transfused the red cell to top up the haemoglobin of the donor one day before the harvest. We attempted to collect the stem cell from this donor by lowering the collection preference to a deeper location with a colour algorithm of 3-4% and raising the centrifugation speed but the rate was ultimately limited by the size of venous access. Although the product yield was sufficient for the child ($10x10^6$/kg CD34+), it was less than half of the predicted calculated CD34+ cell yield.

A 9-year-old child with beta-thalassaemia major underwent bone marrow backup harvest before a haploidentical PBSC transplant and bone marrow processing was done to reduce the volume via plasma depletion before cryopreservation. To satisfy the requirements of the apheresis machines, an additional irradiated packed cell from a random donor was placed in the BMP bag before the processing. Unexpectedly, the product yield was only $0.64x10^6$ CD34+ cells/kg. Learning from the previous case and after discussing with the medical team, $3.89x10^6$ CD34+ cells/kg was achieved in the repeated processing by collecting at a darker colour of 5-6% according to the colour algorithm.

**Conclusion:** The product yield is influenced by the characteristics of other cells, including red blood cells, in addition to the CD34+ count. To maximise the harvest of peripheral blood or bone marrow stem cell from donors with aberrant RBC, changes to the apheresis parameters are required.

### Are We Compressing and Ventilating Correctly During Cardiopulmonary Resuscitation?

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**Background:** Acute care physicians should be proficient in the resuscitation of any individual including the child. Well established and scientific training programs are available that requires annual or bi-annual recertification in most countries, such as Pediatric Advanced Life support (PALS), and Basic Life Support (BLS). The key elements of a successful CPR are adequate and effective compressions and ventilation.

**Aim:** We audit the effectiveness of CPR performance of students during PALS practice.

**Methods:** Using Gaumard Pediatric HAL®S3004 pediatric simulator, the effective of CPR performance of students during PALS practice. Cardiac compression rate and depth, and ventilation rate, Positive Inspiratory Pressure and inspiratory time were recorded.

**Results:** 34 CPR practice (26 Nurses & 8 doctors; Female: 28, Male: 6) were audited at the Simulation Training Centre, HKCH during 3 PALS course days, July 2022 – August 2022. There is no difference between the doctors versus the nurses in performing CPR, but females appeared to underperformed in ventilation than males. Significantly, effective ventilation was not observed or registered by the simulator in 14 of the 34 students (41% of students). These preliminary observations need validation in prospective research setting.

**Conclusions:** The findings are alarming and serve to remind that ineffective ventilation may also occur in real-life situations, which may jeopardise patient safety. This is especially relevant in paediatric practice as cardiopulmonary failure are primarily respiratory.

### Local Data of Children with Visual Impairment (VI) at Child Assessment Service, Department of Health

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**Background:** It is estimated that at least 2.2 billion people around the world have Visual Impairment (VI). In Hong Kong, according to the latest data from the Census and Statistics Department in 2021, 3100 children under 15 years old had VI. The multidisciplinary assessment team at CAS provides comprehensive developmental assessments, recommendations on rehabilitation service referrals and interim parental support.

**Purpose:** To review the epidemiological data of children with VI at the CAS.

**Methods:** CAS data on children diagnosed with severe visual impairment (VA between 3/60 and 6/60) and blindness (VA from no light perception to 3/60) according to ICD-10 categorisation were retrieved from 2013 to 2018. Incidence, causes and comorbidities of these children at our service were analysed.

**Findings:** During 2013-2018, the CAS has assessed 201 children with VI, 135 (67.2%) had severe VI and blindness. The number of new severe VI/blind cases at the CAS was 15 to 30 per year. Most of these children were referred to our centres before the age of two. In our setting, the major causes of severe VI/blindness were cerebral visual impairment (63.7%), retinal disorders (15.6%) and disorder of the whole globe and anterior segment (10.4%). 73.3% of those with severe VI/blind had one or more non-ophthalmological disorders or disabilities, namely intellectual disability, epilepsy, cerebral palsy, etc.

**Conclusion:** In CAS setting, the major causes of severe VI/blindness are CVI and retinal disorders. Most of them also suffer from non-ophthalmological disorder or disabilities. Multidisciplinary team assessment and management is necessary in view of their complex needs and heterogenous conditions.

### A Complex Case of Perioperative Anaphylaxis and the First Successful Desensitisation To a Neuromuscular Blocking Agents

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**Introduction:** Perioperative anaphylaxis is rare yet life-threatening. Neuromuscular-blocking-agents (NMBAs) are common culprits and cross-sensitivity among different NMBAs exists, whether due to IgE or MRGPRX2 mechanisms. While culprit drugs should be avoided in future anesthesia, it may not be possible for patients sensitised to multiple agents. We report the first cisatracurium desensitisation for a series of technically challenging surgeries in a complex syndromic patient with multiple NMBAs allergy.

YKT was born with CHARGE syndrome requiring multiple operations since birth. At 3-year of age, he developed anaphylactic shock during his seventh operation under general anaesthesia. Skin-tests, basophil activation tests and subsequent drug challenges were negative to other medications but confirmed allergy to all NMBAs available locally. Muscle relaxation was critical for perioperative care of the upcoming multi-staged surgeries for mandibular distraction, bone-anchored hearing-aid placement, and cleft-palate repair.

**Method:** Transdisciplinary planning took place collaborating Harvard Medical School team to elaborate the first cisatracurium desensitisation protocol. Patient was premedicated with Omalizumab two weeks and one day before desensitisation. Cetirizine, famotidine and montelukast were started 30 minutes before desensitisation. A prolonged 12 steps protocol targeting the optimal cisatracurium dose to maintain muscle relaxation during and after surgery was implemented successfully.

**Finding:** Desensitisation procedure was performed smoothly. Surgical procedures were done safely and without complications and the patient recovered uneventfully.

**Conclusion:** NMBA sensitisation either through IgE or MRGPRX2 mechanisms may be difficult to address when non-cross-reactive drugs are unavailable. Desensitisation can be useful and successful in sensitised patients in need of muscle relaxation for optimal surgery.

### Nursing Enhancement Programme on Prevention of Unplanned Extubation

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**Background:** Patients with endotracheal intubation are common in paediatric intensive care units (PICU). However, unplanned extubation (UE) is one of the most frequent complications associated with airway management. It may lead to serious complications, or even death.

**Purpose:** The aim of this project were to：(1) identify contributing factors of unplanned extubation in a PICU of a regional hospital; (2) enhance nurses' awareness in the prevention of unplanned extubation and to improve the quality of care to intubated patients.

**Methodology:** Data of the past UE events during 2017 to 2020 in the PICU of a regional hospital was collected. The contributing factors of these UE incidents were identified and analysed. A nursing enhancement program was developed and incorporated with the findings and proposed preventive measures. All nurse of this PICU were recruited in the training. The nurses' knowledge was evaluated by a questionnaire conducted before and after training.

**Result:** The nurses performed better in the identical post-test questionnaire. Furthermore, there is no more UE event in PICU for 2 years after the training programme.

**Conclusion:** Unplanned extubation may cause several adverse consequences, but it is preventable. The education programme could reinforce the knowledge and increase the awareness of nurses.

### Severe Hyponatraemia in an Infant with a Urinary Tract Infection

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**Introduction:** Hyponatraemia with hyperkalaemia is an uncommon finding in infancy but represents a medical emergency. Although congenital adrenal hyperplasia (CAH) remains the most common cause, it is pivotal to consider other adrenal causes such as pseudohypoaldosteronism (PHA). We report an infant presenting with features of PHA in the setting of a urinary tract infection.

**Purpose:** We aim to highlight the association of transient type I PHA with urinary tract anomalies or infections, shedding light on the clues for the diagnosis and the implications for management.

**Method:** Clinical data was gathered from a case admitted to our intensive care unit.

**Findings:** A 6-month-old boy with a good past health presented with failure to thrive, emesis and dehydration. Investigations showed severe hyponatraemia, hyperkalaemia and a mixed high and normal anion-gap metabolic acidosis. Serum creatinine and urea were mildly elevated, and lactate was normal. CAH profile was normal. Serum renin and aldosterone were markedly elevated, which together with the low transtubular potassium gradient and high fraction excretion of sodium implied aldosterone resistance. Catheterised urine grew *Escherichia coli*. Right hydroureteronephrosis was seen on ultrasonography, but there was no obstruction or vesicoureteral reflux. He was treated with fluid resuscitation and a course of intravenous antibiotics with good response, evident by the normalised electrolytes, blood gas, serum renin and aldosterone. Type I PHA is caused by aldosterone resistance. The transient form entails a distinct pathophysiology in which renal tubular immaturity may play a role, given its prevalence in younger infants. Early diagnosis hinges on the recognition of the typical laboratory abnormalities and the exclusion of the more common CAH. Management is supportive.

**Conclusion:** Evaluation of young infants with urinary tract anomalies or infections should include serum electrolytes and blood gases, given the possibility of transient secondary pseudohypoaldosteronism.

### "SH... PLEASE BE QUIET" – The Effectiveness of Educational Talk on Nurses' Awareness on Noise Reduction in Neonatal Intensive Care Unit

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**Introduction:** The preemies stayed in the neonatal intensive care unit (NICU) for the first weeks to months of their lives. The high (>45db(A)) noise levels in a local Level III NICU were identified and this leads to the adverse effect on physical changes and brain development of patients. This project aimed to improve the environment - through noise reduction in NICU.

**Objectives:** The objective was to increase nurses' awareness of noise reduction after the educational talk for critically ill patients.

**Methodology:** A convenience sample consisted of forty-one nurses working in a Level III NICU in Kowloon East Cluster. The participants completed the validated questionnaire before (pre-test) and after (post-test) an educational talk conducted by the team members.

Quantitative and descriptive statistics were used to analyse the data, and all were transcribed onto Fisher's exact test using SCIPY.

**Results:** A total of 41 (100%) questionnaires were collected. The findings demonstrated that the noise level was reduced to 20-29% in NICU. The mean score of awareness of noise level in the post-test (9.88±1.33) was higher than the pretest (5±2.77) ($P<0.001$). Moreover, the participants indicated that they had a positive behavioural change on noise reduction, initiated noise-reducing interventions, and recognised their vital role in providing an optimal environment for the growth and development of the patients.

**Conclusions:** This project achieved a significant increase in nurses' awareness of the importance of noise reduction in the NICU. In addition, this acts as a precursor for evidence-based nursing practice to provide a "quieter" environment for vulnerable patients on the way forward.

### Home Scene Reconstruction in COVID-19 Era Rehabilitation - From Virtual to Reality

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**Introduction:** Home discharge is the ultimate goal for patients, and is facilitated by medical teams and physiotherapists. Under the COVID-19 pandemic, Hong Kong Children's Hospital physiotherapists adopted the concept of Virtual Tour to rebuild home scenes within the physiotherapy department for home environment-targeted precision training.

**Purpose:** This report aims to introduce a new method of virtual home visit under the COVID-19 pandemic as an alternative to conventional home visit to minimise infection risk while maintaining the quality of physiotherapy service.

**Methods:** A virtual home visit was carried out for a 13-year-old patient with rheumatology disease who had been hospitalised for three months. Discussion was carried out among the patient, the caretaker and physiotherapists regarding patient's daily needs. The caretaker was invited to prepare a video of their home. Physiotherapists reviewed the video for accessibility assessment, home scene reconstruction within the physiotherapy department and treatment planning. Simulated training on self-care and safety were performed for 10 sessions. Patient and carer satisfaction level on a numerical rating scale of 0 to 10 were measured post-discharge.

**Findings:** The patient and caretaker acquired proper handling skills and felt competent to return home in a month's time. Both reported the home return to be smooth and safe. Overall satisfaction level for the simulated training was reported to be 9 and 10 by both.

**Conclusion:** The advantages of virtual home visit included infection control, manpower optimisation and the conferral of active role to caretaker. With the employment of technology, discharge home planning was not restricted by infection control concerns.

## MOG-IgG Associated Brainstem Encephalitis in a Chinese Boy: Complication of *Mycoplasma pneumoniae* Infection

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**Background:** MOG-IgG associated disorder (MOGAD) is a rare disease entity in paediatric population. It is recently recognised as a distinct clinical entity due to the emergence of highly specific cell-based assay. MOG antibody was found to be encephalitogenic and a wide range of clinical manifestations have been described, but brainstem manifestation was infrequent at any age. On the other hand, despite *Mycoplasma infection* is well known to be complicated with encephalitis, its relationship with MOGAD is not well reported. Moreover, the optimal acute treatment of MOGAD in children is still lacking evidence.

**Case Description:** We reported a 5-year-old Chinese boy presented with slurred speech, dysphagia and unsteady gait, 10 days after *Mycoplasma pneumoniae* infection. Physical examination revealed patient had slurred speech with associated gurgling sound. Neurological examination of the cranial nerves, upper limbs and lower limbs were unremarkable. Magnetic resonance imaging (MRI) brain showed fluffy T2 hyper-intense signals over the brainstem. Serum anti-MOG was positive. He was diagnosed to have MOG-associated brainstem encephalitis, as sequelae of Mycoplasma pneumoniae infection. Patient was started on intravenous cefotaxime and intravenous immunoglobulin (IVIg). He showed good response to treatment that his symptoms gradually improved. There is no recurrence of symptoms and follow-up MRI brain 6 months later showed resolution of brainstem encephalitic change.

**Conclusion:** Our case demonstrated that MOGAD can be a complication of Mycoplasma infection. Also, we highlighted the potential use of IVIg as first line treatment during acute phase of MOGAD.

## Scenario-based Training on Intravenous Chemotherapy Administration for Paediatric Oncology Nurses

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**Introduction:** Chemotherapy medication incidents ranked high in hospital. There are various factors in occurrence of medication incidents. A scenario-based training on administration of intravenous (IV) chemotherapy was implemented to all nurses who were working in the unit and found effective to improve compliance rate in medication administration.

**Method:** Chemotherapy protocol with twelve scenarios would be randomly assigned to each nurse. They had to perform the administration of intravenous chemotherapy as usual as the unit guideline. At the end of the training session, a debriefing and evaluation would be carried out. A satisfaction survey was distributed to the participants to rate the training programme.

**Results:** The scenario-based training gained over 95% compliance in administration of IV chemotherapy, and a high satisfactory score among the participants. There was a decreasing trend of medication incidents related to chemotherapy after the training programme was implemented. The result also showed nurses with less clinical experience had lower compliance in the assessment. Clinical experience with 0-3 years scored compliance rate 96.1% which with over 3 years scored 99.2-100%.

**Conclusion:** Medication safety is a part of the daily routine nursing care requiring a high standard of practice. There were various factors affecting the quality and safety of health care. It is necessary to undertake a new training method to nurses so as to enhance memory and improve situation awareness of nurses. Further, it may be effective to reduce medication incidents.

### An Uncommon Cause of Haemoptysis – Rasmussen's Aneurysm with Cavitary Tuberculosis

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Haemoptysis is a common presentation of tuberculosis. Rasmussen's aneurysm, an aneurysm arising from the pulmonary artery secondary to cavitary tuberculosis infection, is a rare yet fatal cause of massive hemoptysis. Early identification and timely intervention are of utmost importance to reduce modality. We present a case of a 14-year-old girl who presented with persistent fever, cough and massive haemoptysis which required intubation and intensive care, later confirmed to be pulmonary tuberculosis. Computed tomography (CT) of the thorax and pulmonary angiogram showed extensive pulmonary tuberculosis infection with necrotising pneumonia and lung abscess, also the presence of pseudo-aneurysm in the left upper and lower lobe in which oriented from the left pulmonary artery. The one over the lower lobe was successfully embolized, which was followed by a significant improvement in the severity and the frequency of hemoptysis. She was also treated with anti-tuberculous therapy for more than one year with no recurrence of haemoptysis. Follow-up CT thorax demonstrated extensive destruction of the left lung. To conclude, Rasmussen's pseudo-aneurysm is an important vascular complication of cavitary tuberculosis and need to be considered as one of the differential diagnosis in patients present with pulmonary tuberculosis and massive haemoptysis.

### An Image Protocol to Validate ASAP (Airway Sonographic Aiding Pad)

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**Objective:** Sonographic assessment of the upper airway is highly operator dependent in children due to movement of the child, softness of the laryngeal cartilage framework, the curved anterior surface of the neck, and air blocking transmission of ultrasound signals. Our project aims to facilitate operators to perform airway ultrasound by inventing a physical aid.

**Methods:** We have designed and manufactured Airway Sonographic Aiding Pad (ASAP) and we are reporting its pilot validation test results. The ASAP was engineered from a block of sonographic hydrogel using 3D printing technique. It has a flat surface anteriorly to engage the ultrasound probe, while its curved undersurface obliterate airspace between the probe and subject's neck. An imaging protocol was developed to quantify the standard of airway ultrasound images. A total of 10 operators (4 experienced, 6 inexperienced) were recruited to perform airway ultrasound on a paediatric volunteer. They were randomised to perform the examination with ASAP or with standard hydrogel first. The time taken to complete the set of examination was recorded, and user experience was documented by questionnaires. The images were then presented to a blinded third-party reviewer to compare the image quality.

**Results and Conclusion:** ASAP enables both experienced and inexperienced operators to perform ultrasonographic assessment of upper airway in children more swiftly and confidently. The effectiveness brought about by ASAP is more obvious with inexperienced users. Sonographic images obtained with ASAP are of higher clarity. It can be a useful tool for training and clinical practice.

## Paediatric Anaphylaxis Management in Schools – Current Issues and Challenges in Hong Kong. A Scoping Review

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**Background:** Hong Kong has the highest prevalence of food allergies compared to Mainland China, Russia, and India. There has been a twofold increase in anaphylaxis incidence between 2009-2019, of which 20% occur in day-care and school settings.

**Objectives:** A scoping systemic search was performed with the aim of reviewing existing literature in the Asia-Pacific region regarding food allergy management in the school setting. Current loopholes and inadequacies on governmental policy regarding school anaphylaxis management were explored.

**Methodology:** The search in Medline & EMBASE via OVID revealed 195 registers. Results were later compared with a PRISMA scoping review published in 2022 having similar search terms but focusing on Canada, America, Australia, and European countries. Furthermore, current loopholes and inadequacies on governmental policy regarding school anaphylaxis management in Hong Kong were explored. An internet search was later conducted to supplement the information on governmental policies for school anaphylaxis management.

**Results:** Most publications identified focused on assessing food allergy prevalence and causative agents. However, there is an evident lack of literature on emergency action plans and school training programmes.

Existing governmental policies regarding school anaphylaxis were reviewed and compared. Hong Kong currently lacks legal protection for bystanders and policies encouraging school staff training for anaphylaxis management. Governmental regulations and subsidisation are also absent in encouraging schools to purchase back-up stocks of unassigned epinephrine autoinjectors.

**Conclusion:** Raising awareness, improving guidelines and policies in schools are integral in the management of food-induced adverse events and anaphylaxis. Governmental support through policymaking and legislation can significantly enhance and hasten the process, thus minimising the impact adverse food reactions bring to the paediatric population.

## The 'PREFER' Elements of Rehabilitation in PICU Liberation Bundle

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**Background:** Despite improved survival rates in critically ill children, admission to a Paediatric Intensive Care Unit (PICU) can be physically and emotionally stressful for children. As such, the Society of Critical Care Medicine launched the PICU liberation bundle campaign, which aims to free patients from the harmful effects of pain, sedation, delirium, immobility, and sleep disruption in PICU. However, there is no consensus on the elements of rehabilitation to promote patient recovery.

**Purpose:** Identifying rehabilitation elements in the PICU liberation bundle and assessing clinician consensus

**Methods:** A three-step modified Delphi method was used to establish consensus. In step 1, the elements of rehabilitation were identified via literature search including **P**ain management (P), **R**espiratory optimisation (R), early **E**xercises and mobilisation (E), **F**amily engagement (F), **E**mpowerment of patients (E) and **R**ealisation of rehabilitation goals among clinical team, patients and family (R), which were referred to as **'PREFER'**. In step 2, clinicians were asked to complete an online survey to rank their level of agreement (LOA) (1-9; 1=totally disagree, 9=totally agree) and level of importance (LOI) (1-9; 1=least important, 9=most important) on including these rehabilitation elements as the role of Physiotherapists in the PICU. In step 3, the findings were summarised and discussed among the panel members.

**Findings:** Forty-five clinicians completed the survey (44.4% Physiotherapists, 22.2% Medical officers & 33.3% Nurses; Years of PICU experience: majority 3-9 years: 62.5%). The following were the results (mean / SD): Pain management: (LOA: 8.0.9±1.1 /LOI: 7.6±1.4), Respiratory optimisation (LOA: 8.8±1.1/LOI: 7.9±1.0), early Exercises and mobilisation (LOA: 8.9±0.6 /LOI: 8.9±0.5), Family engagement (LOA: 8.4±1.2/LOI: 8.0±1.3), Empowerment of patients (LOA: 8.3±1.2/LOI: 8.1±1.3) and Realisation of rehabilitation goals among clinical team, patients and families (LOA: 8.2±1.1/ LOI:8.2±1.2). The panel members reached an agreement on the important rehabilitation elements of the PICU liberation bundle.

**Conclusions:** This abstract describes the development of a new rehabilitation PICU liberation bundle - PREFER through the use of a consensus approach. The implementation strategies and clinical care pathways could be developed based on these identified elements in the future.

### Infant's Gut Microbiome is Associated with Brief Infant Sleep Questionnaire Outcomes at 1 and 4 Months of Age

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**Background:** Both sleep and gut microbiome (GM) develop rapidly during infancy. To our knowledge, no prior studies have examined the association between GM and sleep during the first few months of life. In this analysis, we examined the association between 16 infants' GM at 1 month old and their concurrent and 4-month-old sleep outcomes.

**Methods:** We included a subset of 16 infant-mother pairs who were recruited postnatally at the Prince of Wales Hospital to participate the CUHK Stanley Ho Developmental Cohort Study. All subjects have completed the Brief Infant Sleep Questionnaire (BISQ) at 4-month-old and have provided at least one stool samples at 1- or 4-month-old. We characterized the gut microbiome composition by performing 16S rRNA sequencing on the collected samples.

**Results:** Higher richness in GM is associated with the concurrent sleep problem, nocturnal sleep duration (NSD), and crying problem before sleep at 4 months old. A lower proportion of Bacteroides was found in infants with crying problem before sleep and a lower abundance of Corynebacterium_1 and Bacteroidales in infants with longer settling time. Longer nocturnal wakefulness was associated with a higher abundance of Rothia. At 1 month, infants with short daytime sleep and total sleep duration have a higher abundance of Enterococcus while those with short NSD have a lower abundance of Parabacteroides and a higher abundance of Lactobacillales. At 4 months old, infants with longer and more frequent nocturnal wakefulness have a lower abundance of Rothia and a lower abundance of Collinsella. Longer settling time is associated with lower abundances of Veillonella.

**Conclusions:** GM at 1 month old is associated with concurrent and future sleep outcomes. At 1 month, the GM of short sleepers is different from those with normal sleep duration, and this association is attenuated but persisted to 4 months old. Increased abundance of species from Firmicutes and a decrease in the abundance of species from Bacteroidetes are found in poor sleep infants.

### Iron Deficiency (ID) and Its Impact on Fatigue and Attention Function Among School-aged Adolescents in Hong Kong

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**Purpose:** Studies have shown that ID may manifest as mild cognitive impairment, fatigue and poor school performance among school-aged adolescents. This study aimed to evaluate the association of iron status with attention function and fatigue in this population.

**Methods:** We recruited 183 boys and 340 girls (mean age=17.55, SD=1.05 years) from 16 schools through the Red Cross blood donation campaigns. Their serum ferritin (Fe) levels and complete blood counts were measured. ID is defined as Fe <30 pmol/L. They completed the Conners Continuous Performance Test-III to assess impairments in 3 attention domains (impulsivity, inattention and sustained attention) and the PEDsQL Multidimensional Fatigue Scale. Multivariable modeling was conducted to evaluate the association between Fe and attention/fatigue, adjusting for age and sex.

**Findings:** The overall prevalence of ID was 9.9%. None of the boys had ID. Among girls, the rate of ID was 15.2%. Overall, 249 adolescents (47.6%) had normal attention function, while a minority showed moderate to severe impairment in impulsivity (30.0%), inattention (27.6%) and sustained attention (24.9%). Adolescents who demonstrated sustained attention impairment had significantly lower Fe than adolescents with intact attention function (median [IQR] 115 [61-240] pmol/L vs 166 [85-310] pmol/L; P=0.0020). Trends were also observed when comparing Fe levels in adolescents with intact attention function with adolescents with impulsivity (P=0.067) and inattention (P=0.014) impairment. Multivariable analysis showed that every 10 units increase in Fe was associated with a 1.8% decrease in the risk of

sustained attention impairment (RR=0.82; 95% Cl=0.73-0.94; P=0.040). There was also significant association between lower Fe and general fatigue (Est= 0.0092, SE=0.0038; P=0.016).

**Conclusions:** Low iron status is significantly associated with sustained attention impairment and fatigue, with trends suggesting its adverse effect on impulsivity and inattention too. As ID is highly correctable with oral iron supplements and an iron-fortified diet, the identification of adolescents affected by ID is an important public health issue.

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### The Use of Complementary Medicine in Chinese Paediatric Patients Receiving Palliative Care: A Multi-Centre Study

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**Background:** Children receiving palliative care face unique physical and psychosocial needs. Parents may turn to complementary medicines (CM); however, related studies are limited.

**Purpose:** This study examined the pattern of CM use among paediatric patients receiving palliative care.

**Methods:** Between April 2021 and June 2022, parents of patients (<19 years old) receiving palliative care were recruited from 3 hospitals in Hong Kong. They completed a structured survey on sociodemographic data, CM use, the child's symptom burden (Rotterdam Symptoms Checklist) and TCIM integration. Clinical data were collected from health records. Differences in characteristics were compared among CM users and non-users.

**Findings:** We recruited 61 patients (60.7% male; mean age=10.4 years). Overall, 68.9% reported CM use, most commonly Chinese herbal medicine (39.3%) and massage/ TuiNa (34.4%). Non-oral CM approaches, especially acupuncture, were less popular among patients with cancer than non-cancer patients (29.2% vs 64.9%, p=0.01). Among CM users, patients with cancer had higher physical symptom distress than non-cancer users (22.0 vs 17.3, p<0.01). More parents of children with cancer consulted their doctors before using CM than non-cancer users (73.3% vs 33.3% p=0.03), and considered CM to be effective in relieving nausea, vomiting and indigestion symptoms (p<0.05). Most parents (73%) spent ≥HKD$1000 (USD 130) on CM monthly. The most common reason for not using CM was the lack of information (57.9%). Multivariable analyses showed that older parents (adjusted odds ratio[aOR]=1.15, 95%CI=1.02-1.31) were more likely to use CM, while children who received bone marrow transplant were less likely to use CM (aOR=0.16, 95%CI=0.03-0.73). Most parents (82.5%) supported CM integration, especially Chinese medicine and acupuncture, into palliative care service. Qualitative analysis of the structured interviews suggested that parents had difficulty finding reliable CM providers and were concerned about the cost of CM. They also encouraged early initiation of CM services.

**Conclusions:** A significant proportion of parents expressed interest in CM. Policy makers should consider multidisciplinary and collaborative integration of CM in routine palliative care.

### A Case of *FGFR1*-Related Disorder with Hypogonadotropic Hypogonadism and Split Hand/Foot Malformation

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**Background:** Concomitant hypogonadotropic hypogonadism and split hand/foot malformation are rare conditions and to date, there is only one overseas' case series reporting 8 patients with such condition. Among them, 7 (88%) had a *FGFR1* loss-of-function variant.

**Clinical report:** Our patient is a 20-year-old gentleman born full-term of a non-consanguineous Chinese couple with a birth weight of 3.25 kg in Shanghai. Antenatal history was unremarkable. He was noted to have right hand ectrodactyly and oligodactyly; left hand cutaneous syndactyly involving 3rd and 4th digits; bilateral syndactyly between hallux and 2nd toe; and right foot mesoaxial polydactyly. Orthopaedic surgery was later performed to separate left middle and ring fingers. He also

had micropenis. Karyotype done in Shanghai was normal. He later came to Hong Kong and was found to have short stature and delayed puberty by Student Health Service and subsequently referred to paediatricians. Further investigations revealed hypogonadotropic hypogonadism and testosterone injection was commenced since 15 years old. Other hormonal axes were normal and MRI pituitary showed no focal lesion. His development was normal all along with no learning difficulties. There was no family history of short stature or delayed puberty and the mid-parental height was 165cm. On examination, he had bushy eyebrows with synophrys, short nose, mid-face hypoplasia, hypodontia with one absent lower incisor, low set ears and smooth philtrum. Whole exome sequencing was performed with peripheral blood and found a heterozygous missense variant in the *FGFR1* gene [NM_001174067.1(*FGFR1*):c.662G>C p.(Trp221Ser)]. This variant has not been reported in literature or public databases, but parental testing showed that the variant was de novo. By ACMG guideline, this variant was classified as likely pathogenic.

**Conclusions:** We reported a Chinese patient with hypogonadotropic hypogonadism and split hand/foot malformation who has a novel *FGFR1* variant. This echoes overseas' findings and reinforces the importance of considering *FGFR1*-related autosomal dominant genetic disorder when encountering patients with both hypogonadotropic hypogonadism and split hand/foot malformation.

### Level of Urinary Catecholamine in Children with Sleep Disordered Breathing

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**Background:** The role of catecholamines in Sleep Disordered Breathing (SDB) has been postulated by several studies. However, whether there is a difference in catecholamine levels in children with and without SDB remains unclear.

**Purpose:** To compare the levels of different urinary catecholamines amongst paediatric patients with and without SDB as well as across the severity spectrum.

**Methods:** Literature searches were conducted on PubMed and EMBASE until 25/06/2022. Inclusion criteria were original human studies, English language, paediatric subjects diagnosed with SDB/Obstructive Sleep Apnoea (OSA). The main outcome measured was standardised mean difference (SMD) of urinary catecholamine between patients with and without SDB, as well as OSA and controls.

**Findings:** 9 studies with a total of 838 subjects were included in the quantitative analysis. Urinary levels of noradrenaline and adrenaline were higher in children with OSA: SMD=1.45(95%CI=0.91-2.00; I2=75%, P<0.001); SMD=1.84(0.00-3.67; I2=97%, P=0.05). Urine level of noradrenaline was higher in all SDB patients, including Primary Snoring (PS): SMD=0.86 (95%CI=0.32-1.41; I2=85%, P=0.002), and in subjects with moderate/severe OSA compared to the milder form: SMD=0.55 (95%CI=0.10-1.00; I2=0%, P=0.02). Urinary dopamine was not associated with SDB regardless of severity.

**Conclusions:** Urinary adrenaline was higher in OSA patients. Urinary noradrenaline was higher in all SDB patients including PS and could reflect OSA severity in children. Hence, both noradrenaline and adrenaline may be markers of sympathetic overtone in SDB patients and could potentially act as surrogate markers for the associated complications. Further studies are needed to assess this association.

### Age and Gender-specific Flow-mediated Dilation Reference Values for Children and Adolescents from a Large Community-based Study

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**Background and Purpose:** To establish normative reference values of brachial artery flow-mediated dilation (FMD) response by age and gender in children and adolescents, and to identify predictors of FMD in this population.

**Methods:** A representative sample of 1498 healthy children and adolescents aged 8 to 17 years was recruited from schools in Hong Kong. Subjects underwent sonographic brachial artery assessment and blood sampling for glucose and lipid profile. Smoothed gender-specific FMD percentile curves were constructed using the Lambda-Mu-Sigma (LMS) method. Predictive factors of FMD were identified using linear regression analysis.

**Findings:** Mean FMD among healthy children and adolescents in the community setting was 8.57± 0.90%. Smoothed gender-specific FMD in centiles were constructed as a reference benchmark. Regression analysis after adjustment for age, gender, body mass index (BMI) z-score, and baseline artery diameter, when applicable, demonstrated that FMD is positively correlated with age (β=0.142, p=<0.001) and high density lipoprotein (HDL) (β=0.103, p=0.001), while negatively correlated with baseline artery diameter (β=-0.117, p=0.001), diastolic blood pressure (DBP) (β=-0.053, p=0.047), glucose (β=-0.091, p=0.004) and triglyceride (TG) (β=-0.138, p=<0.001). Multivariate regression analysis showed that age (p=0.001), baseline artery diameter (p=0.049), DBP (p=0.048), glucose (p=0.028) and TG (p=0.005) were independent predictors of FMD.

**Conclusion:** Normative reference values for FMD were constructed for children and adolescents with predictive factors identified for this population.

### Impact of COVID-19 on the Sleep-Wake Patterns of a Prospective Cohort of Preschool Children

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**Objective:** To explore the changes in sleep-wake patterns, physical activity, and screen time in pre-schoolers during the COVID-19 pandemic.

**Methods:** A follow-up study was carried out on pre-schoolers who participated in a sleep education project before the COVID-19 pandemic. These children were invited to join the current study, where data including their demographics, their own and parental sleep-wake patterns, physical activities, and screen time were collected through an online questionnaire from August to September 2020. A comparison was made on the collected data from the same cohort of children before and during the pandemic.

**Results:** 497 (13%) children from the baseline cohort of 3720 children were included in the current study. They showed a delay in their bedtime and wake-time on both weekdays and weekends with a 15 to 30 minutes' increase in nocturnal sleep duration. However, with a reduction in nap time, average daily sleep duration was shortened by 16.3±64.3 minutes (p<0.001) and 27.5±72.9 minutes (p<0.001) during weekdays and weekends, respectively. Screen time was increased while outdoor duration was decreased. Parental' sleep/wake times were also delayed with an increase in sleep duration. Children's sleep habits were associated with screen time and their mothers' sleep/wake patterns.

**Conclusion:** Despite school suspension during the COVID-19 pandemic, pre-schoolers were not sleeping longer. Screen time and parental' sleep/wake patterns were the major factors driving the preschoolers' sleep habits. Health education is required to control screen time in children and to promote good sleep hygiene among all family members.

### Epidemiology, Risk Factors and Outcome of Bacterial, Fungal, and Viral Infections in Paediatric Allogeneic and Autologous Haematopoietic Stem Cell Transplantation Recipients

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**Introduction:** With recent changes in haematopoietic stem cell transplantation (HSCT) practices, such as the increasing use of alternative donors and *ex vivo* T-cell depletion, how various risk factors interplay and affect the timeline of infections have not been well elucidated.

**Method:** We retrospectively reviewed the first 100 consecutive HSCTs in the new Hong Kong Children's Hospital.

**Findings:** The vast majority of the allogeneic

transplant recipients (69/74, 93.2%) had infections after HSCT, amongst which bacterial, viral and fungal infection rate was 35.1%, 90.5% and 9.5% respectively. In contrast, only 30.8% (8/26) of autologous transplant recipients had infections (rate of bacterial, viral and fungal infection was 19.2%, 15.4% and 3.8%). HHV-6 and BKV typically occurred early after HSCT, ADV and VZV thereafter, and CMV and EBV throughout the entire 2.5-year observation period. Ex vivo T-cell depletion was a general risk factor for viral infection with HHV-6 (HR=3.03), BKV (HR=3.36), CMV (HR=4.45) and EBV (HR=7.15); all p<0.02. Cancer in second- compared with first-complete remission was a risk factor for bacterial infection (OR=6.0, 95%CI=1.12-32.2, p=0.037). Patients with gut graft-versus-host disease were at risk for fungal infections (OR=12.3, 95%CI=1.33-114.4, p=0.027). The infection-related mortality (IRM) rate was 10%, which occurred only in allogeneic HSCT patients with haematological malignancies receiving cord blood (n=4) or haploidentical HSCT (n=6). History of bacterial infection after HSCT was associated with an increased risk of IRM (HR=4.13, 95% CI=1.05-16.3, p=0.042).

**Conclusion:** Our findings in paediatric patients after contemporary HSCT support both time-dependent and risk-adapted measures against infective complications to improve transplant outcome.

### Medication Prescriptions in Children and Adolescents with COVID-19 Infection: A Propensity Score-Matched Study

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**Background and Purpose:** Long COVID is a term describing the lingering symptoms the patients experience beyond the initial phase of a COVID-19 infection. The impact of the pandemic on mental health in the paediatric population has also been well reported. Medication prescription data provide an objective perspective to evaluate the impact of COVID-19. Therefore, we aimed to investigate the medication prescriptions in children and adolescents with COVID-19 infection and compare them with COVID-negative controls.

**Methods:** A retrospective cohort study was performed using the Clinical Data Analysis and Reporting System. Cases were all patients aged below 18 years with a positive PCR test for SARS-CoV-2 and/or a diagnosis code of COVID-19. Controls were patients who tested negative for COVID-19 and were hospitalised during the same period. The prescription data were retrieved up to 12 months after the initial hospitalisation. Propensity score (PS), the conditional probability of having COVID-19 infection given patients' clinical characteristics, was estimated by a generalised boosted model to balance the differences in the clinical characteristics between cases and controls. The prescription data were compared. Statistical analyses were performed using R software V4.1.3.

**Findings:** 1,035 children and adolescents with COVID-19 infection and 3,272 controls were identified. After PS matching, 852 cases and the same number of controls were included. The prescriptions of bronchodilators, inhaled corticosteroids, cromoglycate, leukotriene, phosphodiesterase type-4 inhibitor, and antihistamines were higher in the controls than in the cases. Hypnotic and anxiolytic prescriptions increased in both COVID-19 patients and control. A more prominent increase in prescriptions for psychosis and antidepressants was observed in non-COVID controls. COVID-19 cases did not obtain more prescriptions for analgesics when compared to controls.

**Conclusion:** This study showed that COVID-19 infection had a limited impact on medication prescriptions among children when compared to COVID-negative controls. An increase in psychotropic medication use was observed in both COVID cases and controls, reflecting the impact of the pandemic on the mental well-being of children and adolescents.

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### Tunneled Femoral Central Venous Catheters as an Alternative Venous Access in Children with Mediastinal Tumours Complicated By Superior Vena Cava Obstruction

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**Background:** In children with malignant mediastinal tumours with superior vena cava obstruction (SVCO), jugular access cannot be used for central venous catheter (CVC) insertion. Tunneled femoral CVC is an alternative in these cases for administration of chemotherapy and blood draw.

**Purpose:** To determine if tunneled femoral CVC can be used as a durable and safe venous access in children with SVCO.

**Material and Methods:** A retrospective analysis of all paediatric oncology patients with tunneled CVCs from June 2020 to June 2022 was performed. Primary outcomes were treatment completion rate and CVC-related complication incidence rate (IR) per 1000 catheter-days for tunneled femoral CVCs. The incidence rate ratio (IRR) was calculated by comparing complication IRs between tunneled jugular and femoral CVCs using Poisson regression model.

**Results:** 159 tunneled CVCs (femoral – 20; jugular – 139) were inserted in 146 patients. 55% of tunneled femoral CVCs were event-free until completion of chemotherapy and 20% were still in-situ at time of analysis. Overall complication IR is 2.3 for tunneled femoral CVC, with exit site infection being the most common (IR 1.1), followed by dislodgement (IR 0.4), tip malposition in lumbar vein (IR 0.4) and inadvertent femoral artery injury (IR 0.4). No catheter-related bloodstream infection or thrombosis was identified. Tunneled femoral CVCs were not associated with higher risk of complications compared to tunneled jugular CVC (IRR 1.07, 95%CI 0.41-2.8).

**Conclusion:** Tunneled femoral CVC is a safe alternative venous access for children with mediastinal tumour complicated by SVCO with comparable complication rate to tunneled jugular CVC.

### Congenital Broncho-esophageal Fistula: An Evasive Vacterl Variant?

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**Introduction:** The common foregut anomaly relating to VACTERL-association is tracheoesophageal-fistula (TEF), while congenital broncho-esophageal fistula (BEF) is rare. Presenting symptoms of H-type TEF and BEF are sometimes indolent and there is no single investigation with promising sensitivity, causing delayed diagnoses and protracted respiratory morbidities. This report presents a case of H-type BEF with predicaments in diagnostic and operative considerations.

**Case Report:** A boy first presented at 6-month-of-age with recurrent fevers and respiratory symptoms with history of mild choking. Workups found positive VACTERL features including cervical plus thirteenth-rib with hemivertebrae, hypoplastic right-pulmonary-vessels, horseshoe-kidneys, but tracheobronchoscopy-esophagoscopy failed to identify any TEF. After recurrent pneumonia, radiographs and computed-tomography revealed gross hypoplasia and consolidations of his right-upper-lobe, with mediastinal lymphadenopathy. Progress computed-tomography at 16-month-of-age eventually reported a fistula between distal esophagus and bronchus to right-upper-lobe with absent middle-lobe. Definitive surgery was proceeded after optimisation with exclusive nasogastric tube-feeding. Besides fistula isolation, surgical challenges included isolation/resection of mediastinal mass plus all adherent diseased lobar segments ideally without risking pneumonectomy.

**Findings:** At esophagoscopy, fistula was identified at distal esophagus and stained endoscopically. Thoracoscopy revealed absence of right-upper and middle-lobe. Fistula dissection was uneventful. Diseased apical segment of the solitary right-lower-lobe in connection was resected completely, as was a large reactive mediastinal lymph-node, while preserving inferior segment. The boy recovered and thrived well after anatomical cure to his symptoms.

**Conclusions:** BEF and associated airway malformation is an evasive and extremely rare variant of foregut anomaly. Timely diagnosis, which can be challenging, is paramount for optimal surgery and outcome.

## The Application of Extracorporeal Blood Purification (EBP) Techniques for Hyperbilirubinaemia in Children

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**Introduction:** Hyperbilirubinaemia and elevated levels of bile acids are associated with increased risk of morbidity and mortality in critical care setting. Various extracorporeal blood purification (EBP) techniques have been used for bilirubin and bile acids removal among critically ill patients but data from paediatric populations remain scarce. We here reported our experience of applying different EBP modalities for children with hyperbilirubinaemia.

**Methods:** The medical records of children requiring either single-pass albumin dialysis (SPAD) or haemoadsorption (HA) for hyperbilirubinaemia in the Hong Kong Children's Hospital between 3/2019 to 7/2022 were reviewed. The clinical features, outcomes, and technical details of the EBP procedure of each patient would be recorded.

**Results:** Among the 14 episodes of EBP from 6 patients with a median age (interquartile range) of 9.3 (5.5) years old, 57.1% of them received HA, 33.3% received SPAD and 7.1% received combined SPAD and HA. All HA episodes employed the Cytosorb® column. The median pre-EBP total bilirubin level was 405 (204) μmol/L. For SPAD, the median treatment duration was 22.5 (10.5) hours and the median albumin dialysate rate was 35.5 (4.5) ml/kg/hour. For HA, the saturation duration per episode was significantly shorter than the corresponding total treatment duration (8 vs 24 hours, p=0.012), and the median total dose and effective doses were 9.8 (6.8) L/kg and 300.0 (163.4) ml/kg/hour respectively The overall bilirubin removal efficacy was 5.3 (16.2)% for SPAD and 44.6 (14.5)% for HA. A higher HA effective dose (p=0.004) and a higher pre-HA bilirubin level (p=0.003) were significantly associated with better removal efficacy. No major EBP-specific complication was encountered. The liver enzymes showed improvement in all children. No patients required liver transplantation and no EBP-related mortality was recorded.

**Conclusion:** Both SPAD or HA were safe and effective modalities for bilirubin removal among children. Future studies should investigate the impact of bilirubin removal on clinical outcomes and explore the factors responsible for better removal efficacy.

## Age of Asthma Onset and the Risk Cardiovascular Disease: A Two-sample Mendelian Randomisation Study

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**Background:** Asthma as a collective disease is observationally associated with cardiovascular diseases (CVDs) in adulthood. Yet causality has not been confirmed due to the effects of confounders, reverse causality, and the impracticality of carrying out randomised controlled trials (RCTs). Mendelian randomisation (MR) is a statistical method that mimics RCTs through using genetic variants to proxy the effects of complex traits on health outcomes, generating causal estimates to guide clinical interventions.

**Purpose:** This study aims to investigate the causal impacts of childhood- and adulthood-onset asthma (COA and AOA) on four adulthood-onset CVDs (coronary artery disease (CAD), heart failure (HF), atrial fibrillation (AF), and any stroke).

**Methods:** Two-sample MR was performed using publicly available summary statistics. A total of 123 and 56 independent genetic variants were used to predict the risk of COA (cases: 13,962; controls: 300,671) and AOA (cases: 26,582; controls: 300,671) respectively. Summary statistics for AF were obtained from a genome-wide association meta-analysis study, while data from the CARDIoGRAM, HERMES and MEGASTROKE consortium were retrieved for CAD, HF and stroke respectively. Odds ratios (ORs) for risk of CVDs in asthmatic cases were calculated using the inverse variance weighted method. Sensitivity analyses (MR-Egger, MR-PRESSO and weighted median) were conducted to evaluate pleiotropy.

**Findings:** Genetically predicted COA was causally associated with AF after accounting for horizontal pleiotropy under the MR-Egger estimate (OR: 1.06, 95% confidence intervals: 1.02 to 1.10). Contrastingly, genetically predicted AOA was not causal for any CVDs across all analyses.

**Conclusions:** These findings support the early-life origins of adult diseases hypothesis and should be triangulated with observational evidence to support the unique management of COA to reduce potential comorbidity with CVDs.

## Multiple Autoimmune Syndrome (MAS): A Rare Combination

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**Introduction:** Around 25 percent of patients with autoimmune diseases have a tendency to develop another autoimmune disorder. When more than one autoimmune disease coexist, this is defined as polyautoimmunity. When three or more autoimmune diseases coexist, this is known as multiple autoimmune syndrome (MAS).

**Purpose:** This report summarises the clinical presentations, laboratory values and treatments of a patient with MAS.

**Findings:** A 12-year-old Chinese adolescent boy presented with polydipsia, polyuria, nocturnal enuresis and weight loss. He was diagnosed with type 1 diabetes and found to have Hashimoto thyroiditis incidentally. One year later, he presented with a prolonged remittent and intermittent fever. He had a painless swelling at his right neck which progressively increased in size. He had no constitutional symptoms. He had no family history of autoimmune diseases. The lymph node biopsy was suggested of necrotising histiocytic lymphadenitis. His blood test showed elevation of predominantly aspartate transaminase (AST) and alanine aminotransferase (ALT). In addition, he had leukopenia, presence of antinuclear antibodies (ANA), anti-double-stranded DNA (dsDNA), anti-cardiolipin, lupus anticoagulant, low complement 4 and positive direct Coomb's test. He was diagnosed with systemic lupus erythematosus (SLE) which was manifested by necrotising histiocytic lymphadenitis and lupus hepatitis. He had three autoimmune diseases, namely type 1 diabetes mellitus, Hashimoto's thyroiditis and SLE, which were a rare combination among MAS.

**Conclusion:** We highlight the importance for physicians to be aware of the need of continued surveillance for the development of new autoimmune diseases in patients who have multiple autoimmune diseases.

## Hesitancy, Reactogenicity and Immunogenicity of BNT162b2 and Coronavac in Paediatric Patients with Neuromuscular Diseases

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**Background:** Vaccination can reduce severe COVID-19. However, some patients with neuromuscular diseases (NMDs) have yet to receive COVID-19 vaccines due to concerns over limited safety and immunogenicity information for NMDs.

**Purpose:** To study precisely the factors for hesitancy, reactogenicity and immunogenicity of BNT162b2 and CoronaVac in paediatric NMDs.

**Methods:** NMDs aged 8-18 years and 2-21 years were invited to complete two longitudinal surveys and join our vaccination research program, respectively, in early 2022. Electronic diary was used to record adverse reactions 7 days after vaccination. To evaluate the antibody response, Blood samples were drawn before each dose and after 28 days to measure levels of SARS-CoV-2 S-RBD IgG and surrogate virus neutralisation test (sVNT), which were compared with healthy children and adolescents.

**Findings:** Forty-one NMDs completed the surveys, while 22 NMDs joined our reactogenicity and immunogenicity vaccination study. More respondents indicated they had or intended to receive the vaccines in April compared to January 2022 (97.6% vs 73.3%, p=0.002). Two or more family members vaccinated against COVID-19 was significantly associated with vaccination (odds ratio: 11.7, 95% CI: 1.81-75.1, p=0.010). Reasons for vaccine hesitancy included a higher chance of encountering adverse effects and safety (n=9/41, or 22.0%). Most adverse reactions were mild, and no severe adverse event was reported. Geometric mean ratios of S-RBD IgGs and sVNT for NMDs and healthy population were above the levels of detection.

**Conclusion:** Overall, the safety and immunogenicity profiles of BNT162b2 and CoronaVac for paediatric NMDs were comparable to healthy children.

### Social Determinants of Paediatric Food Allergy in Hong Kong: A Population-based Cross-sectional Study Through the Lens of Health Equity

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**Background:** Since the COVID-19 pandemic, there has been increased awareness of health inequities caused by disparate disease burdens. The skyrocketing rates of food allergy (FA) worldwide has underscored the importance of understanding the social determinants of FA.

**Purpose:** To perform the first study examining the social determinants and health equity of paediatric FA in the Asia Pacific region.

**Methods:** Questionnaire responses were collected from 4432 children aged 2-7 studying in 32 randomly sampled kindergartens from 2020-2021. Self-reported data collected include demographics, past health, dietary habits, environmental factors, and adverse food reactions (AFR), which is used as the surrogate marker of FA.

**Findings:** Using the median district income as a proxy for household income, income does not have a significant effect on the risk of AFR (p=0.987). There is no significant difference in the odds of AFR for different ethnicities of the child (p=0.797) and whether the child is born in Hong Kong (p=0.573). However, the father being born locally is associated with higher odds of AFR for the child (OR=1.47, 95%CI [1.15, 1.90], p=0.02) and increased burden of AFR on daily life (p=0.002). These effects are not seen for mothers (p>0.05). There is no correlation between the father's and mother's educational attainment and their child's risk of AFR (p=0.055 & p=0.780). Children with AFR have significantly (p=0.000) more siblings (mean=0.67, SD=0.715) than those without (mean=0.82, SD=0.778).

**Conclusions:** Social determinants do not significantly contribute to FA and with minimal health inequity in Hong Kong, in contrast to results reported in North America.

### Influence of the PDA Exposure Duration on the White Matter Maturation in Preterm Infants

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**Objective:** To investigate the effect of exposure duration of patent ductus arteriosus (PDA) on early white matter development in preterm infants.

**Methods:** This study was a retrospective analysis. The subjects were very low birth weight preterm infants with patent ductus arteriosus of preterm infants and complete brain MRI data admitted to NICU of our hospital from January 2014 to January 2020. The comprehensive score of white matter morphology and FA values of key white matter regions were compared between the PDA-closed group and the PDA-open group.

**Results:** A total of 67 children were enrolled in this study, with a gestational age of 28.8±2.3 weeks and an average birth weight of 1176±289 g. Forty-five children were included in the PDA-closed group within 28 days, and 23 children were included in the PDA-open group at 28 days. There were no significant differences in gestational age, birth weight and MRI scan time between the two groups (P>0.05). The scores of white matter comprehensive score of the two groups were 3.37±2.18 and 4.26±1.48, respectively, and the difference was not statistically significant (P>0.05). The FA values of the left posterior limb of the internal capsule were 0.524±0.119 and 0.438±0.128, respectively, while the FA values of the right posterior limb of the internal capsule were 0.525±0.115 and 0.439±0.100, and there was no significant difference between the two groups (P>0.05).

**Conclusion:** The exposure duration of PDA in preterm infants does not affect the developmental maturity of white matter.

### Identification of Common Genetic Variants in Lower Vitamin D Supplementation Response in Hong Kong Hypovitaminosis D Infants

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**Background:** Hypovitaminosis D is associated with long-term health issues and disease development in infants and oral supplementation is an effective approach for minimising the risk. Previous findings in vitamin D genetics identified genes function in the vitamin D metabolic pathway is a major determinant of the serum level. Infants with an identified non-risk haplotype demonstrated a greater vitamin D supplementation response by prolonging 25(OH)D half-life in plasma, which indicates the importance of prior genotyping of the selected loci if vitamin D supplementation is recommended.

**Objective:** To illustrate the dose effects of the selected variants among vitamin D deficient infants after 400IU/d oral supplementations for a month, providing the pharmacogenetic information specifically for vitamin D-deficient infants and patients.

**Methods:** A vitamin D deficient cohort composed of 58 infants, aged 2 to 24 months, were recruited randomly from a voluntary screening program in the community and referred to clinical follow-up with detected vitamin D deficiency(<25 nmol/L). Genotyping of the selected variants was performed on the extracted leukocytic DNA by allelic discrimination. Serum 25(OH)D was measured in the collected samples during the program recruitment and after clinical visits by LC/MS-MS method. The accuracy passed the proficiency testing of the Vitamin D External Quality Assessment Scheme (DEQAS) by Endocrine Laboratory, UK. The lower efficacy alleles of the selected SNPs were identified by a significantly lower average post-supplemented 25(OH)D concentration in the corresponding alleles carriers

**Results:** Seven selected common variants function in the core vitamin D metabolism were genotyped. All the hypovitaminosis D infants showed elevated serum 25(OH)D levels after supplementation. The functional variants rs7041 and rs4588 demonstrated significant allelic differences in the post-supplemented serum 25(OH)D levels, suggesting that VDBP is a major determinant in the variations of the supplementation response in infants with GC2 was identified as the lowest efficacy VDBP isoform. Further identification of the lower dose response alleles was performed by combined haplotype of GC2 isoforms with the selected polymorphisms' alleles and results revealed significantly lower post-supplemented serum 25(OH)D concentration in the carriers of GC2-VDR FokI T, GC2-CYP2R1 G, GC2-CYP27B1 G, GC2-DHCR7 A, GC2-VDR T, when compared to its GC1S-corresponding alleles counterparts, suggesting an efficacy variability not only exerted by VDBP, but also involved genes in the vitamin D metabolic pathway in infants.

**Conclusion:** Genetic factor was demonstrated as a determinant of supplementation response in Hong Kong infants. VDBP, and its genetic interaction with other elements in the metabolism, were found in regulating the infant supplementation response. This preliminary study provided infant-specific vitamin D genetics data in Hong Kong and laid down a basis for genomic medicine in this field.

### The Role of JAK/STAT Signalling in Neuroblastoma Cells Survival and the Promoting Effect of Mesenchymal Stromal Cells

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Neuroblastoma (NB) is the most common extra-cranial solid tumour of children and frequently metastasizes to bone and bone marrow. A close interaction between metastatic tumour cells and the bone marrow microenvironment, especially the mesenchymal stromal cells (MSCs), has been proposed as a key step in the growth and metastasis in several tumour types. However, little is known regarding the role of bone marrow MSCs in neuroblastoma. Here, we investigate that JAK/STAT signalling pathway is important for NB cells proliferation and migration, as well as the promoting effect of MSCs. Small-molecular inhibition of STAT3 suppressed NB cells growth and blocked the effect of MSCs. However, genetic inhibition of STAT3 in NB cells enhanced the tumor proliferation and may be due to the crosstalk with STAT1. Subsequently, we found that MSCs derived factors activated both STAT1 and STAT3 in NB cells, and single genetic inhibition of STAT1 or STAT3 didn't block the pro-tumourigenic activity of MSCs. Whereas, blocking JAK/STAT pathway using JAK inhibitor or knockdown of both

STAT1 and STAT3 dramatically attenuated the NB cells proliferation and the promoting effect of MSCs. Similarly, MSCs also enhanced NB cells migration and could be suppressed by the treatment of JAK inhibitor. Therefore, interference with JAK/STAT signalling might offer a therapeutic strategy for neuroblastoma patients.

### Ex-vivo TCRαβ/CD45RA Depleted Haematopoietic Stem Cell Transplantation for Chronic Granulomatous Disease – First Cases in Hong Kong

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**Background:** Chronic granulomatous disease (CGD) is a rare inherited primary immunodeficiency caused by CYBB, CYBA, NCF1, NCF2 and NCF4 mutations. Patients with CGD are at increased risk of life threatening bacterial and fungal infections, and inflammatory disease. Allogenic hematopoietic stem cell transplantation (HSCT) is a curative treatment for CGD. Haploidentical HSCT could be an option in some CGD patients for whom HLA-matched donors are not available.

**Objective:** To describe the clinical course of two CGD patients undergoing haploid HSCT

**Methods:** Clinical, laboratory and immunological data were collected from medical records. Conditioning regimen consisted of Campath (0.2 mg/kg/day from D-13 to D-10), Busulfan (4.8 mg/kg/day from D-7 to D-5) or Treosulfan (12 g/m$^2$/day from D-7 to D-5), Fludarabine (40 mg/m$^2$/kg/day from D-7 to D-4) and Thiotepa (5 mg/kg/dose Q12H D-4). Peripheral blood stem cells were collected by leukapheresis from the parents after G-CSF mobilisation. Harvest product was processed by TCRαβ and CD45RA depletion by magnetic beads using the ClinicMACS device. The processed stem cells were infused on D0. Dihydrorhodamine (DHR) assay was used to evaluate the function of neutrophils on D+30.

**Results:** P1 is a 4-month-old girl with NCF1 mutation, who presented with pulmonary aspergillosis at 2 months. P2 is a 26-month-old boy with exon 1-3 deletion of *CYBB* gene and McLeod neuroacanthocytosis syndrome (MLS), who experienced recurrent lymphadenitis and perianal abscess. Both underwent haploidentical HSCT in September 2022 and received conditioning regimen as described above. The number of CD34+ cells reinfused were 10×10$^6$/kg and 12×10$^6$/kg respectively. Neutrophil engraftment occurred on D+12 and D+11 respectively, platelet engraftment on D+7 and D+9 respectively. For P1, whole blood chimerism was 100% on day 14 and remained at 100% on D+60. For P2, whole blood chimerism was 100% on D+14 and down to 98% at D+30, so Donor Lymphocyte Infusion was given. DHR on D+30 showed normal results in both patients. P1 showed resolution of pulmonary aspergillosis and P2 had recovery of perianal abscess and lymphadenitis post-HSCT. Both patients did not have graft-versus-host disease (GVHD), viral reactivations or organs complications. P1 was discharged on D+33 and P2 was discharged on D+28.

**Conclusions:** Haploidentical HSCT with ex-vivo TCRαβ/CD45RA depletion is an effective and safe treatment option for clearance of pre-existing infections in CGD. Neutrophil function recovers rapidly upon engraftment, and the incidence of GVHD and viral infections is low in the early post-HSCT period.

### Large Cell/Anaplastic Medulloblastoma with Heterozygous Variants in *MSH6* Gene: A Case Report

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**Background:** Constitutional mismatch repair deficiency (CMMRD) is caused by bi-allelic germline mutations in mismatch repair genes: MSH6, MSH2, PMS2 or MLH1. This entity is a childhood cancer predisposing syndrome with autosomal recessive mode of inheritance. Many patients with CMMRD syndrome additionally develop signposting noncancerous features, most frequently cutaneous pigmentation alterations.

**Case presentation:** We report on a 6-year-old boy with a large cell/anaplastic medulloblastoma and a conspicuous skin pigmentation that included multiple café-au-lait spots, freckling and Mongolian macules. He has no obvious family health history of Lynch syndrome-associated cancers. The diagnosis of CMMRD syndrome was based on next generation sequencing analysis showing heterozygosity for MSH6 variant c.2150_2153del (p.V717fs*18) and MSH6 variant c.3261del

(p.F1088Sfs*2).

**Conclusions:** CMMRD should be suspected in patients who present with childhood cancer and skin features resembling neurofibromatosis type I. DNA-sequencing analysis is of value in identifying people who may have CMMRD syndrome.

### Empowerment Program of Developmental Positioning in Preterm Babies for NICU Nurses

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**Background:** Preterm infants are the majority in Neonatal Intensive Care Unit (NICU), they face challenges when growing up in the extrauterine environment. As there is a lack of a definite official guideline on preterm infant positioning in our department, and positioning is a rather subjective measure, some nurses with less experience may feel confused about learning different practices of positioning a preterm infant. According to Santos et al. (2017), following a standardised procedure while positioning infants brought a positive effect on physiological and behavioural factors that could influence preterm infants' development. Thus, it is important to use an objective tool for guiding fellow nursing staff in positioning a preterm infant.

**Aims:** The project aimed to reinforce the importance of developmental care to NICU staffs, and to improve their nursing skill in positioning preterm babies.

**Objectives:** (1) Explore the existing evidence for ideal positioning in preterm babies; (2) Educate frontline nurses whose year of experience <5 years and without NICU Post-registration Certification by face-to-face teaching and hands-on practice sessions; (3) Provide standardised and clear instructions on routine repositioning of preterm babies; (4) Introduce the use of Infant Positioning Assessment Tool (IPAT) in assessing competence in positioning

**Methods:** A prospective observational study was conducted to investigate the performance of handling developmental care on positioning preterm babies for NICU staffs after education program implementation. Thirty NICU nurses at the Department of Paediatrics & Adolescent Medicine of Queen Mary Hospital (PAM, QMH) were recruited by convenience sampling to perform positioning for eligible preterm babies. Their competency was scored under IPAT by assessors. The reliability and effectiveness of the IPAT was guaranteed (Coughlin, Lohman & Gibbins, 2010). Cronbach's alpha 0.972 was calculated and indicated that the IPAT had internal consistency (Spilker et al., 2016). A written pre- and post-test was used to assess their baseline knowledge and confidence on infant positioning. An educational program was delivered after pre-assessment, on 1:1 or 1:2 basis, with educational talk, PowerPoint slides training material and poster. Demographics data was collected with primary outcome as IPAT score. Secondary outcome included pre- and post-assessment scores, staff confidence level and program evaluation score among nurses.

**Findings:** (1) The average IPAT scores before and after the program were 8.90 and 11.20 respectively. (2) The average total pre-test score was 20.53 / 27.00 whereas the average total post-test score was 25.06 / 27.00. (3) The mean scores confidence level ranged from 3.50 to 3.90 before the program and was around 4.50 (95%). Significant improvements were observed in baby positioning, staff knowledge in positioning strategies and confidence level.

**Conclusions:** This empowerment program successfully improved staff skills, knowledge, and confidence on developmental positioning of preterm babies. However, the target population of this program only covered nurses without specialty training, while competency and knowledge in experienced staff were not assessed. Further study to include all the NICU nurses into the educational program might be needed. To maintain sustainability of the program, adoption of IPAT could be considered as an assessment tool into daily practice to ensure the proper positioning to be carried out in preterm infants.

## A Two-year Review of Functional Outcomes at PICU Discharge in Haemato-oncology Children at a Tertiary Oncology Centre in Hong Kong

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**Background:** Advancements in cancer treatment have resulted in longer survival in haemato-oncology patients, often comes at the expense of therapy-associated new morbidities. Functional outcomes of these survivors can significantly impact their quality of life. The prevalence of morbidity in general PICU survivors at discharge is 4-8%. The aim of this study is to evaluate functional outcomes of haemato-oncology patients at PICU discharge, and to identify risk factors associated with these outcomes.

**Methods:** This is a retrospective observational study of all children with a haemato-oncology diagnosis or undergoing haematopoietic stem cell transplantation who have been admitted to the Hong Kong Children's Hospital PICU over a 2-year period. Functional status upon admission and discharge were evaluated by validated instruments and compared. Univariable and multi-variable analyses were employed to identify risk factors associated with the development of new morbidities.

**Results:** Out of 288 PICU admissions, there were 277 live discharges, of which 52 (18.8%) developed new morbidities. Emergency admission, severity of illness at admission, organ dysfunction and support were all associated with new morbidities with univariable analysis. After adjusting for associated factors, higher Paediatric Logistic Organ Dysfunction 2 score at admission was significantly associated with development of new morbidities ($p<0.001$).

**Conclusions:** Critically-ill children with haemato-oncological diseases had a higher rate of developing new morbidities compared with the general PICU population. This was significantly associated with severity of illness at admission. Further work is warranted to understand the lasting effects of these new morbidities and interventions that may mitigate them.