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ORAL PRESENTATIONS

OP 01

LEVELS OF VITAMIN D AND SELECTED CYTOKINES AS POTENTIAL PROGNOSTIC BIOMARKERS OF ACUTE ISCHAEMIC STROKE: A CASE-CONTROL STUDY

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Background and Objectives: Inflammation-derived oxidative stress is postulated to contribute to neuronal damage leading to poor clinical outcomes in acute ischaemic strokes (AIS). This study evaluated the role of selected serum immune-mediators (IL-1β, IFN-γ, IL-4, and vitamin D) in ischaemic stroke progression and their accuracy in predicting AIS prognosis.

Methods: Sixty AIS patients admitted to the National Hospital of Sri Lanka after stroke onset at <6, 6-24, 24-48, and 48-96 hours, and age- and sex-matched healthy controls (n=15/group) were recruited. Serum cytokine and vitamin D levels were quantified using Sandwich ELISA and Competitive ELISA, respectively. CombiROC analysis established optimal prognostic biomarker combinations for AIS. Patients were clinically evaluated for stroke severity on admission, based on the National Institute of Health Stroke Scale while 30-day functional outcome (FO) was assessed using the modified Rankin Scale (mRS; 0-2 =good FO, 3-6 =poor FO).

Results: Compared to controls, significant upregulation of serum IL-1β and IFN-γ was recorded at all stages after stroke onset ($p<0.05$). Conversely, upregulation of IL-4 was detected in the recovery phase ($p<0.05$). Polarization of the T_H1: T_H2 (IFN-γ:IL-4) cytokine ratio towards a T_H1 bias with AIS progression and towards T_H2 with AIS recovery was established ($p<0.05$). A majority of test and control subjects (69.3%) had insufficient serum vitamin D levels (<30 ng/ml). Lower serum levels of IL-1β and higher levels of IL-4 were associated with a good FO ($p<0.05$), while lower Vit D levels were related to poor FO ($p=0.001$). The triple-biomarker panel, IL-4- IFN-γ -Vit D, demonstrated excellent accuracy in predicting AIS prognosis with 100% sensitivity and 91.9% specificity.

Conclusions: We found a prototypical association between the immunologic mediators IL-1β, IFN- γ, IL-4 and vitamin D, and AIS prognosis. Supplementation with vitamin D may improve the prognosis of AIS.

OP 02

INCREASED MUSCLE RESISTANCE IN CHILDREN WITH CEREBRAL PALSY OBJECTIVELY MEASURED BY CLINICALLY APPLICABLE TECHNIQUE

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Background and Objectives: Increased resistance to passive movement is often associated with cerebral palsy (CP). Clinical assessments e.g., the Modified Ashworth Scale (MAS) lack reliability, affecting the accurate quantification of resistance. The hand-held assessment device is a major new development, intended to improve the reliability of assessments. The device measures joint displacement, force, and electromyogram activity providing measurements of overall passive stiffness (PS), passive resistance (PR), the non-neural components, and reflex-mediated active resistance (AR) the neural component of increased resistance. The reliability and validity of measurements of the device have not been tested in children with CP previously.

Objective: To investigate the validity and reliability of a clinically applicable assessment method for increased muscle resistance of children with CP, using a hand-held device.

Methods: A validation study among 50 normal and 50 kids with CP was conducted. Slow and fast dorsiflexion stretches of the ankle joint were performed using the device to evaluate PS, PR, AR, and ROM of the triceps surae muscle group.

Results: Average age of children with CP was 7.8 years. Their Gross motor function scales were distributed evenly across 5 grades. They were hemiplegic in 16, diplegic in 22, and balance were quadriplegic. Very good to excellent intra-rater (ICC 0.97) and inter-rater (ICC 0.97) reliability of PS measurements of the device were elicited. Mean PS values were significantly higher among the cases (34.23 SD±17.1) than controls (12.0 SD±6.9) (P<0.01). Further, a significant negative correlation was found in PS and ROM in both cases (P<0.01) and controls (P<0.01) while a significant positive correlation was found between PS and MAS (P<0.01) and PR (P<0.01). PS cut-off of 14.8 would identify CP with 88% sensitivity and 80% specificity.

Conclusions: The hand-held device provided valid and reliable measurements of PS and PR of the triceps surae muscle, in a group of children with CP.

OP 03

NEURO-ENDOCRINE CHANGES IN GUILLAIN-BARRE SYNDROME AS A CRITICAL ILLNESS AND THEIR RELATIONSHIP WITH SEVERITY

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Background and Objectives: To assess the neuroendocrine changes that occur in patients with GBS and their association with the severity of the disease.

Methods: A prospective study was carried out on all GBS patients who fulfilled the Brighton criteria, who were admitted to National Hospital of Sri Lanka over a 10-month duration. Details on demographics, clinical presentation, investigations, and treatment were obtained. Blood was tested for general haematology, biochemistry, and endocrine parameters. The results were analysed using SPSS and a comparison was made between mild and severe groups according to their modified Rankin Scores (mRS).

Results: Forty-nine patients who fulfilled the Brighton criteria for diagnosis of GBS were included in the study. Out of them, 27 were males (55.1%). Their mean age was 55 years. The nerve conduction studies confirmed 65.9% to have AIDP, 20.5% to have AMAN, and 13.6% as having non-specific F wave abnormalities. A mild disease was seen in 28.6% and a balance of 71.4%, suffered a severe disease.

Out of the statically significant parameters obtained, mean serum albumin level was 19.17 g/dL, and inversely correlated with the severity of the disease (Pearson correlation -0.548, <0.01). Mean serum 9 am cortisol level for the total population was 452.9 nmol/L and showed a direct correlation to the severity of disease (Pearson correlation-0.534, <0.01) and a significant difference between the means of mild and severe groups.

In males, mean testosterone level was 168.80 ng/dL and showed a significant inverse correlation with the severity of disease [Pearson correlation -0.657, <0.01) p=0.008]. There was no significant correlation between the age and severity of disease.

In females there was no significant difference in endocrine results between the groups.

Conclusions: There is no literature available on detailed endocrine changes in GBS other than syndrome of inappropriate ADH secretion. This study reveals a significant inverse relationship between serum albumin concentration and a direct relationship of serum cortisol concentration with severity. In males, serum testosterone concentration shows a significant inverse relationship with the severity independent of age.

OP 04

COGNITION, ELECTROENCEPHALOGRAM, VISUAL-EVOKED POTENTIALS AND PERIPHERAL NERVE CONDUCTION IN LONG-TERM EXPERIENCED MEDITATORS: A CROSS-SECTIONAL COMPARATIVE STUDY

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Background and Objectives: Meditation is a self-entrainment that influences neuronal plasticity. We compared electroencephalography (EEG) and visual evoked potential (VEP) wave patterns, measures of cognition and peripheral nerve conduction (NCS) parameters of long-term experienced meditators (LTEM) with meditation naïve controls (MNC) to assess central and peripheral neurological effects of long-term meditation.

Methods: Thirty LTEM (regular practice of >3 years) were selected using a validated intake interview. Thirty matched MNC were recruited from the community. All had a Montreal cognitive assessment (MoCA) score of

>26/30. A validated Sinhala version of the repeatable battery for the assessment of neuropsychological status (RBANS) was used to assess cognition. Using 10-20 system, EEG was recorded with eyes closed for one-minute followed by 19 minutes of meditation among LTEM while in MNC, total 20 minutes of EEG recording was in an eyes-closed relaxed state. EEG wave frequencies in both groups were analysed from six regions. Latencies for N75, P100, N145 were measured on VEP. Median and tibial nerve conduction velocities and amplitudes were recorded via NCS.

Results: Sum of index scores of cognitive domains were higher among LTEM than MNC: immediate memory, ($p<0.001$); visuospatial, ($p<0.001$); language, ($p<0.001$); attention, ($p<0.001$); delayed memory, ($p<0.001$).
In EEG, significantly higher right temporal alpha activity was observed among LTEM (44.5%) at rest compared to MNC (33.9%) ($p=0.005$). During meditation, frontopolar predominant higher frequency wave activity over left and right was found in LTEM (37.1% and 39.2%) compared to MNC (23.9% and 21.8%) ($p<0.001$).
NCS showed significantly higher conduction velocity (LTEM 41.9m/s, MNC 33.8m/s; $p<0.001$) and amplitude (LTEM 6.7mV, MNC 5.6mV; $p=0.007$) for tibial nerve among LTEM than MNC.VEP showed significantly shorter latencies bilaterally for all 3 deflections in LTEM than MNC (Right latencies of N75 , $p=0.006$; P100, $p=0.04$; N145, $p=0.035$: Left latencies of N75, $p<0.001$; P100, $p<0.001$; N145, $p<0.001$).

Conclusions: Long-term meditation enhances cognitive domains and produces significant changes in EEG frequencies, VEP and NCS.

OP 05

DIAGNOSTIC VALIDITY OF THE GENERAL MOVEMENTS ASSESSMENT (GMA) AND THE HAMMERSMITH INFANT NEUROLOGICAL EXAMINATION (HINE) IN THE EARLY DETECTION OF CEREBRAL PALSY IN HIGH -RISK INFANTS: A MODIFIED APPROACH FOR SRI LANKAN PATIENT CARE SETTING

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Background and Objectives: The General Movement Assessment (GMA) and the Hammersmith Infant Neurological Examination (HINE) are currently being used as a modified approach for the early detection of cerebral palsy (CP) among high-risk infants in Sri Lankan settings. This study was conducted to assess the diagnostic validity of the GMA, the HINE, and their combined use.

Methods: A cross-sectional study was conducted among 416 high-risk children, 24-42 months of age, attending the paediatric neurology clinic, Teaching Hospital, Karapitiya. Each child had undergone the GMA (writhing and fidgety movements) with or without the HINE in early infancy. CP status was assessed by a consultant paediatric neurologist. Diagnostic validity of the GMA was assessed based on sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), false positive rate (FPR), and false negative rate (FNR). Diagnostic validity of HINE and cut-off score for the detection of CP were determined using ROC analysis.

Results: The HINE had a strong diagnostic accuracy (AUC=0.876; 95%CI=0.802 to 0.950) at a cut off score of 57.5 with a satisfactory sensitivity (81.6%) specificity (83.1%), NPV (97.1%), FNR (18.4%) and FPR (16.9%). PPV of the HINE was low at 39.2%. Regarding the GMA, fidgety movements had a higher sensitivity (92.9%), specificity (87.7%), PPV (46.4%) and NPV (99.1%) and a lower FPR (12.2%) and FNR (7.1%), compared to writhing movements (sensitivity=84.6%, specificity=50.3%, PPV=16.7%, NPV=96.5%, FNR=15.4%, FPR=49.7%). By combining GMA and HINE scores, the sensitivity of the detection of CP increased up to 99.8% whereas other combinations also had very high sensitivity, writhing with fidgety (98.9%), HINE with fidgety (98.7%) and HINE with writhing (97.1%) respectively.

Conclusions: Both the GMA and the HINE had high diagnostic validity while the combination of GMA and HINE scores was identified as the best approach. Combination of the GMA and the HINE scores, an approach currently practiced, is recommended for Sri Lankan settings with this evidence.

OP 06

ACUTE SYMPTOMATIC SEIZURES AFTER STROKE IN A SRI LANKAN TERTIARY CARE CENTRE

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Background and Objectives: Seizure is a life-threatening complication of acute stroke. There is limited data on seizures after stroke from South Asia, and no data in Sri Lankan patients. We sought to evaluate the frequency and characteristics of acute symptomatic seizures (ASS) after stroke in a Sri Lankan tertiary care hospital.

Methods: Data of all consenting stroke patients with ASS admitted to the Stroke Unit of a Sri Lankan tertiary care centre over four years (July 2019 to June 2022) was analysed. Seizure mimics, and seizures due to systemic disturbances or non-stroke aetiologies were excluded. Demographic data, stroke characteristics, seizure characteristics, treatment and functional outcomes were obtained using interviewer-administered questionnaires. Descriptive analysis of data was performed using IBM SPSS version 22.

Results: Out of 416 admissions, 10 (2.4%) developed ASS [60% males, mean age (SD) 68.9 years (6.2)]. Ischaemic and haemorrhagic strokes were 5 (50%) each; 8 (80%) were right sided, and 7 (70%) each were cortical and large lesions. Mean admission NIHSS score (SD) was 14.33 (8.8). Four (40%) patients had seizures within first 24 hours; three (30%) presented with seizures as first symptom of stroke. Majority 6 (60%) had a single seizure; 2 (20%) had status epilepticus. Focal seizures were commoner (60%). All were treated with antiseizure medications; 7 (70%) were given multiple drugs. Majority [8 (80%) each] had poor functional outcome at discharge (mRS >2, BI <90). One patient died.

Conclusions: To our knowledge, this is the first data on ASS after stroke from Sri Lanka. Frequency of ASS in our study was similar to published data. Haemorrhagic, cortical, large strokes and those on right were commoner among those with ASS. Functional outcomes were poor in patients with ASS after stroke.

OP 07

THE TRIAD OF DRUG RESISTANT EPILEPSY, CORTICAL VISUAL IMPAIRMENT AND PARIETO-OCCIPITAL DAMAGE: AN OVERLOOKED EPILEPSY SYNDROME?

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Background and Objectives: Perinatal ischemia can damage and induce scarring in immature brain; especially in the parieto-occipital zone, founding epileptogenic foci. Additionally, occipital lobe damage may result in Cortico-Visual-Impairment (CVI). However, the syndromic association of CVI with epilepsy due to parieto- occipital damage is not defined as a specific entity. Therefore, in this series we aimed to delineate the characteristics of patients with Drug-Resistant-Epilepsy (DRE), CVI and parieto-occipital damage to establish the clinical triad.

Methods: Patients were recruited from an epilepsy-surgery clinic from 31/07/2020–31/07/2022. Seizure-semiology, magnetic resonance imaging and electroencephalograms were reviewed by the authors. CVI was graded using Roman-Lantzy's CVI Range assessment tool.

Results: 25 were included out of 173 clinic registrants. Mean age 9.08 years (1.5-20 years), males 20. All had perinatal complications; neonatal convulsions (56%), hypoglycaemia (44%) and sepsis (30%) were the commonest. All had Drug-Resistant-Epilepsy; 15/25 had focal-onset motor seizures, 05/15 with elementary visual aura. Generalized-onset seizures occurred in seven, and in the balance three the onset is unknown. Twenty four percent experienced daily seizures, 10/25 had weekly and 09/25 had monthly seizures. Severity of CVI was III in 56%, II in 28% and I in 4% .

All had posterior interictal epileptiform EEG activity, focal discharges in 21/25, (bilateral-independent discharges in 08), diffuse discharges in 04/25, and epileptic-encephalopathy in 06.

In MRI, parieto–occipital gliosis was seen in all, unilaterally in 19/25 and bilateral in rest, Occipital-Ulegyria was seen in 05/25; periventricular leukomalacia in two, colpocephaly and hippocampal sclerosis in one patient each. Sixty percent of those with occipital ulegyria had daily seizures. MRI and EEG findings were concordant in 20/25 patients. One

child underwent occipital lobectomy and achieved Engel class 1A outcome.

Conclusions: Triad of Drug-Resistant-Epilepsy, Cortical-Visual-Impairment and Parieto-Occipital damage is a specific entity. Cortical-Visual-Impairment should be an active diagnosis. Suggest screening at-risk populations to locate the triad.

OP 08

EFFECT OF THE CURRENT ECONOMIC CRISIS ON HEALTH AND HEALTHCARE SERVICES RELATED TO EPILEPSY

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Background and Objectives: Impending collapse of Sri Lanka's free healthcare system is likely to have a catastrophic effect on vulnerable groups such as epilepsy patients. To describe the effect of the economic crisis on health and healthcare related to epilepsy in persons with epilepsy (PWE), their caregivers and healthcare providers for PWE.

Methods: A cross-sectional study was performed among PWE, their caregivers and care providers at five hospitals of three different tiers in Western Province. Eighty patients and caregivers selected using systematic sampling from those attending clinic in each setting and all practicing neurologists (adult and paediatric) in the national healthcare system were invited to participate. The following were assessed by recall before and after the economic crisis: health of patients using modified 9-item questionnaire for epilepsy status and ILAE epilepsy control scale; patients' mental wellbeing using DASS21(adults) and Strengths and Difficulties Questionnaire (SDQ) for children; caregivers' mental wellbeing using Kessler Psychological Distress Scale (K10); and care providers' mental wellbeing using DASS21.
The healthcare provision was assessed using different questionnaires for PWE, caregivers and care providers. Availability of anti-seizure medications (ASM) in all hospitals served by a neurologist was assessed by a survey.

Results: Four hundred and seven patients and caregivers and 39 (78%) neurologists participated. Health in PWE indicated an increased seizure frequency in 31.1%, admission due to seizures in 14.75%, difficulty in obtaining ASMs in 90%, and missing ASM doses in 57.5%. Severe or extremely severe mental illness was seen in 16% of adult PWE and abnormal SDQ in 17.8% children. Thirteen percent of caregivers had indicated severe mental distress. 46% of care providers were suffering from mental distress; 22% severe.
Health care provision related to clinic reviews, patient admissions, transfers and provision of medications were significantly affected ($p<0.05$) after the crisis. Availability of anti-seizure medications was affected in all settings, abortive therapy for status epilepticus in 85%. All three care settings were equally affected except Lady Ridgeway Hospital where availability of ASM was less restricted.

Conclusions: The current economic crisis has substantial effect on the health of PWE and their caregivers. The data will be useful to exert pressure on administration towards maintaining essential services of our health-care system.

OP 09

INITIAL EXPERIENCES AND RESULTS FROM A COMPREHENSIVE PAEDIATRIC EPILEPSY SURGERY PROGRAMME AT THE NATIONAL EPILEPSY CENTRE OF SRI LANKA

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Background and Objectives: Drug-Resistant Epilepsy (DRE) is associated with serious consequences in children including, poor intellectual, psychosocial outcomes and poor health related quality of life. Epilepsy surgery is a proven treatment option for children with DRE.
National-Epilepsy-Centre of Sri Lanka (NECSL) is the state of art epilepsy management institute in Sri Lanka. Paediatric clinic was incepted at NECSL in July-2020 and considered the national reference centre for children with DRE. The initial experiences and the surgical results are presented herein.

Methods: This is a descriptive, retrospective cohort study. All clinic registrants from 31/07/2020–31/07/2022 were included. Medical records, video-EEG files and imaging studies were reviewed by the authors. Quality of life was assessed using the Quality of Life in Childhood Epilepsy (QOLCE). Behaviour was assessed using Strengths and Difficulties Questionnaire (SDQ).

Results: One hundred and seventy three children with DRE were registered, mean age 10.4 years, 58.3% were males.
Epilepsy-Surgery-Meeting (ESM) discussions were carried in 38/173 (21.9%), while 41/173 (23.7%) were awaiting ESM. 19/173 (10.9%) underwent surgery, 19/173 (10.9%) were lined up for surgery. Eleven children (6.3%) had pseudo-DRE.
Of the children who underwent surgery, 16/19 had focal motor seizures. All had MRI lesions, 02/19 underwent fMRI and 01/19 had Diffusion-Tensor-Imaging-Tractography. Semiology-Imaging and EEG were concordant in 15/19.
Four children underwent occipital lobectomies, eight tailored resections, two modified hemispherectomies, one modified temporal lobectomy and four Vagus-Nerve-Stimulator insertions.
All resections were done under neuro-navigation, guided by intra-operative electro-corticography (ECOG) monitoring. Mean follow up duration was 12.2 months.
Engle's class I outcome was seen in 13/19 patients, class II in 02/19 and class III in the four patients who had VNS insertions. QOLCE scores correlated with duration of seizures, epileptic encephalopathy and Engle's outcome. SDQ did not show significant change at postoperative 3-months review. Two had post-operative transient hemiplegia.
Histology revealed, Focal-cortical-dysplasia in eight, gliosis in two, dysembryoplastic-neuroepithelial-tumour in four and diffuse-astrocytoma in one.

Conclusions: Preliminary results demonstrated satisfactory post operative seizure control, better quality of life, with minimal complications in the paediatric cohort.

OP 10

UTILITY OF THE GENERAL MOVEMENTS ASSESSMENT (GMA) AND THE HAMMERSMITH INFANT NEUROLOGICAL EXAMINATION (HINE) IN THE EARLY DETECTION OF NON-CEREBRAL PALSY DEVELOPMENT DELAYS AMONG HIGH-RISK INFANTS

Hewawitharana GP[1], Darshana ILAN[2], Madhushani ULAN[1], Chathuranga DPS[1], Priyangika DLN[1], Madumadhavie RGY[1], Kumara RDS[1], Mihiran DKD[1], Nilukshika KVK,[3] Hewawitharana BDR[1], Wijesinghe CJ[2], Phillips J[4], Kodituwakku PW[4]
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Background and Objectives: The General Movement Assessment (GMA) and the Hammersmith Infant Neurological Examination (HINE) are currently being used for the early detection of cerebral palsy (CP) but not for non-CP development disorders. This study was conducted to assess the utility of these instruments in the early detection of non-CP development delays among high risk- infants.

Methods: A cross-sectional study was conducted among 368 high- risk children, 24-42 months of age, attending the Paediatric Neurology Clinic, Teaching Hospital, Karapitiya. Children diagnosed with CP were excluded from the study. Each child had undergone the GMA (writhing and fidgety) with or without the HINE in early infancy. Development delays were assessed using the Bayley Scales of Infant and Toddler Development (4th Edition), which is considered the gold standard of developmental assessment. Diagnostic validity was assessed for "cognitive", "speech and language" and "motor" impairment with the GMA, the HINE, and the combination of the two tests.

Results: The GMA (writhing and fidgety) or the HINE alone did not have a satisfactory degree of sensitivity in the early detection of non-CP development disorders (sensitivity less than 55% for all). In contrast, the Fidgety items on the GMA and the HINE had strong specificity (around 90% for cognitive and motor impairment) while writhing, fidgety and the HINE had strong negative predictive value (around 90% for cognitive and motor impairment and around 70% for speech and impairment). When the two methods are combined, sensitivity was increased up to 70% for detection of motor impairment and cognitive impairment but not for speech and language impairment. However, when combing all three methods, sensitivity exceeded 75% for the early detection of non-CP development disorder (87.4% for motor, 83.6% for cognition and 78.2% for speech and language respectively).

Conclusions: The GMA (writhing or fidgety) or the HINE alone did not have place in early detection of non-CP development disorders among high-risk

infants. However, the combined use of these instruments allows clinicians to identify children at-risk for developmental delays more effectively.

OP 11

IMPACT OF THE ECONOMIC CRISIS ON STROKE ADMISSIONS AND OUTCOMES IN A SRI LANKAN TERTIARY CARE CENTRE

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Background and Objectives: There is no data on the impact of the economic crisis in Sri Lanka on healthcare seeking behaviour and outcomes. We sought to evaluate the impact of economic crisis on admissions, delays in seeking care and functional outcome in stroke patients in a tertiary care hospital.

Methods: We studied all patients with stroke admitted to a tertiary care hospital during the economic crisis (March 2022-October 2022), and during the same 8-month period in a previous year (March-October 2019). Year 2020 and 2021 were not considered due to the impact of COVID-19 pandemic. Data on stroke admissions, patient characteristics, admission delays and functional outcomes were collected using interviewer-administered questionnaires. Data on economic status of the country was obtained from the Central Bank of Sri Lanka.

Results: Admissions were significantly higher during economic crisis period [2019 - 244, 2022 – 296; mean (SD) admissions – 30.5(10.97) in 2019, 37(12.36) in 2022, p=0.025]. Patients with admission delay >3h was significantly higher during the economic crisis [2019 - 43.4%, 2022- 56.6%, p=0.02]. Stroke severity was significantly higher during economic crisis [mean admission NIHSS score (SD): 2019- 8.36(6.46), 2022- 9.11(6.10), p=0.03].

More patients were functionally dependent both on admission and on discharge during the crisis period: admission modified Rankin Scale (mRS) 3-5: 2019- 41.5%, 2022- 58.5%, p<0.001, admission Barthel index ≤60: 2019- 42.6%, 2022- 57.4%, p=0.005; discharge mRS 3-5: 2019- 33.9%, 2022- 66.1%, p<0.001, discharge BI ≤60: 2019- 36.8%, 2022- 63.2%, p<0.001). No difference was noted in case fatality between the two periods.

Conclusions: This is the first data on stroke admissions during economic crisis in Sri Lanka. Stroke admissions were significantly higher, presentation delays were longer, stroke severity was higher and patients had more disability during the economic crisis period.

POSTER PRESENTATIONS

PP 01

ESTIMATION OF BODY WEIGHT USING ANTHROPOMETRIC PARAMETERS IN SRI LANKAN ADULTS

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Background and Objectives: Body weight is an important clinical parameter for accurate dosing of drugs with a narrow therapeutic window. This study aimed to validate existing anthropometrics-based weight estimation equations and develop a new equation for the same purpose for Sri Lankan adults.

Methods: This prospective study was conducted at the National Hospital of Sri Lanka, split into a development and a validation phase. During the development phase, estimated body weight of patients by doctors and nurses and patients themselves were noted and compared against their actual body weight. In addition, 13 anthropometric measurements were taken, which were used to validate 12 anthropometrics-based equations to estimate body weight described in literature previously. Two new gender specific regression models to estimate the body weight in the local population was also derived and validated.

Results: A total of 502 (males=249) and 217 (males=108) patients were recruited for the development and validation phases respectively. Both doctors and patients had comparable accuracy in predicting body weight ($p>0.05$). All anthropometric based equations were significantly correlated with actual body weight (correlation coefficients: 0.741 – 0.869), and the new equations derived from the local data performed similarly to the best performing equation identified from the literature during validation phase.

Conclusions: When the patient weight cannot be measured, an estimate by the patient may be the best substitute. If the patient cannot speak or is unconscious, the equations derived here may be the next best alternative for Sri Lankan adult males and females.

PP 02

A GENETICALLY CONFIRMED CASE OF MITOCHONDRIAL ENCEPHALPATHY LACTIC ACIDOSIS AND STROKE (MELAS) WITH COEXISTING NEMALINE MYOPATHY - FIRST REPORTED CASE IN THE WORLD

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Background: MELAS is a maternally inherited mitochondrial disorder characterized by encephalomyopathy, lactic acidosis, and stroke-like episodes. We report a case of MELAS with coexistent nemaline myopathy with genetic confirmation of both conditions. This is the first case in literature with coexistence of both these rare disorders.

Case Report: A 11-year-old previously healthy Sri Lankan male child, product of a non-consanguineous marriage with normal development presented with acute onset short lasting (three minutes each) recurring episodes (six episodes) of right sided eye deviation with impaired consciousness. In between episodes he regained consciousness. There was no fever, jerky limb movements, tongue biting or incontinence. Family history was significant for a similar presentation in the mother developed at the age of 36 years.

Examination was significant for short stature and proximal upper and lower limb weakness. His plasma and cerebrospinal fluid lactate were elevated. MRI brain had evidence of an acute infarction in the right occipital territory.

Targeted mitochondrial mutation analysis for MELAS revealed him to be heteroplasmic for a pathogenic variant in *MT-TL1* gene at m.3243A>G. Whole exome sequencing revealed compound heterozygosity for nemaline myopathy with pathogenic variants in the two alleles of *NEB* gene at c.20089G>A and c.11300A>G. His asymptomatic sister (8-year-old) and mother were both found to be heteroplasmic for MT-*TL1* gene variant.

Acute presentation was managed with antiepileptics, hydration and mitochondrial supplementation including co enzyme Q, creatine, carnitine, vitamin E and K, vitamin B complex, and arginine. Currently he

is stable on daily supplementation of arginine. He is being monitored closely both clinically and with serum lactate level.

Discussion: MELAS is a mitochondrial disorder with heterogeneous presentation. Currently there is no specific therapy. Arginine is effective in managing and preventing stroke-like episodes.

PP 03

A CASE OF SMITH-MAGENIS SYNDROME (SMS) DUE TO A *DE NOVO* POINT VARIANT IN THE *RAI1* GENE.

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Background: Smith-Magenis syndrome (SMS; OMIM 182290) is a rare autosomal dominant neurodevelopment disorder. It is characterized by intellectual disabilities, sleep disturbance, behavioural problems, and a variety of craniofacial, skeletal and visceral anomalies. The estimated prevalence of SMS is 1:25,000. While the majority of the cases are due to 3.5 Mb deletion on 17p11.2 of the *RAI1* (retinoic acid induced-1) gene, some patients carry small intragenic deletions or point variants.

Case Report: A 3 years and 6 months old girl was born as the only child to non-consanguineous parents. She presented with developmental delay. She could only speak single words. Behavioural issues including head banging, face slapping, bruxism, self-injurious behaviours and temper tantrums were noted. There was no history of sleep disturbance or seizures. On examination, she had subtle dysmorphic features including brachycephaly, midface hypoplasia, broad-square coarse face, low set ears, flat nasal bridge and knock knees. She had mild hypotonia and increase joint laxity. Gower's sign was positive. Her IQ level was slightly low. There were no organomegaly. Apart from a squint, she had no vision or hearing impairment.
Karyotype was 46, XX. Whole exome sequencing was performed which detected a likely pathogenic nonsense variant in the *RAI1* gene denoted as c.847C>T, p.Gln283* (rs2032120646), thus confirming the diagnosis of SMS. This is the first reported case of a Sri Lankan patient with SMS due to a point variant of *RAI1* gene.

Discussion: There are many undiagnosed cases of SMS due to single nucleotide or indel variants in the *RAI1* gene, as they will not develop full spectrum of the disease. Therefore, more clinical suspicion and sequencing for variants in *RAI1* gene in these patients who do not have a cytogenetically detectable deletion for SMS will help in diagnosis of the disease.

PP 04

EPILEPSY-RELATED KNOWLEDGE AMONG PUBLIC HEALTH MIDWIVES (PHMs); A STUDY FROM A SEMI-URBAN PROVINCE OF SRI LANKA

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Background and Objectives: Public Health Midwives (PHMs); are the first contact health care workers in the Sri Lankan hierarchy of public health. PHM is accountable for child and maternal-health of her area. Assessing the knowledge of PHMs on epilepsy related topics relevant to her target population is the scope of this study.

Methods: A descriptive; cross-sectional, cohort study was performed. Sample was selected using randomized stratified sampling method. Data were collected using a pretested, custom designed, self-administered questionnaire.

Results: The sample comprised a total of 262 PHMs; 104 (39.7%) admitted having prior knowledge on epilepsy, 105 (40%) had a kith-kin with epilepsy and 114 (43.5%) had a person with epilepsy (PWE) in her area. Sex and epilepsy: a minority of five (2%) believed PWEs should abstain from sex; nine indicated that epilepsy could deter sexual activity (3%), 73 sexual intercourse always facilitates seizures (27.9%); and marriage should be postponed till seizure control is achieved by13 (5%). Popular contraceptive options prescribed for a patient who is on anti-seizure medications (ASMs) were IUCD 196 (74.8%), condom 105 (40%), and permanent contraception 56 (21.3%). Regarding pregnancy and epilepsy: eleven felt ASMs should be stopped during pregnancy (4.2%); LSCS should always be the mode of delivery 67 (25.6%); breast feeding is contraindicated while on ASMs was indicated by five (2%). Regarding immunization: Japanese Encephalitis vaccine cannot be given for one

year post seizure was accurately indicated by 229 (87.4%); post vaccination fever can facilitate a seizure by 64 (24.4%).
Out of the cohort 131 (50%) has witnessed an acute convulsion; however, only 36 (13.7%) acknowledged to know the first aid. Service experience has a positive correlation with the knowledge, R =0.128 (P<0.05). Having a kith-kin with epilepsy t=2.40 (P<0.05), and a patient-with-epilepsy in the area t=2.18 (P<0.05) have enhanced epilepsy related knowledge.

Conclusions: Only a minority of PHMs had prior knowledge on epilepsy; service experience has significantly enhanced the knowledge. Majority have given correct answers under the domains of epilepsy versus sex, pregnancy and immunization. Unfortunately, only very few acknowledged to have knowledge on first-aid.

PP 06

A RARE CASE REPORT OF COVID-19 RELATED RECURRENT OPSOCLONUS MYOCLONUS ATAXIA SYNDROME

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Background: Opsoclonus myoclonus ataxia syndrome **(**OMS) is characterized by spontaneous arrhythmic conjugate saccades in all directions of gaze, multifocal myoclonus, and ataxia with or without other cerebellar signs. Nearly one third is paraneoplastic, the rest is para/post infectious, toxic-metabolic or idiopathic. Infective causes include mycoplasma pneumonia, streptococcus, Borrelia, HIV, cytomegalovirus, human herpes virus 6, arboviruses and enteroviruses.

Case Report: We present a case of OMS in a young female who presented with mild COVID-19. She developed neurological symptoms eight days after the onset of respiratory symptoms. Pharyngeal swab SARS-CoV-2 PCR was positive. As she was severely disabled, we treated her with intravenous immunoglobulin (IVIG), and she recovered completely by four weeks. Seven months later she again developed mild COVID-19 with positive PCR. Interestingly she developed OMS again on day five of respiratory symptoms. She responded well to IVIG. MRI brain and cerebrospinal fluid analysis were normal. Workup for an underlying malignancy was negative.

Discussion: SARS-CoV-2 infection is known to associate with a variety of neurological complications including OMS. However, OMS is limited to case reports. Respiratory symptoms do not corelate well with neurological symptoms.
The strong host immune response to the virus leads to a hyperinflammatory phase and the resultant immune response is thought to trigger OMS. Immune mediated pathogenesis is further supported by response to immunomodulatory treatment such as IVIG, plasma exchange and steroids.
To the best of our knowledge this is the first COVID-19 related OMS reported in Sri Lanka. Even among the globally reported cases recurrence of OMS with recurrent infection is not reported. Our case with recurrent OMS emphasizes the need to dissect the underlying pathophysiology of COVID-19 related OMS as genetics may have a role.

PP 07

FAVOURABLE OUTCOMES FOLLOWING THE USE OF IV IMMUNOGLOBULIN THERAPY IN A PATIENT WITH OSMOTIC DEMYELINATION SYNDROME

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Background: Osmotic Demyelination Syndrome (ODS) embraces the common entity of Central Pontine Demyelination and less common extra pontine demyelination, and is characterized by destruction of neuronal myelin sheaths leading to severe irreversible neurological deficit.
Publications on treatment of ODS is mostly limited to case reports. They mainly describe immunomodulatory therapy with IV immunoglobulin (IVIG), steroids and plasmapheresis, leading to favourable outcomes, often completed recovery. This is the first case report of an infant making complete recovery following treatment with IVIG alone.

Case Report: A 9-month-old previously well baby boy presented with a five day history of Rota viral diarrhoea. He was drowsy, tachypnoeic, and febrile and had severe hypernatraemic dehydration with serum sodium of 196 meq/L, metabolic acidosis and acute renal failure. During the initial management he had several episodes of rapid fluctuations of serum sodium concentration. The baby was referred to neurology with encephalopathy, gradual onset asymmetrical quadriparesis with exaggerated deep tendon reflexes and rotatory nystagmus. Clinical diagnosis of ODS was made and this was confirmed by MRI which showed

features of pontine and extrapontine demyelination. Immediate treatment was started with IVIG and the child completely recovered without any residual neurological deficit.

Discussion: Due to the variety of presentation and delayed imagine changes, ODS is initially not suspected in most cases. So, the prompt diagnosis and treatment may modulate its progress and long-term neurological outcome. Due to the rarity of the syndromes and clinical heterogeneity standard treatment guidelines are not available. However, considering the devastating nature of the disease IVIG therapy can be considered as an effective treatment modality in ODS in infants.

PP 08

KNOWLEDGE OF THE ANTI-SEIZURE-MEDICATION MANAGER ON THE PRESCRIBED REGIMEN

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Background and Objectives: Epilepsy needs long term regular treatment with Anti-Seizure Medication (ASM). Insufficient/wrong knowledge on ASM, can lead to poor event control and other unwanted health issues. Scope of this study is to determine the knowledge of the ASM-manager on the prescribed regimen. ASM-manager is defined as the key person who is responsible for the medication of the patient (self or by-proxy).

Methods: A descriptive; cross-sectional, cohort study was performed. Sample was selected from an epilepsy clinic in a semi-urban province of Sri Lanka. ASM manager was asked to answer a questionnaire based on the current ASM prescription. Given answers were crosschecked with the original prescription by the author.

Results: The sample comprised a total of 262 PHMs; 104 (39.7%) admitted having prior knowledge on epilepsy, 105 (40%) had a kith-kin with epilepsy and 114 (43.5%) had a person with epilepsy (PWE) in her area. Sex and epilepsy: a minority of five (2%) believed PWEs should abstain from sex; nine indicated that epilepsy could deter sexual activity (3%), 73 felt sexual intercourse always facilitates seizures (27.9%); and marriage should be postponed till seizure control is achieved by 13 (5%). Popular contraceptive options prescribed for a patient who is on anti-seizure medications (ASMs) were IUCD 196 (74.8%), condom 105 (40%), and permanent contraception 56 (21.3%). Regarding pregnancy and epilepsy: 11 felt ASMs should be stopped during pregnancy (4.2%); LSCS should always be the mode of delivery was indicated by 67 (25.6%); breast feeding is contraindicated while on ASMs was indicated by five (2%). Regarding immunization: Japanese Encephalitis vaccine cannot be given for one year post seizure was accurately indicated by 229 (87.4%); post vaccination fever can facilitate a seizure was stated by 64 (24.4%).

Of the cohort 131 (50%) had witnessed an acute convulsion; however, only 36 (13.7%) acknowledged to know the first aid. Service experience had a positive correlation with knowledge, $r = 0.128$ ($P<0.05$). Having a kith-kin with epilepsy $t=2.40$ ($P<0.05$), and a patient-with-epilepsy in the area $t=2.18$ ($P<0.05$), have enhanced their epilepsy related knowledge.

Conclusions: In a majority, ASM were managed by proxy managers. Majority claimed to know the dosing frequency; level of education of the ASM manager reduced the number of drug mistakes; polytherapy increased mistakes; the fewer the drug mistakes the better the seizure control.

PP 09

POINT OF CARE ULTRASOUND SCANNING TO MEASURE OPTIC NERVE DIAMETER IN TWO PATIENTS WITH INCREASED INTRACRANIAL PRESSURE

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Background: Monitoring of the intracranial pressure (ICP) is very important in decision making about patients who are suspected to have raised ICP. There are invasive and non-invasive methods of monitoring ICP. Bed side ultrasound measurement of optic nerve sheath diameter is one of the non-invasive methods.

Case Reports: Case 1 - 19-year-old boy managed for super refractory status epilepticus developed unequal,

poorly responding pupils. Necessity to undergo repeated ICP monitoring arose with need of repeated NCCT brain imaging.
Case 2 - 22-year-old girl with CSF culture positive CNS TB presented with lack of energy and on and off headache. MRI brain revealed obstructive hydrocephalus. Patient was started add-on anti TB medications. Plan was to closely observe her clinically rather than going for early neurosurgical intervention to reduce hydrocephalus.
We used monitoring of optic nerve sheath diameter as a surrogate marker of increased intra cranial pressure. In these two cases, we attempted to see whether the optic nerve diameter changes correlated with the radiological features.

Discussion: Frequent monitoring of ICP is challenging in certain clinical settings where clinical signs can be misleading. Invasive methods bare their risks and repeated imaging holds practical difficulties. We suggest USS guided measurement of optic nerve sheath diameter as a useful marker of increased ICP specially in a resource poor setting. Although the sensitivity of the study is operator dependent, frequent utilization of this method will make the clinicians more familiar with the technique and this can greatly help in day-to-day practice. Further studies will be useful in validating this method for the Sri Lankan population.

PP 10

CHIARI-TAC; A SURGICALLY CURED SYMPTOMATIC TRIGEMINAL AUTONOMIC CEPHALALGIA

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Background: Trigeminal autonomic cephalalgia (TAC) encompasses four primary headache disorders according to the International Classification of Headache Disorders 3β (ICHD3β). Rarely, TAC headaches may occur secondary to structural pathology. We describe a patient with TAC headache as the initial manifestation of Type 1 Chiari malformation, which resolved after decompressive surgery.

Case Report: A 36-year-old female presented with a one-month history of worsening headache. Headache frequency was 4-5 times per week. She described a severe, left side locked headache with associated redness and tearing of the ipsilateral eye. She also complained of numbness of the ipsilateral face and weakness of the body. Duration of the headache was unusual, lasting between 4-6 hours and occurring 2-3 times per day. She denied any increase in headache with the Valsalva manoeuvre. Examination revealed left- sided partial ptosis and conjunctival suffusion. However, there was no pupillary involvement. She had impaired pain sensation over the ophthalmic and maxillary divisions of the left trigeminal nerve. Fundoscopy was normal and there were no other cranial nerve abnormalities. The Hoover test uncovered that her hemiparesis was a functional neurological deficit.
Magnetic resonance imaging (MRI) of the brain revealed a 6mm tonsillar descent, compatible with the diagnosis of Type 1 Chiari malformation. She was started on a combination of indomethacin, verapamil, and prednisolone, to which she had suboptimal control of headaches. A neurosurgical input was obtained, after which she underwent successful cranio-vertebral decompression. She remains headache free at 2 months of follow up.

Discussion: All patients with trigeminal autonomic cephalalgia should undergo MRI as symptomatic TAC headaches may be indistinguishable from primary headache disorders. However, the presence of atypical features, such as in our patient suggest the former. We wish to highlight that although symptomatic TAC headaches are rare, they may be amenable to surgery with excellent outcomes.

PP 11

SURGICAL OUTCOME OF PATIENTS WITH PHARMACO-RESISTANT EPILEPSY AT THE NATIONAL EPILEPSY CENTRE IN SRI LANKA: A SINGLE CENTRE EXPERIENCE

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Background and Objectives: Epilepsy surgery is considered for drug-resistant focal epilepsy, and results in good outcome rates. The National Epilepsy Centre (NEC) in Sri Lanka which offers an epilepsy surgical program was established in 2017.

Methods: We report post-operative seizure outcomes, surgical complications, and impact on quality of life among 15 adult (≥ 16 years) patients who underwent epilepsy surgery at NEC. We used Engel classification and QOLIE 31 questionnaire to assess seizure outcome and quality of life (QOL).

Results: Six female and nine male patients underwent epilepsy surgery (mean age 27.7 years, range 16-44 years) from October 2017 to June 2022. The mean follow-up duration was 32 months (range 6-55 months). Ten patients underwent temporal lobe surgeries. At the 6-month follow-up, 80% (8/10) of them had favourable seizure outcomes with Engel class I/II. At 1-year follow up 6 patients (75%) and at 2-year follow-up, 5/7 patients (71%) had a favourable outcome. Five patients had extratemporal lobe surgeries, and one was lost to follow-up. The rest were seizure-free at six months. Out of them, three patients had one-year follow-ups, and two had 2-year follow-ups, all remaining seizure-free.
None had long-term post-operative neurological complications. Four patients (28.6%) experienced minor post-operative surgical site infection.
Meaningful improvement in QOL (change in QOLIE 31 score ≥ 11.8) was observed irrespective of seizure outcome or type of surgery. The mean difference of QOL among patients without favourable seizure outcomes was 13, whereas, among patients with a favourable outcome, the mean change was 24.8. Only one patient did not have a clinically significant change.

Conclusions: Epilepsy surgery is effective for selected group of patients with focal onset drug-resistant epilepsy. It is an option for patients in developing nations. Seizure outcomes in our patients were comparable to outcomes worldwide. Meaningful QOL improvement was observed in our series.

PP 12

RITUXIMAB THERAPY IN THE TREATMENT OF MULTIPLE SCLEROSIS: A SHORT CASE SERIES

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Background and Objectives: The availability of Disease Modifying Therapies (DMTs) for multiple sclerosis (MS) remain limited in developing countries such as Sri Lanka. While interferon beta-1a is the DMT of choice for patients with Relapsing Remitting Multiple Sclerosis (RRMS), rituximab is increasingly being used for patients with a more aggressive disease course. The use of rituximab has been reported as a cost-effective strategy in the management of MS in other developing countries.
Objective: To highlight the option of rituximab as a DMT for multiple sclerosis due to its efficacy, wide availability and relative low cost.

Methods: Five patients diagnosed with RRMS using the 2017 McDonald criteria were selected for treatment with rituximab at a dose of 500mg every 6-12 months. At the time, four patients were being treated with interferon beta-1a but had had a clinical relapse and/or new lesions seen on Magnetic Resonance Imaging while on treatment. The remaining patient was treated with rituximab at the first presentation due to the severity of clinical signs and the high lesion load on imaging, suggesting a more aggressive form of the disease.

Results: Rituximab infusions were well tolerated in all five patients. At 9 months post-treatment, all patients had not experienced any clinical relapses. The Expanded Disability Status Scale remained unchanged. Repeat MRI study of one patient showed a reduction in T2 signal intensities. Gadolinium enhancement of lesions couldn't be elicited due to the non-availability of MRI contrast at the time.
One patient suffered from an opportunistic fungal infection of the lung as a side effect of drug induced immunosuppression, 6-weeks post initial treatment. The patient responded to anti-fungal therapy and made a satisfactory recovery. No cases of severe SARS-CoV-2 infection were recorded.

Conclusions: Rituximab appears effective and safe in the treatment of MS in developing countries like Sri Lanka where affordability plays a crucial role in the choice of DMT for each patient.

PP 13

PAEDIATRIC STROKES. THE CLINICAL AND AETIOLOGICAL SPECTRUM. A RETROSPECTIVE CROSS SECTIONAL STUDY FROM THE NEUROLOGY DEPARTMENT OF A TERTIARY CARE CHILDREN'S HOSPITAL, COLOMBO, SRI LANKA.

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Background and Objectives: The clinical spectrum and aetiological factors vary widely among different

geographical regions worldwide. There is no data from Sri Lanka as there have not been any studies on paediatric strokes up to now.
To determine the clinical and etiological spectrum of paediatric strokes in Sri Lanka.

Methods: The study was conducted as a retrospective observational study with the use of clinical data of patients admitted and referred to the neurology department of Lady Ridgeway Hospital, Sri Lanka. Children from one month to fourteen years with the diagnosis of stroke were recruited for the study for a period of three years from January 2019.
On the basis of history of presenting complaint, clinical examination, relevant laboratory investigations and radiological imaging, aetiology for the strokes were determined. Appropriate statistical methods were used to analyse the data.

Results: The most common clinical presentation from a total of 72 cases was of hemiparesis or motor weakness (n=47, 67%) followed by seizures (n=18, 25%) and speech abnormality which could be either slurring or aphasia (n=5, 7%). Vasculopathies were the most common aetiological factor (n= 56, 78%) from which inflammation counted the most, followed by thromboembolic events (n= 7, 10%) and post-surgical (n- 5, 8%).
Ischaemic strokes accounted for most of the cases studied (n=61, 84%). The remainder were of haemorrhagic strokes (n=8) and there were three cases of cerebral venous sinus thrombosis.

Conclusions: The commonest aetiology of paediatric strokes presented in this study were ischaemic strokes due to intracranial vasculopathy.

PP 14
ACUTE ISCHAEMIC STROKE SUCCESSFULLY MANAGED WITH THROMBECTOMY IN A RESOURCE LIMITED SETTING – A SINGLE CENTRE EXPERIENCE

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Background: Management of acute ischemic stroke (AIS) has revolutionized over the years. With the advent of intravenous thrombolysis, neurons in the penumbra are salvageable to a larger extent with excellent long term functional outcomes. In Sri Lanka, many hospitals have the facilities necessary to administer intravenous thrombolysis for patients, presenting within 4.5 hours.
Thrombectomy is performed for patients with large vessel occlusions in the anterior circulation usually after administration of intravenous thrombolysis. Thrombectomy needs special expertise, catheter-laboratory facilities and equipment making its use limited to a few specialized centres even in more developed countries.

Case Reports: We report three patients with AIS involving the middle cerebral artery (MCA) territory aged 35, 55 and 60 years who were successfully managed with emergency thrombectomy in a timely manner. Diagnosis of large artery occlusion was made by clinical presentation and non-contrast CT brain. They underwent thrombectomy at 3.5 hours, 4 hours and 4.5 hours respectively with no post-surgical complications. While one patient had excellent functional outcome with a modified Rankin score (mRS) of 1 at 3 weeks, other two patients had satisfactory outcome with mRS of 3 and 4 respectively. We believe that in all three of them catastrophic progression to malignant MCA territory infarct was prevented.

Discussion: It was a team effort primarily involving the neurologists and interventional radiologists. We report these cases with the intention of encouraging specialized teams to utilize this highly effective form of treatment when dealing with appropriate cases of AIS, overcoming numerous obstacles and limitations, as the outcomes can be excellent.

PP 15
FAMILIAL ATAXIA SYNDROME: FRIEDREICH ATAXIA GENETICALLY CONFIRMED BY A LOCALLY DESIGNED MOLECULAR DIAGNOSTIC TEST

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Background: Friedreich ataxia is the commonest hereditary ataxia accounting for approximately 50% of all cases. We describe two siblings with cerebellar ataxia who fulfil the clinical criteria of this syndrome. The diagnosis was confirmed by a Polymerase chain reaction (PCR)-based genetic assay designed by the authors.

Case Report: An 18-year-old girl presented with progressive unsteadiness and recurrent falls over a 1-year period. Examination revealed thoracic scoliosis, ataxic gait and bilateral dysmetria. There was symmetrical wasting of distal lower limbs, with absent ankle reflexes and an extensor plantar response. Romberg's sign was positive with loss of joint position sensation bilaterally. Although she had dysarthria, there was no nystagmus or deafness. Precordial examination was abnormal, and echocardiography revealed hypertrophic obstructive cardiomyopathy (HOCM). Ophthalmologic evaluation demonstrated thinning of the retinal nerve fibre layer. Cerebellar and upper cervical cord atrophy was noted on Magnetic Resonance Imaging (MRI) and nerve conduction studies revealed axonal sensory neuropathy. Screening for diabetes was negative.
Interestingly, the patient's elder sister had identical clinical features with symptoms onset at 16 years of age. Unfortunately, this sibling had a sudden cardiac death at the age of 32 years. Their parents were non-consanguineous. This suggested an autosomal recessive inheritance, with the most likely diagnosis being Friedreich ataxia. A molecular diagnostic test targeting this diagnosis was conducted and found to be positive.

Discussion: Causation of Friedreich ataxia is due to repeat expansion of GAA in intron 1 of the frataxin (*FXN*) gene. We performed a PCR based genetic test using primers designed to anneal to the flanking regions of the abnormal repeats. PCR products visualized by Agarose gel electrophoresis revealed bands indicative of GAA repeat sequence, confirming our diagnosis. We wish to highlight the importance of making a phenotypic diagnosis in hereditary neurological disorders, to guide genetic testing. This will enable utilization of available resources at low cost.

PP 16

OSMOTIC DEMYELINATION SYNDROME FOLLOWING RAPID FLUCTUATIONS IN THE GLYCAEMIC STATUS

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Background: Osmotic demyelination syndrome (ODS) is a life-threatening neurological disorder characterized by non-inflammatory demyelination in the pontine and extra-pontine regions. It has been a recognized complication of rapid correction of hyponatremia. However, a variety of other conditions have been associated with the development of ODS, independent of changes in serum sodium. It has seldom been reported as a complication of rapid correction of hyperglycaemia. Here, we report a case of osmotic demyelination syndrome in a patient with rapid fluctuations in the glycaemic status.

Case Report: A 51-year-old male with poorly controlled diabetes mellitus, hypertension, ischaemic heart disease, and CKD stage III presented with unsteadiness, slurred speech, and dysphagia for one week. Two weeks back he has been admitted to a local hospital with osmotic symptoms and generalized body weakness and was found to have a random plasma glucose level of 515 mg/dl and treated with insulin infusion. Blood sugar was corrected to 265 mg/dl within twenty-four hours. The neurological examination revealed cerebellar signs on the left side with gait ataxia and bilateral partial ptosis without ophthalmoplegia or pupillary abnormalities with a normal examination of the upper and lower limbs. The magnetic resonance imaging of the brain revealed heterogenous T2 high signal intensities in the central pons with diffusion restriction suggestive of central pontine myelinolysis. The patient improved with supportive management.

Discussion: This case report suggests that variations in plasma glucose levels can cause osmotic demyelination as a result of abrupt shifts in osmolality, independently of the serum sodium levels, and highlights the necessity of avoiding abrupt corrections of any metabolic alteration that may influence osmolality.

PP 17

CREUTZFELDT JAKOB DISEASE A GREAT MASQUERADER IN NEUROLOGY- A SINGLE UNIT EXPERIENCE

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Background: Creutzfeldt Jakob disease (CJD) is a great masquerader in neurology, mimicking various neurological disorders. Here we describe three cases of probable CJD with different presentations encountered at our neurology unit.

Case Reports:

Case 1: A 64-year-old hypothyroid man presented with reduced responsiveness for two days. Further history revealed subtle memory impairment, visual hallucinations and psychotic symptoms for two months. He had bilateral asymmetrical myoclonic jerks without obvious startle myoclonus. Anti- TPO antibody titre was > 100 IU/ml but he had poor response to immunotherapy. Although the initial EEGs were nonspecific, later he developed periodic sharp waves. CSF studies were normal. MRI-DWI and FLAIR revealed high signal in thalamus and striatum.

Case 2: A 72-year-old-male initially developed rapidly progressive left eye painless vision loss two months ago. He had altered behaviour subsequently including wandering, aggression, visual hallucinations and reduced cognition. He developed drowsiness, generalized rigidity and frequent myoclonic limb jerks. His MRI brain revealed T2/FLAIR hyper-intensities with minimal diffusion restriction of both caudate and lentiform nuclei with serial EEGs showing Periodic lateralized epileptiform discharges (PLEDs).

Case 3: A 64-year-old lady presented with acute onset left homonymous hemianopia. Her initial neuroimaging was normal. Within two months she developed rapidly progressive dementia and admitted with drowsiness. She later developed myoclonus. Initial EEG showed diffuse slowing and serial EEG revealed PLEDs. Her repeat MRI showed T2/FLAIR hyperintensities in the basal ganglia with diffusion restriction and cortical ribboning.

Discussion: The above cases highlight the variable clinical manifestations and nonspecific preliminary investigational findings, a major challenge when diagnosing CJD. However, evolution of neuropsychiatric features and serial EEG changes facilitate the diagnosis. Hence high index of suspicion is needed to diagnose CJD especially in the resource poor setting with the unavailability of CSF biomarkers.

PP 18

OUTCOMES AND CHALLENGES OF ACUTE STROKE THROMBOLYSIS; A SINGLE TERTIARY CARE CENTER EXPERIENCE

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Background and Objectives: Thrombolysis for acute ischemic stroke is performed in 31 centres across all provinces in Sri Lanka. We report our thrombolysis experience over a 2-year and 10-month period, from a high influx neurology unit at the National Hospital of Sri Lanka.

Methods: This was a prospective study conducted between January 2020 to November 2022 on all patients who received thrombolysis for acute ischemic stroke in our unit. Data was recorded regarding demographics, stroke severity, timing of alteplase (rTPA) administration and clinical outcomes.

Results: Of the 166 patients (69% male, mean age 61 years) thrombolysed over this period, there were 8.4% (n=14) young strokes and 3.6% (n=6) were above 80 years of age. Most patients had moderate severity stroke with a median NIHSS of 10. There were 12% (n=20) of minor strokes. The majority (54%) of patients received alteplase between 3 and 4.5 hours of symptoms onset. The average door to needle time was 1 to 2 hours with only 4.8% (n=8) achieving the target of ≤60 minutes. Time from arrival to CT scanning took 41-60 minutes on average, with only 3.6% (n=6) being imaged within 25 minutes of arrival. However, 54% (n=83) had a favourable outcome (MRS 0-2) after 24 hours of thrombolysis. There were no significant differences in outcome according to age, stroke severity or time to thrombolysis. There were no reports of allergy to alteplase. Haemorrhagic transformation occurred in 9% (n=15) and 3.6% (n=6) had minor bleeding manifestations. There were only 3% (n=5) deaths attributed to thrombolysis.

Conclusions: Despite less-than-ideal timings of imaging and thrombolysis, we report good outcomes in our patients, regardless of age and stroke severity. Rates of complications were comparable to worldwide figures. Thus, our thrombolysis practice is safe and efficacious with room for improvement in timeliness of delivery.

PP 19

BRAINEOCARE: A LOW-COST AND USER-FRIENDLY EEG MONITORING SYSTEM FOR SCREENING NEONATAL SEIZURES

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Background and Objectives: The neonatal period is the most vulnerable time for seizures to develop. Seizures in the immature brain lead to detrimental consequences, therefore early diagnosis and prompt management are needed. Continuous electroencephalography (cEEG) is the gold standard for neonatal seizure detection. However, cEEG and clinical expertise are often limited to resourceful settings while countries with limited per capita expenditure on health services lack such technology. Hence, cost-effective cEEG with latest technologies will help clinicians to make accurate diagnosis and advocate prompt treatment.
This study aims to develop a cost-effective seizure monitoring device capable of long duration real-time monitoring of EEG specifically considering features of neonates.

Methods: A novel user-friendly EEG headset and a wireless dry-contact electrode-based EEG signal acquisition system was developed as a reusable design. The headcap design ensures safety and comfort of the neonate while allowing adequate scalp contact with an adjustable design to support varying dimensions of head sizes. Acquisition hardware includes a battery-operated portable design with the capability of storing recordings in backup storage.
To support seizure detection, we tested an explainable machine learning algorithm on a separate dataset with the intention of integrating to the device at a later stage. This algorithm was fed with extracted features corresponding to seizure instances from a previously published neonatal EEG dataset.

Results: The proposed machine learning algorithm can differentiate ictal periods from non-seizure EEG patterns with an accuracy of 89.5% with sensitivity and specificity values of 86.35% and 92.56% respectively. We have designed and 3D printed the EEG headcap and fabricated the printed circuit board for EEG acquisition.

Conclusions: A proof-of-concept design developed to address critical limitations in currently used cEEG systems is presented. Further testing and confirmation of performance will help medical professionals in low-resource settings to diagnose neonatal seizures efficiently.

PP 20

CLINICAL AUDIT ON INVESTIGATION AND MANAGEMENT OF NEONATAL SEIZURES

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Background and Objectives: Current literature on neonatal seizures is predominantly based on studies from Neonatal Intensive Care Units. Aetiology of neonatal seizures vary according to time from birth. This study aimed to understand aetiology and audit the management of neonatal seizures presenting to a tertiary paediatric facility.

Methods: A prospective study was performed at the Lady Ridgeway Hospital to audit the current management of neonatal seizures. First half of study was performed during pre-COVID period in 2019 and completion of study was during the first six months of 2022. Babies admitted to general medical wards or the Premature Baby Unit of Lady Ridgeway Hospital after being discharged from the delivering institution were recruited.

Results: Thirty-one babies were reviewed; seven born pre-term. Median age at presentation was five days (1-29). Fifty four percent reported some form of preceding concern in the peripartum. Nearly 50% experienced less than five seizures; 30% had numerous seizures (>10). The commonest attribute as cause was CNS infection however, none had positive isolates. Other aetiologies included possible hypoxic event in five, maternal abstinence in two, possible inborne error of metabolism in one, congenital structural abnormality in one, intracranial haemorrhage in one, no obvious cause identified in the balance. Majority did not undergo a vigorous evaluation. Only 12 had at least a routine 30-minute EEG, none were monitored for longer duration. Ultrasonography was the universal imaging offered, only 3 underwent MRI and another 3 CT brain. None had any genetic evaluations. Only two babies were observed without treatment. Eighty-four percent were given initial medication via intravenous route; 93% receiving phenobarbitone. However, only 37.5% received a loading dose. Therapeutic drug monitoring was unavailable.

Discussion: Considering the developments and advances in neonatal neurology, evaluation of neonatal seizures has a large room for improvement. Detailed EEG monitoring and investigations to identify the aetiology should be pursued. Efforts should be made to avoid both over and under treatment since both are detrimental to the developing brain.

PP 21

A GENETICALLY AUTHENTICATED SRI LANKAN CASE OF GLYCYL-TRNA SYNTHETASE 1 (*GARS*1) RELATED HEREDITARY NEUROPATHY WITH VARIABLE PHENOTYPE IN THE SAME FAMILY

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Background: Hereditary motor sensory neuropathy (HMSN) is the most common inherited neuropathy which is clinically and genetically heterogeneous. Next generation sequencing (NGS) based whole exome sequencing (WES) is being increasingly utilized for precise molecular diagnosis. We report a genetically confirmed case of *GARS*1 related neuropathy with variable phenotype within the same family.

Case Report: A 36-year-old previously healthy female, product of a non-consanguineous marriage presented with progressive weakness and wasting of right hand for three years. She reported mild affect to instrumental activities of daily living. Her 65-year-old father is having progressive distal muscle wasting of bilateral upper and lower limbs with moderate difficulty in instrumental activities and mobilization for more than 30 years. Rest of the systemic inquiry was unremarkable. There were no abnormal environmental or toxin exposures.

Examination of the patient revealed wasting and weakness of the small muscles of the right hand with normal sensation and reflexes. Rest of the neurological examination was normal. Nerve conduction study (NCS) was normal, but electromyography showed chronic denervation in the right first dorsal interosseous, abductor pollicis brevis and deltoid muscles. MRI scan of brachial plexus and cervical spine were unremarkable.

Examination of her father revealed distal wasting and weakness of bilateral upper and lower limbs with bilateral foot drop. Ankle reflexes were absent with normal sensory findings normal. NCS was compatible with motor and sensory axonal neuropathy with predominant motor involvement.

WES revealed her to be heterozygous for a likely pathogenic variant in *GARS*1 gene at c.643G>C (p.Asp215His). Sanger sequencing confirmed her father to be harbouring the same variant.

Discussion: *GARS*1 gene variants implicate in an autosomal dominantly inherited neuropathy with variable phenotype even within the same family. This could be either distal spinal muscular atrophy or HMSN phenotype like in the case of our patient and her father.

PP 22

NEUROMYELITIS OPTICA SPECTRUM DISORDER (NMOSD) ASSOCIATED WITH OVARIAN TERATOMA - AN UNUSUALLY AGGRESSIVE FORM SENSITIVE TO RITUXIMAB THERAPY

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Background: Neuromyelitis optica spectrum disorder (NMOSD) is found to be paraneoplastic in only 3-5% of cases[1]. Within this category, NMOSD associated with ovarian teratoma is a neurological rarity. We report a case of seropositive NMOSD with ovarian teratoma, with aggressive disease despite tumour resection.

Case Report: A 19-year-old girl underwent resection of a large ovarian mass, detected during evaluation for persistent nausea and vomiting. This was histologically proven to be an ovarian teratoma with elements of neuroepithelium. She continued to have persistent symptoms and developed ataxia within two weeks of surgery. Magnetic resonance imaging (MRI) revealed a T2/FLAIR hyperintensity in the dorsal medulla extending to the pons and cerebellar peduncles suggestive of demyelination. Anti- aquaporin-4 (AQP-4) antibodies were positive with negative anti- NMDA receptor antibodies. She responded to intravenous methylprednisolone pulse therapy. Despite maintenance corticosteroids, she relapsed within three weeks with bilateral visual loss, neuropathic pruritus, and episodic tonic spasms. MRI spine revealed multiple short segment hyperintensities in the cervical cord. Bilateral visual evoked potentials (VEP) were delayed with only perception of hand movements. There was no significant response to repeated steroid pulse therapy or therapeutic plasma exchange. A PET-CT scan excluded tumour recurrence. We then administered intravenous rituximab in divided doses to which she had excellent recovery with return of visual acuity to 6/36 bilaterally. On follow up she remains neurologically stable and functionally independent on maintenance immunotherapy.

Discussion: There are several common clinical features of NMOSD with ovarian teratoma reported in the literature which were demonstrated in our patient. This includes age less than 30 years, presentation with area postrema syndrome and presence of neural tissue in the teratoma[2]. However contrary to previous cases our patient continued to have aggressive disease despite tumour resection. We wish to highlight the value of rituximab in recalcitrant disease especially in those with seropositivity.

PP 23

SEIZURE REMISSION FOLLOWING EPILEPSY SURGERY IN A PATIENT WITH LATE ONSET LESIONAL LENNOX-GASTAUT SYNDROME: A CASE REPORT

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Background: Lennox-Gastaut Syndrome (LGS) is an epileptic encephalopathy with a peak age of onset at 3-5 years. It is characterized by multiple, generalized seizure types and an inter-ictal electroencephalogram (EEG) showing bilaterally synchronous slow spike-and-wave, and progressive cognitive regression. Surgical options in LGS can be either curative or palliative.

Case Report: A 12-year-old girl having multiple seizure semiologies with a significant seizure burden despite five anti-seizure medications was referred for evaluation. She had an antecedent history of right focal clonic seizures with impaired awareness associated with left frontal empyema at the age of eight years. After a six-month quiescent period, she experienced generalized tonic clonic, atonic and absence seizures.
Her inter-ictal EEG showed numerous long runs of generalized high voltage spike wave discharges at 1.5-2Hz frequency and an irregular persistent slow background. The ictal EEG was generalized electro-decrement with or without brief runs of generalized alpha range activity with myoclonic seizures and 1.5-2Hz spike wave runs with absences. A 3T MRI brain showed left middle frontal gyrus gliosis.
She underwent resection of gliotic tissue under electrocorticography (ECoG) guidance and MRI neuro navigation. At three months post-op she remains seizure free. Her EEG demonstrated complete abolishment of epileptiform discharges, however with background slowing.

Discussion: Resective surgery can be promising in LGS patients with localized seizure foci identified on EEG even in the absence of corresponding MRI lesions. However, our patient did not demonstrate focality on EEG during pre-surgical evaluation.
Initial focal semiology at the onset supported seizure origin to be of left frontal. This case lends support to the concept of LGS phenotype as being a network epilepsy, where key cerebral networks become autonomously unstable and that cortical lesions can establish and maintain this abnormal unstable network behavior. Thus, resection of the lesional zone can abolish the focus of seizure genesis.

This case highlights the importance of considering lesionectomy in LGS despite failure to demonstrate clinico-electro-anatomical concordance even at a later point of evaluation.

PP 24

ATTITUDES AND EPILEPSY-RELATED KNOWLEDGE AMONG PRIMARY SCHOOL TEACHERS; A STUDY FROM A RURAL PROVINCE OF SRI LANKA

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Background and Objectives: Children with epilepsy (CWE) are an educationally vulnerable group due to many unjustifiable reasons. Positive attitudes and epilepsy-related knowledge of teachers are crucial in healthier educational experiences and safety of CWE. To identify attitudes and epilepsy-related knowledge among primary school teachers from a rural province of Sri Lanka.

Methods: A descriptive; cross-sectional, cohort study was performed. Sample was selected using a randomized stratified sampling method from the rural North-Western-Province of Sri Lanka. Data were collected using a pretested, custom designed, self-administered questionnaire as the tool.

Results: The sample comprised a total of 373 teachers; commonest age category was 20-29 years (42.4%), 77.2% (n=288) females. Amongst all, only 7.2% (n=27) teachers professed to have adequate knowledge on epilepsy. Eighteen percent (n=68) was known to a patient with epilepsy, 55% (n=205) had witnessed epileptic-seizures. Said variables independently associated with better scores on attitudes and knowledge; perceived adequate knowledge (p=0.042), known patient with epilepsy (p=0.0001), witnessed an epileptic seizure (p=0.0001).

Merely 41.3% (n=154) acknowledged epilepsy as a neurological disease. Few 4.3% (n=16) considered epilepsy as a mental disease, and 7.0% (n=26) believed it a supernatural disease. Majority 60.6% (n=226) accepted epilepsy as a non-contagious disease.

Symptomatology of epilepsy was correctly identified only by very few (n=3). Amongst all, 29.4% (n=110) alleged epilepsy a medically controllable disease.

Minority 2.5 % (n=84) believed CWE always have lower IQ, and 63% (n=235) suggested them to follow main stream education. 67 % (n=250) of teachers were prepared to accept CWE in their class room with no uncertainty. Few 29% (n=108) considered CWE as a risk to other children in the class room. Forty five percent (n=166) wanted epilepsy to be fully under control before attending school. Twelve percent (n=43) deemed confident on first aid management of a seizure.

Conclusions: Knowing a patient with epilepsy enhanced positive attitudes and epilepsy related knowledge. Only a minority were confident about the knowledge they have. Majority of the teachers are willing to accept a CWE into their class.