Annual Academic Sessions 2022

Colombo South Clinical Society

The Book of Proceedings
and
Abstracts

25th March 2022

Dr W. G. Gunawardene Auditorium, CSTH

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MESSAGE FROM THE PATRON

Dr Sagari Kiriwandeniya
Director
Colombo South Teaching Hospital
Patron
Colombo South Clinical Society

It is a great pleasure for me to write a message as the Patron of the society for the abstract book of the Annual Academic Sessions 2022 of the Colombo South Clinical Society. I am indeed very happy to see the progression of the event since its inception in 2015 to the current status. It is indeed a great achievement to be able to have an event of this caliber despite the enormous barriers created by the pandemic.

This annual academic session will give an opportunity for the researchers to platform academic work. I am glad to see the good quality academic presentations of our doctors at this forum. On behalf of the Colombo South Teaching Hospital, I would like to take this opportunity to express my gratitude to Professor Mohan de Silva, Dr Ravindra Randeniya and all the oral and poster presenters for their valuable contribution to make this event a success. The first academic event of the Colombo South Teaching Hospital for this year, the joint clinical session with Sri Lanka Medical Association was held successfully on 25th of February. This has been a good start for the Colombo South Clinical Society.

I take this opportunity to congratulate and express my sincere gratitude to the president of the Colombo South Clinical Society, Dr Ruwanthi Perera and her council of spirited members, for all the effort and hard work in making this event a success.

I wish the organisers all the very best, and the participants, an enriching academic session that will aid you to broaden your medical perspectives. Hope to see you all tonight at the Golden Rose Hotel, Borelasgamuwa at the Annual Doctors' Get together and the Dinner Dance.

MESSAGE FROM THE PRESIDENT, COLOMBO SOUTH CLINICAL SOCIETY

Dr Ruwanthi Perea
President
Colombo South Clinical Society

It gives me great pleasure to convey this message at the Annual Academic Sessions of the Colombo South Clinical Society. It is indeed a very happy moment for all of us at Colombo South Teaching Hospital to witness how the academic event which took place as a guest speech during the dinner dance has gradually progressed to the large-scale event held today.

We are honoured by the presence of our chief guest today, Dr Asela Gunawardane, Director General of Health Services, amidst his busy schedule. We are fortunate to have Emeritus Professor Mohan De Silva, former Dean and the Chairman of the University Grants Commission as a guest speaker at this occasion to deliver a lecture on an area of his expertise on chronic pancreatitis. The presence of our second guest speaker, Dr Ravindra Randeniya, much beloved senior artist of our country will undoubtedly touch the hearts of all the participants.

The academic session consists of two free paper sessions and a poster session. This has provided a platform for the postgraduate trainees and medical officers of Colombo South Teaching Hospital to showcase their research and share the knowledge. I take this opportunity to congratulate all oral and poster presenters for their excellent pieces of work. This academic session will be followed by the annual get together and the dinner dance taking place at Golden Rose Hotel, Boreslesgauwa.

Such an undertaking would not be possible without the unstinted support of many. I therefore wish to extend my sincerest gratitude to all those who have contributed towards the success of both the events. The editorial team who put a lot of energy in compiling the abstract book and the newsletter is acknowledged with much gratitude. The generous financial support extended by all sponsors is very much appreciated.

A special thank you goes to all the members of my council who put a lot of effort to make this event a success despite their busy schedules.

MESSAGE FROM THE DIRECTOR GENERAL OF HEALTH SERVICES

Dr Asela Gunawardena
Director General of Health Services

It gives me great pleasure to pen a few words for the abstract book of the Annual Academic Sessions of Colombo South Clinical Society. I am indeed overwhelmed to see the development of the Colombo South Clinical Society from its infancy in 2015 to how it has developed up to now. This annual academic session is proof of the professional status achieved by the clinical society, as it has made remarkable progress over the past few years and is clearly spearheading the development and enhancement of knowledge, attitudes and skills of healthcare professionals in the field of medicine.

What does an annual academic session mean to a professional society? It can be looked upon as an opportunity to improve on the existing scientific knowledge, introduce new skills and technology, enhance the quality-of-service delivery and renew personal interactions with an exchange of ideas on clinical practices. The end result would be that the recipient, i.e., the general public should reap the benefits of the efforts of the membership of the society which have been planned and executed in a very committed manner.

The society has now matured into a well-recognized and reputed professional organization. It is a timely endeavor that research is focused on the development of the medical sciences in patient care services.

I take this opportunity to congratulate and express my sincere gratitude to the President of the Colombo South Clinical Society, Dr Ruwanthi Perera and her council members, for all the effort and hard work in making this event a success.

I wish the organizers all the very best, as well as the researchers who are enriching the academic sessions, who will aid you to broaden your medical perspectives.

ACADEMIC PROGRAMME

08:30	Poster viewing and judging
09:30	Lighting of the oil lamp, National anthem
09:45	Welcome address by President, Colombo South Clinical Society
09: 50	Address by Director, CSTH
09:55	Launch of the Newsletter
10:00	Guest Lecture: "**Chronic pancreatitis- diagnostic and therapeutic challenges**" by Professor Mohan de Silva
10:30	Oral presentations - Session 1
11:10	Entertainment
11:15	Guest Lecture: "**Doctor-patient relationship: A patient's perspective**" by Dr Ravindra Randeniya
11:45	Oral presentations - Session 2
12:25	Award ceremony
12:30	Vote of thanks

Oral Presentations

Poster Presentations

Poster Presentations

GUEST LECTURES

Prof. Mohan de Silva
MS, FRCS Edin, FCSSL
Professor Emeritus- Surgery
Faculty of Medical Sciences
University of Sri Jayewardenepura

Chronic Pancreatitis – Diagnostic and therapeutic challenges

Management of Chronic Pancreatitis is challenging to clinicians. To understand the diagnostic and therapeutic challenges, in this presentation, I hope to highlight few noteworthy features in the aetio-pathogenesis, clinical features, newer diagnostic modalities and treatment strategies, the understanding of which have resulted in achieving good outcomes.

Dr Ravindra Randeniya
PhD
Professional Actor in Cinema,
Theatre and Television

Doctor-patient relationship: A patient's perspective

Dr Ravindra Randeniya will give an insight into a patient's perspective on the much talked about, salient topic in medical practice; Doctor-patient relationship.

ABSTRACTS

Colombo South Clinical Society
AAS 2022

Oral Presentations

Poster Presentations

INNOVATION TO IMPROVE THE PROCESS OF CARDIOVASCULAR DISEASE (CVD) RISK ASSESSMENT RATE

Vidanage UA[1], Elapatha GEST[2], Thirimanna HDGS[2]

[1] *Ministry of Health, Sri Lanka*
[2] *University of Sri Jayewardenepura, Sri Lanka*

Introduction: Non-communicable diseases (NCDs) are estimated to account for 83% of all deaths in Sri Lanka resulting in a high disease burden. National policies have been developed to overcome this situation. To achieve the national target, all the clients attending Healthy Lifestyle Centres (HLCs) should be assessed for CVD risk. Current CVD risk assessment rate in Matara district is not satisfactory. Objective of the study was to assess the process of CVD risk assessment in the NCD prevention programme in Matara district.

Method: A software was developed and a cohort study was conducted in 34 HLCs in the Regional Director of Health Services, Matara for 6 months (15/11/2020 to 15/04/2021). *Qualitative Component*: Depending on the importance in CVD risk assessment process, participants were purposefully selected for pre and post-interventional Key Informant Interviews (KIIs) and Focus Group Discussions (FGDs). A narrative analysis was done for KIIs and FGDs. *Quantitative component*: Desk reviewing of secondary data relevant for the calculation of the percentage of monthly CVD risk assessments pre and post-intervention in all HLCs, were carried out by accessing NCD monthly returns from HLCs.

Results: According to FGDs and KIIs, most of the participants stated, “The software is very useful, and can be used for speedy assessment.” Desk reviewing of data showed that HLC monthly returns and CVD risk assessment rates have increased in 2021 when compared to 2020.

Conclusion: The main concern was unavailability of a user-friendly screening tool. The software can be used island wide.

AN AUDIT ON COMPLETION OF COVID-19 DEATH NOTIFICATION FORMS SENT FROM THE JUDICIAL MEDICAL OFFICER'S OFFICE AT COLOMBO SOUTH TEACHING HOSPITAL

Renushanth T[1], Dassanayake PB[1]

[1]*Judicial Medical Officer's office, Colombo South Teaching Hospital*

Introduction and Objectives: The COVID-19 pandemic has lasted for more than two years, challenging all countries in every aspect. In Sri Lanka, there has been a significant number of deaths due to COVID-19, especially after the spread of the delta variant. Since COVID-19 is new to the entire world, accurate notification of deaths is important to find out the epidemiology of the disease. Objective of the study was to audit the completion of COVID-19 death notification forms sent from the Judicial Medical Officer's (JMO's) office, Colombo South Teaching Hospital.

Method: A total of 813 consecutive notification forms sent from the JMO's office during four months were collected. Altogether, 22 sections, including name of the deceased, age, and cause of death were descriptively analysed. Data was analysed using SPSS software and the results were presented as frequencies and percentages.

Results: Out of 813 notification forms, 9 were excluded based on exclusion criteria. There were 481 (59.82%) completely filled forms. There were 332 (40%) notification forms which did not mention comorbidities and this was the commonest error. In 81 cases, abbreviations were used to write the cause of death. The time between the presumed onset of the condition and death was never recorded in the cause of death section.

Conclusions: Proper completion of the COVID-19 death notification form is necessary to serve its purpose. However, inadequately filled death notification forms remain a challenge. Hence, it is of utmost importance that all relevant medical officers fill the death notification form appropriately. An awareness programme among the medical officers regarding this would be beneficial.

ADEQUACY OF PAIN MANAGEMENT IN THE POST-OPERATIVE PERIOD FOLLOWING LOWER SEGMENT CAESAREAN SECTION: A CLINICAL RE-AUDIT

Senanayake HGS[1]

[1]*Postgraduate Institute of Medicine, University of Colombo*

Introduction and Objectives: Inadequate pain management during the acute post-operative period following Lower Segment Caesarean Section (LSCS) is associated with adverse outcomes interfering with care for the newborn. The adequacy of acute post-operative pain management following LSCS was audited in the Professorial unit of Colombo South Teaching Hospital (CSTH) 6 months previously and the results revealed that mothers were in considerable pain until the analgesics took effect as they were administered only after the onset of pain. Therefore, a protocol was recommended based on the PROSPECT guideline. The objective of this re-audit was to determine whether the recommended protocol was implemented in this unit.

Method: Forty consenting mothers who had undergone LSCS were randomly selected from the Professorial unit, CSTH over one month. An interviewer based questionnaire was used to collect data from the mothers regarding their pain perception based on the Numerical Pain Intensity Scale (1-10) and the Wong Baker Facial Grimace Scale. Medical records maintained in the ward were used to assess the pain management received by these mothers during the post-operative period. Management given in the ward was compared with the protocol recommended according to the PROSPECT guideline following the initial audit.

Results: Most frequently used analgesic (97.5%) for the post-operative pain was diclofenac suppository. None of the mothers received paracetamol suppositories. Only 12.5% were given oral diclofenac for rescue pain. Oral paracetamol was given up to a maximum of 2 doses during this period and the first dose was received by 32.5% while only 23.1% received a second dose.

Conclusions: Results reveal that implementation of the recommended protocol is not successful and needs modifications.

AUDIT ON DOOR TO ECG TIME AT THE EMERGENCY TREATMENT UNIT OF COLOMBO SOUTH TEACHING HOSPITAL

Weerakkody DSI[1], Elvitigala KN[1], Munasinghe MP[1], Goonewardene D[1], Epasinghe DP[1]

[1]*Colombo South Teaching Hospital*

Introduction: Emergency Treatment Unit (ETU) of Colombo South Teaching Hospital (CSTH) had a total of 7299 admissions in 2021, 3079 of them presented with chest pain where 409 were thrombolysed and 26 were transferred to National Hospital for Percutaneous Coronary Intervention. They also held the highest mortality in the ETU. Electrocardiogram (ECG) is the principal investigation in diagnosing acute coronary syndrome in candidates requiring thrombolysis. According to American Heart association, standard door to ECG time is less than 10 minutes. Objectives of the audit were to assess the door to ECG time.

Method: Data was collected from of 822 random patients admitted to ETU from October-December in 2021.

Results: Mean door to ECG time in the study population was 17.2 minutes (SD 13.8). Out of patients admitted with chest pain, 41.18% (217/519) achieved the standard door to ECG time and 8.6% (45/519) patients received ECG only after 30 minutes from admission. The mean time was 15.9 minutes (SD 11.7) and the longest time was 100 minutes among them.

Conclusions: CSTH ETU doesn't achieve the standard door to ECG time in more than 50% of patients. Recommendation is to allocate a specific ECG technician for the ETU or allow the ETU staff to perform ECGs. This study will be followed up by a repeat audit in 6-months.

NEUTROPHIL LYMPHOCYTE RATIO AS A MARKER OF IN-HOSPITAL DETERIORATION IN COVID-19: OBSERVATIONS FROM A RESOURCE CONSTRAINT SETTING

Perera N[1], de Silva A[1], Kumbukage M[2], Rambukwella R[2], Indrakumar J[1]

[1]*Department of Medicine, Faculty of Medical Sciences, University of Sri Jayewardenepura*
[2] *Research unit, Family Health Bureau*

Introduction and objectives: COVID-19 pandemic has posed an enormous challenge to health care services and it is vital to have simple, cheap and widely available tests for identifying patients likely to develop severe disease. This study was conducted to assess the association of neutrophil lymphocyte ratio (NLR) in COVID-19 and to identify the cut-off value that predicts mortality, need of respiratory support and admission to high-dependency or intensive care.

Methods: A retrospective observational study was conducted to collect the NLR on-admission and the outcome of confirmed COVID-19 patients admitted to Colombo South Teaching Hospital during July-August 2021. Receiver operating characteristic curve and logistic regression were used to identify the association of NLR with disease outcome and to identify the cut-off value predicting severe disease.

Results: There were 208 patients with a median age of 56 years (IQR 43-67) and 98 (47.1%) males. The median neutrophil count was 4.07 $x10^3/\mu L$ (IQR 2.97-6.79) and the median lymphocyte count was 1.74 $x10^3/\mu L$ (IQR 1.36-4.75). The calculated NLR ranged from 0.12 - 48.28 with a median value of 2.32 (IQR 1.37-4.76). A NLR value >3.6 predicted development of severe disease requiring respiratory support, transfer to a high-dependency or an intensive care unit and/or succumbing to the illness with a sensitivity of 80% and specificity of 80% (area under the curve 0.8, 95% CI 0.72-0.88, $p<0.0001$). The adjusted odds ratio of NLR >3.6 on predicting severe disease was 11.1, 95% CI 4.5- 27.0, $p<0.0001$.

Conclusions: A NLR>3.6 is a useful variable to be included in risk prediction scores in Sri Lanka.

OVERUTILIZATION OF X-RADIOGRAPHS IN SCREENING CERVICAL, ANKLE AND FOOT INJURIES OF POST-TRAUMATIC PATIENTS AT A TERTIARY CARE CENTRE IN SRI LANKA

Prasanga RPS[1], Vishwajith RMV[1], Jayathilake AB[2], Jayasoma SAMM[1]

[1]*Post Graduate Institute of Medicine*
[2]*Lecturer, Professorial Surgical Unit, Faculty of Medicine, University of Rajarata*

Introduction and objectives: Post-traumatic cervical, ankle and foot injuries are an important clinical entity and it is difficult to diagnose such injuries clinically. Overutilization of X-radiographs (X-ray) is a common problem which happens in post-traumatic setting. This study was designed to analyse the accurate use of cervical, ankle and foot X-ray as a screening tool performed in post-traumatic setting.

Methods: This study was performed in an emergency department of a single centre. A total of 200 each of cervical, ankle and foot X-rays were analysed throughout 3-months according to the National Emergency X-Radiography Utilization Study (NEXUS) Cervical Spine rules and Ottawa Ankle and Foot Rules. X-ray findings and the indication to perform X-rays were evaluated. Proper documentation of clinical indication was also assessed.

Results: Out of X-rays assessed, 103(51.5%) cervical, 112(56%) ankle and 110(55%) foot X-rays had an indication to perform imaging. The remaining 97(48.5%) cervical, 88(44%) ankle and 90(45%) foot X-rays were performed without an indication. Furthermore, 121(60.5%) cervical, 107(53.5%) ankle and 118(59%) foot X-rays had no documentation of the indication.

Conclusions: Overutilization of X-ray is an important problem in post-traumatic setting. The rational implementation of NEXUS Cervical Spine rules and Ottawa Ankle and Foot Rules would possibly bring down the number of unnecessary cervical ankle and foot X-rays in the post-traumatic setting. This prevents unwanted radiation and increased cost. Furthermore, poor documentation is another significant deficiency which needs to be corrected. Use of a pro-forma including above criteria would help to minimize this overutilization.

PRIMARY PERITONEAL SARCOIDOSIS: A RARE AND CHALLENGING DIAGNOSIS

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[2]*Department of Obstetrics and Gynecology, Faculty of Medical Sciences, University of Sri Jayewardenepura*

Introduction: Sarcoidosis is a rare multisystem inflammatory disorder which has a predilection towards lungs and lymph nodes. Diagnosis is challenging and is dependent on demonstrating non-caseating epithelioid granulomas with exclusion of common granulomatous diseases.

Case report: A 57-year-old postmenopausal female while being evaluated for a cystocele was found to have ascites on ultrasound scan abdomen along with elevated CA-125 levels. Contrast enhanced CT did not demonstrate adnexal masses nor enlarged lymph nodes. Although Mantoux test was positive, high-resolution CT-chest did not demonstrate evidence of tuberculosis (TB) nor other lung pathology, thus leaving us at a diagnostic dilemma. An exploratory laparotomy was undertaken where multiple white tubercle-like lesions that were consistent with peritoneal TB and marked adhesions were found. Pathological evaluation revealed multiple epithelioid cell granulomata, leaving the possibilities of TB or sarcoidosis and excluding the diagnosis of peritoneal carcinomatosis. Given that TB is common, the patient was started on anti-TB regimen for peritoneal tuberculosis. However, erythrocyte sedimentation rate and CA-125 continued to rise after weeks of therapy and a diagnosis of peritoneal sarcoidosis was made and the patient responded to corticosteroids.

Discussion and Conclusions: Extrapulmonary involvement in sarcoidosis is considered rare and primary peritoneal sarcoidosis is considered to be extremely rare with only a handful of reported cases. Given that TB is widely prevalent, initial diagnosis of TB-peritonitis or widespread carcinomatosis as suggested by elevated CA-125 and ascites is justifiable, but sarcoidosis is a disease entity that should not be overlooked. In conclusion, peritoneal sarcoidosis should be considered as a possible differential diagnosis when noncaseating epithelioid granulomas are demonstrated.

EVALUATION OF THE ChAdOx1 nCoV-19 (COVISHIELD™) VACCINE (1ST AND 2ND DOSES) INDUCED TOTAL ANTIBODY RESPONSE TO RECEPTOR BINDING DOMAIN OF SPIKE PROTEIN (RBDS), IN A COHORT OF PARTICIPANTS IN COLOMBO

Jinasena TMRR[1], Gunawardane SA[2], Katulanda GW[2], Agampodi SB[3], Dodampahala H[4], Thushyanthi P[1], Athapaththu AMTU[1], Samarasinghe M[1], Jayasinghe IN[1], Dissanayake DJGGN[2], Inthujah T[2], Samarakoon SMPP[2], Balasooriya BMCM[2] ,Sujeeva N[2], Thowfeek ZTM[2], Prashanthan S[2], Ediriweera TW[2], Hewa SP[1]

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Introduction: As a part of the global effort to combat SARS CoV-2 infection, the first immunisation programme using Covishield vaccine was launched in Sri Lanka, in early 2021. To evaluate the success of the initiative, it is essential to assess the immune response of recipients.

Methods: Adult participants (190) were randomly selected at vaccination centres, National Hospital of Sri Lanka and Colombo South Teaching Hospital. Blood samples were collected on day-0,14,21,45 and 90 following the first dose and on day0, day14 and 3months after the 2nd dose. Total antibody levels to RBDS were measured using Advia XP chemiluminescent assay.

Results: Following a single dose, seroconversion was observed in 87.5%, 95.0% and 100% on day 14,21 and 45, respectively. Greater response was noted on day 45 following the 1st dose (median 14.43-Index, IQR 5.5-34.3) and at 2-weeks following the 2nd dose (median 237.14-Index, IQR 194.73-322.64). No significant difference was noted with all age groups up to 70-years and in the presence of comorbidities. Participants with an extreme of BMI (<18.5kg/m^2 or >30kg/m^2) showed significantly low response. Participants who received the 2nd dose 3 months following the 1st dose, had a significantly higher response at post 2-weeks (mean 259-Index) compared to individuals who received the 2nd dose 45 days later (mean 83.42-Index). Considerable amount of antibodies persisted in 95% of the participants who did not receive the 2nd dose within the scheduled time period (median 4.03-Index, IQR 2.22-8.12). Three months following the 2nd dose, a significant reduction of antibody levels (median 67.68-Index, IQR 35.33-107.27) were noted in almost all participants with a p value <0.05.

Conclusion: Two doses of the vaccine showed a high antibody response irrespective of the age, gender, BMI and comorbidities. An interval of 12 weeks between 2 doses showed a higher response compared to a 45 days gap.

HOW CAN MEDICATION ADMINISTRATION ERRORS (MAE) IN ICUS BE REDUCED?

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[1]*Ministry of Health, Sri Lanka*
[2]*Faculty of Medical Sciences, University of Sri Jayewardenepura, Sri Lanka*

Introduction and Objectives: This project was carried out to solve management issues and to evaluate the process of Medication Administration (MA) at Intensive Care Units (ICUs). Objective of the study was to reduce occurrence of MAE in ICUs in Lady Ridgeway Hospital, Colombo.

Methodology: The study was implemented in three phases from 15/07/2020-15/06/2021. The existing process of MA was assessed in phase I. Three qualitative methods were used: Focus Group Discussions (FGDs), Key Informant Interviews (KIIs) and Observation of the MA process using a checklist. Quantitative assessment was done by a self-administered questionnaire given to relevant staff in pre and post intervention phases, FME proactive analysis and desk review of data. Interventions to improve MA procedures were designed and implemented in phase II which included development of a guideline booklet, training programmes for staff, wearing "Please do not disturb tags", working in a well-lit environment. The effectiveness of the interventions was evaluated in phase III (post-intervention).

Results: However, even after the intervention requirement of in-service training on MA for better performance (p value 0.417), availability of adequate guidelines and checklists on MA (p value 0.708), adequate record keeping about medication administration errors (p value 0.007), important to maintain a record of MA errors for each patient (p value 0.320) has not improved significantly after the intervention.

Conclusions and Recommendations: The introduced intervention was successful in improving the MA process. Introducing a guideline booklet has improved MA by giving due attention to MAs, therefore this will lead to reduction of MAEs and healthcare cost.

OVERLAP OF DELLEMAN-OORTHUYS SYNDROME AND GOLDENHAR SYNDROME

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[2]*University Paediatric Unit, Colombo South Teaching hospital*

Introduction: Delleman-Oorthuys syndrome known as oculo-cerebro-cutaneous syndrome (OCCS), occurs sporadically as a triad of eye, cerebral and dermatological malformations.

Case report: A baby boy born at 37-weeks was noted to have following craniofacial anomalies; his left palpebral fissure was fused with anophthalmia while the right cornea was cloudy. His left ear was normal while the right ear had linear skin grooves on the back of the pinna, a skin tag and two pre-auricular sinuses with an absent external auditory meatus. He also had cutis aplasia over the vertex. Rest of the system examination was normal and hearing was normal on both sides. His cardiac assessment, brain imaging and spine was normal while abdominal ultrasound scan showed a hepatic haemangioma.

Discussion: OCCS which has a male preponderance without a familial inheritance was first described by Delleman and Oorthuys and it is believed to occur due to a defect in the fifth or sixth week of embryonal development. Commonest eye anomalies are orbital cysts, microphthalmia or anophthalmia and palpebral, iris colobomas and congenital cataract have been reported. The commonest dermatological lesions are focal areas of skin aplasia or hypoplasia of variable size. Ventricular system anomalies, cerebral and cerebellar cysts and agenesis of the corpus callosum have also been reported. Some OCCS cases show features overlapping with Goldenhar Syndrome (GHS). GHS is characterized by facial asymmetry, peri-auricular skin tags, dermoid cysts, iris and palpebral colobomas, cleft lip and palate, micrognathia, auditory malformations and vertebral anomalies. Our patient had features of OCCS and some features of GHS.

VARIATION IN DOOR TO NEEDLE TIME OF PATIENTS RECEIVING THROMBOLYSIS FOR ST-ELEVATION MYOCARDIAL INFARCTION AT THE EMERGENCY TREATMENT UNIT OF COLOMBO SOUTH TEACHING HOSPITAL

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[1]*Emergency Treatment Unit, Colombo South Teaching Hospital*

Introduction: An ST-elevation myocardial infarction (STEMI) is a transmural myocardial infarction and is a medical emergency. While percutaneous coronary intervention (PCI) is the treatment of choice for STEMI, due to logistical constraints, fibrinolytic therapy is frequently used instead. The mortality benefit of administration of fibrinolytic agents is highest within 4 hours of symptom onset and declines rapidly with time. This audit aims to evaluate the time taken to administer fibrinolytic therapy after admission of a patient with a STEMI to the emergency treatment unit (ETU) at Colombo South Teaching Hospital (CSTH) to further improve patient care. The expected time is less than 30 minutes according to the ACC/ AHA guidelines for STEMI management. Objectives of the audit were to determine the average time taken for administration of thrombolysis and to determine if time of admission affects time taken for administration of thrombolysis.

Method: Data were collected on all patients receiving thrombolytic therapy in the ETU from October to December 2021. Time of admission, time of administration of Tenecteplase and outcome of thrombolysis were documented. Incomplete data were excluded. Results were analysed with SPSS (version 26.0.0.0).

Results: Average time from admission to thrombolysis was 30 mins (SD 26.4 min). There was no statistically significant difference in time to thrombolysis between day and night admissions. ($p = 0.71$).

Conclusions: Thrombolytic therapy is given within 30 minutes on average at the ETU of CSTH with no significant difference in this time between day and night admissions. Further audits should be carried out to determine the factors contributing to this delay.

INFANTILE-ONSET OF MAPLE SYRUP URINE DISEASE (MSUD) WITH DBT MUTATION

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[2] *Department of Paediatrics, Faculty of medical Sciences, University of Sri Jayewardenepura*

Introduction: Maple syrup urine disease (MSUD) is a rare metabolic disease which occurs due to reduced activity of branched chain alfa ketoacid dehydrogenase (BCKD) complex. Mutations in 4 genes (BCKDHA, BCKDHB, DLD, DBT) are associated with autosomal recessive inheritance of MSUD. Here we discuss the initial presentation and subsequent follow up of a child with MSUD with possible DBT gene mutation.

Case history: A 3-week-old baby presented with abnormal crying, poor feeding, poor weight gain and persistent hypertonia. She was born at term by elective caesarean section as the second child of healthy non-consanguineous parents with an uncomplicated antenatal and immediate postnatal period. She did not have a body odour. Her basic blood test results were normal except marginally elevated transaminases. Electroencephalogram showed epileptic encephalopathy and MRI brain showed bilateral symmetrical markedly increased signal intensity in basal ganglia, thalami, internal capsule and cerebellum. Guthrie test was positive for MSUD and branched chain amino acids (BCAA) were elevated in plasma. Urine organic acid analysis showed elevated branched chain keto acids and succinate which was compatible with MSUD. Genetic testing revealed two heterogeneous variants of DBT gene. She was commenced on oral levetiracetam for seizures, high doses of thiamine and BCAA (valine, leucine, isoleucine) free enteral formula.

Discussion: DBT gene mutation associated MSUD has mild clinical manifestations. Early diagnosis and timely intervention with BCAA free formula and thiamine supplementation improves outcome. Our patient improved clinically with no further seizures, adequate growth and achieved developmental milestones. Repeat plasma amino acid analysis after 2-months showed reduced levels of BCAA in plasma.

INFANTILE NEUROAXONAL DYSTROPHY (INAD) IN A CHILD WITH DEVELOPMENTAL REGRESSION

Abewardana LS[1], Nasoor MFM[1], Wijesekara DS[1,2]

[1]*Professorial Paediatric unit, Colombo South Teaching Hospital*
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Introduction: Infantile neuroaxonal dystrophy (INAD) is a neurodegenerative disorder affecting the axons of the nervous system causing a progressive loss of motor skills, vision, and intellectual capacity. They are asymptomatic at birth with symptoms and signs appearing after the first year of life.

Case report: A 10-year-old girl, the only child of non-consanguineous parents, presented at the age of 4 years with a history of recurrent falls. She was born at term without any antenatal or postnatal complications. Parents noted clumsy movements at 3 years of age, followed by gradual regression of motor milestones and gradual deterioration of vision and language over 1 year. By 6 years, her interactions with caregiver were limited to touch. She was totally bedbound with spastic limbs and contractures with minimal involvement of swallowing and respiration. There is no history of recurrent respiratory tract infections and seizures. Her basic blood investigations, metabolic screening and electroencephalogram were normal. MRI Brain showed cerebral atrophy. Genetic Mutation analysis revealed mutation in the PLA2G6 gene compatible with INAD, her parents being carriers. Currently she is well looked after by parents.

Discussion: INAD is an autosomal recessive disorder with a prevalence of 1 in 1,000,000. It is associated with progressive white matter degeneration leading to global developmental delay followed by developmental regression. Death usually occurs due to respiratory complications between the ages of five to ten years. Currently, only palliative treatment is available. However, there is some recent promising evidence with gene therapy.

PRIMARY ENDOCARDIAL FIBROELASTOSIS WITH CONTRACTED LEFT VENTRICLE (LV): A RARE CASE

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Introduction: Endocardial Fibroelastosis (EFE) is seen among 1:5000 live births. Deposition of acellular fibrocartilaginous tissue leads to diffuse thickening of the ventricular endocardium and classifies as dilated or contracted based on left ventricular (LV) morphology. We present an infant with EFE with contracted LV diagnosed at the post mortem examination.

Case presentation: One-month-old boy born at 36-weeks of gestation (birth weight of 2.76 kg) was brought with respiratory distress, poor feeding and poor activity. He was resuscitated as acute heart failure with cardiogenic shock and transferred to a special care baby unit for further management. Septic screen was negative. Chest radiograph demonstrated massive cardiomegaly. Echocardiogram revealed dilated right ventricle (RV) with biventricular dysfunction (Ejection fraction 20%). Baby expired following a pneumothorax. Antenatal history revealed that concerns were raised regarding fetal cardiac function though the heart was structurally normal at the anomaly scan. Fetal echocardiogram was performed and recommended to have postnatal follow up. However, the baby has been discharged after saturation monitoring without a cardiac assessment following birth. Postmortem revealed an enlarged thick walled RV and a small LV which had diffuse glistening, whitish thickened endocardium with shortened chordae tendineae suggestive of EFE. Multiple pulmonary hemorrhages and pulmonary artery atheroma suggested pulmonary hypertension. EFE was histologically confirmed by cardiac tissue examination.

Discussion: Primary EFE with contracted LV in children is rare. Antenatal and postnatal cardiac assessments are important to plan management of these patients. Clinical outcome could be significantly improved by early diagnosis and appropriate interventions.

'DIPLOPIA AND FATIGABILITY'...CAN IT BE A NORM?

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Introduction: Congenital myasthenic syndromes (CMS) are rare inherited disorders of neuromuscular transmission. We herein report a case of CMS who had bilateral droopy eyelids and difficulties with motor activities assumed as a norm for the child since infancy.

Case report: A 12-year-old girl, born to healthy parents presented with proximal muscle weakness, fatigability, droopy eyes and diplopia. She was born by normal vaginal delivery and had to be resuscitated at birth. She required gavage feeding initially and had gross motor delay. Bilateral partial ptosis was noted at 4-months of age and she had significant bilateral upper limb weakness at 4-years of age. The maternal grandmother had partial ptosis since birth. On examination, bilateral symmetrical partial ptosis and almost complete external ophthalmoplegia was noted. Proximal muscle power in both upper and lower limbs were less than distal. She had a waddling gait and Gower's sign was positive. Electromyogram showed a significant decrement pattern supporting the diagnosis of Myasthenia gravis while the muscle biopsy showed type 2 muscle fiber atrophy. Serum and cerebrospinal fluid lactate levels were normal and did not suggest a diagnosis of mitochondrial myopathy. Other investigations were normal. She was commenced on oral Pyridostigmine and the response was remarkable.

Discussion: CMS present commonly in infancy and childhood. Clinical presentation can be varied and treatment challenging according to the affected gene. Acetylcholinesterase (AChE) inhibitors have a favorable effect in most, but may result in worsening symptoms in some. Our patient improved with AChE inhibitors and she is leading a near normal life with some limitations of outdoor physical activities.

CHILD WITH MULTIPLE CEREBRAL CAVERNOMAS PRESENTING WITH STATUS EPILEPTICUS

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Introduction: Cavernomas are vascular malformations. They are uncommon in children. Most cavernomas are asymptomatic while some can present with hemorrhage (62%) and seizures (35%). This case report illustrates a right temporal lobe cavernoma presenting with status epilepticus in a young boy.

Case report: A 10-year-old healthy boy with no past or family history of epilepsy presented with a generalized status epilepticus preceded by initial staring. He had complained of a headache over right temporal region and vomited once the previous day. Examination revealed hypertonia in left upper and lower limbs with brisk reflexes while the remainder of the examination was normal. Urgent magnetic resonance imaging (MRI) scan of the brain with vascular imaging was performed on the same day which showed a large bleed into a cavernoma over the right temporal region and multiple small cavernomas. Following surgical excision of the cavernoma over the right temporal lobe, he made a complete recovery.

Discussion: Cavernomas occur as solitary or multiple lesions. In the familial variety, cavernomas are multiple. The mode of inheritance is autosomal dominant with variable penetrance. T2-weighted MRI scan is said to be very sensitive in detecting them. Cavernomas in children are less common. Presentation of multiple cavernomas as status epilepticus in children is even rarer. The lesion in the boy was easily accessible surgically and completely removed. However, there were a few small cavernomas detected at surgery that were left untouched as the management options are surgery or conservative. It was decided to observe with follow up MRI in a few years for further evaluation.

THREE SISTERS WITH ATAXIA TELANGIECTASIA

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Introduction: Ataxia telangiectasia (A-T) is an autosomal recessive disorder caused by mutations in the ATM gene, resulting in cerebellar atrophy with progressive ataxia, telangiectasia, immune deficiency and increased incidence of lympho-reticular malignancies and brain tumors. Treatment is mainly supportive and there is no definitive treatment. We report a case of a 13-year old girl who manifested features of A-T with recurrent chest infections.

Case report: A 13-year-old girl with a background history of ataxia and ocular telangiectasia presented with left sided lower lobe pneumonia. She is the third child born to non-consanguineous healthy parents. Previous two siblings were diagnosed with A-T and died in adolescence due to recurrent chest infections. She had ataxia and could walk a distance of 3-5m with support, and used a wheelchair for mobility. She was on trihexyphenidyl and clonazepam for the movement disorder. Her immunoglobulin profile revealed low IgA and IgG and mutation analysis confirmed the diagnosis of A-T.

Discussion: A-T is a hereditary neurodegenerative disorder. Diagnosis is usually clinical and genetic studies confirm the diagnosis. There is no specific treatment for the condition yet. Therefore, management is mainly symptomatic and requires a multidisciplinary approach to treat specific symptoms to minimize complications and to improve the well-being of affected children.

CHROMOSOME 9P DELETION IN A CHILD WITH DYSMORPHIC FEATURES AND CONGENITAL HEART DISEASE

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Introduction: 9p deletion syndrome is a rare chromosomal disorder due to missing genetic material on the short arm (p) of 9th chromosome. Severity, signs and symptoms depend on the size and location of the deletion. Developmental delay, particularly speech and language, craniofacial dysmorphism, hypotonia, congenital heart diseases, disproportionately long phalanges, inguinal hernia, omphalocele and middle ear infections are common presentations.

Case presentation: Our patient is a 2-year-old first born male child of non-consanguineous parents. He was born at term to a 37-year-old insulin dependent type 1 diabetic mother. Diabetes control was optimized prior to the pregnancy and there were no other antenatal or postnatal complications. An atrial septal defect was noted at birth, which resolved on subsequent follow up. At the age of 1.5 years, he was diagnosed to have autistic spectrum disorder. His physical examination revealed mid facial hypoplasia, prominent forehead, epicanthic folds, anteverted nostrils, long philtrum and thin upper lip. His basic blood investigations and metabolic screening were normal. His chromosomal analysis revealed deletion of chromosome 9 with one of the parents being a possible balance translocation carrier (parents' karyotyping is pending).

Discussion: Chromosome 9p deletion has a prevalence of 1:50,000 in newborns, of whom two-thirds are girls. Half of the cases occur sporadically and the remainder result from parental translocation. Cytogenetic analysis remains the mainstay of diagnosis. There is only supportive treatment modalities for correcting underline medical and surgical problems. Genetic counselling is important.

ALAGILLE SYNDROME: AN INCIDENTAL DIAGNOSIS

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Introduction: Alagille syndrome, the most common syndrome with intra hepatic bile duct paucity, has an estimated incidence of 1 in 30,000 live births and an equal incidence between males and females. This multi system condition with autosomal dominant inheritance is caused by mutations in one of the two genes, JAG1 which is commoner and the NOTCH2 gene. In this case report, we present the case of a 2-month-old infant, a known patient with congenital cyanotic heart disease, who was clinically diagnosed with Alagille syndrome during his admission for vaccination.

Case report: A two-month-old baby boy, diagnosed with Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral artery-dependent pulmonary circulation was admitted for his routine vaccination. On examination, he had growth faltering and dysmorphic facies with low set ears, broad forehead and a bulbous tip of nose and he was icteric. There was no hepatosplenomegaly. Further inquiry revealed passing of pale stools. On further investigation, he was found to have direct hyperbilirubinemia with elevated transaminases, alkaline phosphatase and γ-glutamyl transferase. Ultrasound scan of the abdomen showed normal gallbladder without a triangular cord sign. Chest and abdominal radiographs showed thoracic butterfly vertebrae. Eye examination was negative for posterior embryotoxins. Diagnosis of Alagille syndrome was made based on the clinical criteria and the patient was transferred for liver biopsy, where the child died before it was performed. Postmortem was not performed due to no consent.

Discussion: Although rare, Alagille syndrome must also be considered in a child presenting with prolonged direct hyperbilirubinemia.

PRIMARY INGUINAL TUBERCULOUS LYMPHADENOPATHY: A COMMON DISEASE IN AN UNCOMMON SITE

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Introduction: Tuberculosis (TB) is a significant public health burden worldwide and in Sri Lanka. In 2016, 4% of total Sri Lankan cases were in children. Extra pulmonary tuberculosis (EPTB) manifests in 8.4 – 13.7% of all TB cases and it is common among children, with the most common type being tuberculous lymphadenitis. In this case report, we present a case of primary tuberculous lymphadenitis presenting with inguinal lymphadenopathy.

Case report: A 14-year-old girl, diagnosed with rheumatic carditis and iron deficiency anemia presented with exertional dyspnea and a gradually enlarging painless lump in the right inguinal region for 1-month duration. She had subjective weight loss and night sweats but no fever, respiratory symptoms or a contact history of TB. She was not treated with antibiotics prior to admission. She was underweight and had the BCG scar. Right sided firm inguinal lump measuring 3 x 5 cm was noted. Multiple bilateral inguinal lymph nodes were palpable without cervical, axillary or epitrochlear lymphadenopathy. The rest of the system examination was unremarkable without hepatosplenomegaly. Chest radiography was normal and HIV antibodies were negative. Lymph node biopsy revealed chronic granulomatous lymphadenitis with negative Ziehl Nielsen staining and mantoux test was strongly positive. Sputum for AFB and Gene X-pert were negative. She was started on anti-tuberculous treatment and demonstrated satisfactory clinical response.

Discussion: Histopathology should be attempted in indecisive cases like in this presentation, to establish the diagnosis of primary tuberculous lymphadenitis. Caseous necrosis of the lymph nodes is an important characteristic in identifying TB which responds promptly to anti-tuberculous drugs.

LARYNGOTRACHEOBRONCHITIS: A NOVEL PRESENTATION OF COVID-19

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Introduction: The novel Corona virus SARS-CoV-2, was declared as a pandemic by WHO on 11th March 2020. Since then, new clinical presentations of COVID-19 are being described daily. Laryngotracheobronchitis (croup), the most common form of acute upper airway obstruction in children, is caused by viruses. In this case report, we present the case of a 14-month-old child with croup who was found to have positive rapid antigen test for COVID-19.

Case Report: A 14-month-old previously healthy, immunized boy presented to the emergency unit with an acute onset abnormal breathing sound for four hours duration. There was no history of fever, cough, foreign body inhalation, choking or recent contact with COVID -19 positive patients. On arrival at the emergency treatment unit, child was alert and febrile with a loud inspiratory stridor which became more prominent with crying. He was tachypnoeic with a respiratory rate of 44 breaths per minute, without intercostal or subcostal recessions and clear lungs. Saturation was maintained on room air. His heart rate was 136 beats per minute. Rapid antigen test for COVID-19 was positive. A possible diagnosis of laryngotracheobronchitis in a child with COVID-19 infection was made. He was nebulized with racemic epinephrine. IV dexamethasone (0.15mg/kg) and nebulization with budesonide (0.5 mg) was also given. Symptoms improved over next 24 hours with treatment and child was discharged two days after admission.

Discussion: New clinical presentations of COVID-19 are emerging. Further research is warranted to determine optimal management of croup in COVID-19 positive patients.

AN UNUSUAL CASE OF DROWNING AT SEA MIMICKING A SALTWATER CROCODILE ATTACK

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Introduction: There are two main species of crocodiles in Sri Lanka, the marsh crocodile (*Crocodylus paluster*) and the saltwater crocodile (*Crocodylus porosus*). Deaths due to crocodile attacks are relatively uncommon. There are no published reports of deaths due to saltwater crocodile attacks in the sea of Sri Lanka, to the best of our knowledge. We report the first case of saltwater crocodile attack in the sea of Sri Lanka.

Case report and discussion: A diver in the shallow sea off the western coast of Sri Lanka, was “rescued” following an attack by a crocodile. He was pronounced dead on admission to the emergency department of the local hospital. Autopsy revealed patterned bite marks mainly on the left shoulder, thorax and arm. There were no fatal injuries. Features of drowning were present. There was no evidence of natural disease or poisoning. The cause of death was confirmed as drowning. Similar to this case, the most common cause of death in crocodile attack is drowning, followed by, traumatic injuries to vital organs and haemorrhage. The probability of survival depends on the victims’ position during the attack and the size and weight of the crocodile. A threat of a possible human-crocodile conflict in the local region affecting the tourism and fishing industries was observed after this case. Prompt involvement of all relevant authorities is essential to prevent such further attacks and deaths.

A RARE CASE OF PERNICIOUS ANAEMIA WITH HEPATOSPLENOMEGALY

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Introduction: Pernicious anaemia is caused by vitamin B12 deficiency due to its malabsorption. Association of hepatosplenomegaly with pernicious anaemia is a rare occurrence, although its association with splenomegaly is commonly reported.

Case report: A 51-year-old patient with hypertension and hypothyroidism presented due to lethargy and malaise for 2- years duration. On examination, she was pale with hepatosplenomegaly. There was no lymphadenopathy. She had pancytopaenia and the blood picture showed macrocytic red blood cells suggestive of megaloblastic anaemia. Iron studies and coagulation profile were normal. Reticulocyte index was 0.64 indicating a hypo proliferative bone marrow. Lactate dehydrogenase was elevated. Ultrasound scan of the abdomen confirmed hepatosplenomegaly. Gastric biopsy showed chronic antral gastritis with chronic corporal gastritis and glandular metaplasia. Her intrinsic factor antibody was positive. She was diagnosed with pernicious anaemia and started on an intramuscular vitamin B12 supplement. The repeat full blood count performed one month after initiating vitamin B12 supplementation showed improvement in pancytopaenia. Repeat ultrasound scan of the abdomen showed reduction of the sizes of the liver and spleen.

Discussion: Pernicious anaemia is a cause of megaloblastic anaemia due to vitamin B12 deficiency. Intrinsic factor is produced by parietal cells of the gastric lining and it is needed for the absorption of vitamin B12 from the terminal ileum. Autoantibodies are produced against Intrinsic factor in pernicious anaemia which impairs vitamin B12 absorption leading to megaloblastic anaemia. This can lead to splenomegaly but hepatosplenomegaly is reported very rarely.

GAUCHER'S DISEASE

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Introduction: Gaucher's disease is inherited as an autosomal recessive disorder. There is glucocerebrosidase enzyme deficiency, leading to reduced metabolism of glucocerebroside. It is more commonly seen in the Ashkenazi Jewish population. In Sri Lanka, it is rarely seen in the paediatric age group and very rarely detected in the adult population.

Case report:
A 52-year-old diagnosed patient with epilepsy presented with a generalized tonic-clonic seizure while on anti-epileptic drugs. He also had mild cognitive impairment for 1-month duration and abdominal discomfort. There was no history of constitutional symptoms, fever, past history of recurrent infections or easy bruising. He was pale and had moderate splenomegaly on examination, but there was no hepatomegaly or lymphadenopathy. Investigations revealed pancytopaenia. Contrast enhanced CT scan revealed gross splenomegaly with multiple low attenuating lesions. Liver and renal function tests, erythrocyte sedimentation rate, lactate dehydrogenase level, 2D Echocardiogram and iron studies were normal. EEG showed feature of generalised epilepsy. Serum protein capillary immunotyping electropherogram showed IgG kappa bi-clonal gammopathy. Bone marrow aspiration and trephine biopsy showed Gaucher's cells. Beta glucosidase enzyme level was low, leading to the diagnosis of Gaucher's disease. Patient was not treated with enzyme replacement and is presently being followed up for the development of complications.

Discussion: Gaucher's disease is a rare disorder to present in adulthood. Identification of patients is important as close monitoring for development of complications needs to be carried out.

WHEN TO "THINK ECTOPIC": A CASE REPORT ON HETEROTOPIC PREGNANCY IN A PATIENT WITH SPONTANEOUS CONCEPTION WITH NO RISK FACTORS

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Introduction: Heterotopic pregnancy (HP) is a rare and potentially fatal condition where simultaneous gestations occur at multiple implantation sites. It rarely occurs in spontaneous conception but it is becoming commoner with assisted reproduction techniques. Identification of an intra-uterine pregnancy (IUP) can divert attention from the possibility of a concurrent ectopic pregnancy (EP). A delayed diagnosis can result in complications.

Case report: Primigravida with no risk factors for EP presented at 5+2 weeks following spontaneous conception. Initial Trans-vaginal-scan (TVS) raised suspicion of a tubal pregnancy. Patient had abdominal pain without peritonism, while maintaining haemodynamic stability. Serum beta HCG was 1326mIU/ml on admission and was raised by 88% within 48 hours. TVS revealed a gestational sac with minimal free fluid in the pouch of Douglas. Suspicion of HP was raised and a diagnostic laparoscopy was arranged to confirm the suspicion. A right tubal ectopic was identified and salpingectomy was performed. Post-operative and repeated TVS in two weeks revealed a growing IUP.

Discussion: Early diagnosis of HP can be difficult and a careful sonographic examination is the key to diagnosis. In the presence of a re-assuring IUP, HP is easily misdiagnosed as a luteal cyst. Difficulty of an accurate clinical and sonographic diagnosis in asymptomatic patients can justify laparoscopy as a diagnostic & therapeutic tool. The ectopic component is treated surgically, whereas the intrauterine component is expected to develop normally. In conclusion, this rare entity of HP in a woman with no risk factors illustrates the importance of the dictum "think ectopic" even when an IUP is present.

A PATIENT WITH BETA-KETOTHIOLASE DEFICIENCY PRESENTED WITH SEVERE DEHYDRATION

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Introduction: Beta-ketothiolase deficiency (BKTD) is a rare autosomal recessive disorder. This enzyme is involved in the final steps of isoleucine catabolism and fatty acid oxidation pathway.

Case history: One-year-old previously healthy boy presented with vomiting and loose stools for one day. He was the second child of non-consanguineous parents. On examination, he was drowsy and tachypnoeic. He had features of severe dehydration. All the investigations were normal except severe metabolic acidosis with strongly positive urine ketone bodies. Capillary blood sugar was 120 mg/dL. There was a massive elevation of ketotic markers, 2-methylacetoacetate, lactic acid and marked excretion of dicarboxylic acids in Gas Chromatography Mass Spectrometry. When combining the history, examination and laboratory investigations, clinical presentation was more suggestive of BKTD. He showed a good response to fluid therapy with 10% dextrose and bicarbonate infusion. After 12-hours of intensive care, his blood gas analysis results normalized and his clinical condition improved. He was prescribed an isoleucine-restricted diet.

Discussion: Acute ketoacidotic episodes should be managed promptly, aiming at suppression of ketogenesis and correction of acidosis, avoiding rapid alkalinization. A favorable outcome can be achieved with a protein restricted diet. BKTD should be suspected in patients presenting with ketoacidotic attacks. Early diagnosis and treatment is imperative to achieve a good outcome.

MAPLE SYRUP URINE DISEASE IN A BABY WITH POOR WEIGHT GAIN

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Introduction: Maple syrup urine disease (MSUD) is an inborn error of branched chain amino acid (BCAA) metabolism. We report an infant with MSUD who presented with poor weight gain.

Case report: A two-week-old baby girl presented with poor sucking and excessive crying for ten days duration. She was the second child of non-consanguineous parents. Her antenatal period has been uneventful. She was delivered by an elective lower segment cesarean section at term due to past section. Postnatal period was uneventful and the birth weight was 2.87 kg. Baby was discharged on day two of life after establishing breastfeeding. On admission, she had 14% weight loss with a weak cry and cyclical movements of lower limbs. On examination, all four limbs were hypertonic with occasional opisthotonos posture. Other systemic examination was normal. No body odour was present. Her full blood count, c-reactive protein, liver function tests, renal function tests, ultrasound scan abdomen and cerebrospinal fluid analysis were normal. Guthrie test was suggestive of MSUD. Plasma amino acid analysis revealed high alloisoleucine (353 µmol/L), leucine (3731 µmol/L), valine (1047 µmol/L). Ultrasound brain showed mild cerebral oedema. An epileptic focus was noted in the electroencephalogram. She was managed by a multidisciplinary team. She was given oral thiamine, antiepileptics and BCAA-free formula. Baby improved with the given management.

Discussion: MSUD is a rare metabolic disease that should be suspected in neonates presenting with poor weight gain and excessive crying. A high clinical suspicion and an early diagnosis result in a better prognosis.

CEREBRAL TUBERCULOMA: A RARE PRESENTATION

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Introduction: Diagnosis of tuberculoma is difficult due to cerebrospinal fluid (CSF) analysis being normal unless meninges are inflamed. Obtaining a tissue biopsy is invasive. Magnetic resonance (MR) spectroscopy is useful and it shows a lipid peak in tuberculoma that can differentiate it from a brain tumor.

Case report: A 51-year-old previously healthy female presented with recurrent left facial involuntary twitching movements. She did not have any limb weakness. There was no fever, constitutional symptoms or contact history of tuberculosis. Cranial nerves and neurological examination of limbs were normal. Her CSF was acellular. Her magnetic resonance imaging (MRI) brain showed multiple ring enhancing lesions in the cortical region of the right parietal region with diffusion restriction on diffusion weighted imaging. This appearance was suggestive of a tuberculoma. MR spectroscopy findings were consistent with brain abscess formation due to tuberculosis. Diagnosis of cerebral tuberculoma was made. Patient was started on anti-tuberculous therapy for which she responded well.

Discussion: MRI brain finding of peripheral ring enhancement in lesions is important but not pathognomonic to diagnose tuberculoma. MR spectroscopy has a significant role in diagnosis of such cases seen on MRI brain. Central nervous system tuberculosis commonly manifests as meningitis and tuberculoma is a much rare presentation. Tuberculoma appears as a circumscribed granulomatous focus which causes a mass effect without entering the subarachnoid space. Brain biopsy can be performed when there is lack of substantial evidence to suggest active tuberculosis.

RESPIRATORY COMPLICATIONS OF TREATED PULMONARY TUBERCULOSIS PATIENTS DECLARED AS CURED: A CROSS-SECTIONAL STUDY

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Introduction and Objectives: Tuberculosis (TB) is a common disease worldwide. Due to high incidence of TB, number of post-tuberculosis survivors are high. As a result of permanent anatomical changes, many survivors experience post-TB sequelae. Therefore, we evaluated these patients after declared as cured to assess the respiratory complications and their impact on the quality of life.

Methods: A cross-sectional retrospective study was performed on 431 post-TB patients who were registered in the Chest Clinic, Kalutara from 2006-2009. Patients with pre-existing lung diseases were an exclusion criterion for the study. Data were collected using an interviewer-administered questionnaire that included socio-demographic factors, economic factors, patient's health conditions, and the St. George's Respiratory Questionnaire to assess the patient's quality of life. Patients were considered as obstructive airway disease (OAD) if FEV1/FVC <70% and FVC ≥80% predicted in the lung function. Restrictive lung disease (RLD) was diagnosed if FEV1/FVC ≥70% and FVC <80%.

Results: Among study participants, 16.24% (n=70) had OAD, 20.64% (n=89) had RLD and 9.2% (n=40) had mixed type of lung impairment. Co-morbidities were seen in 110 patients. Diabetes mellitus had a good relationship with complications. 52% (n=46) of patients who had complications had more than 12-weeks of symptoms before the diagnosis of TB. Out of patients with OAD, 98.5% (n=69) were smokers. According to the results of the Smoking had a significant relationship with the activity score and total score, and Type 2 diabetes had a significant relationship with symptom score and the activity score according to St.Georges Respiratory Questionnaire.

Discussion: According to results, we can conclude that the time gap between the appearance of symptoms and diagnosis increases the rate of OAD. The relationship of OAD with smoking habits and Type 2 diabetes is significant.

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