

STROKE & NEUROVASCULAR DISORDERS

P424

SPONTANEOUS RECURRENT BILATERAL VERTEBRAL ARTERY DISSECTION: TWO DIFFERENT OUTCOMES OF THE SAME EVENT

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Introduction: Recurrent dissections have been described in patients with connective-tissue disorders. Recurrence risk is highest in the first months, persists for at least 10 years, and involves different arteries. The aim of this report was to present two patients with spontaneous bilateral vertebral artery (VA) dissection.

Patient 1: A 10-month-old boy presented with irritability, vomiting, and torticollis with left hemiparesis. MRI (4th day) revealed ischemic infarcts in cerebellar hemispheres and thalamus. MRA showed disappearance of flow signal in the right VA from its origin.

Diagnosis: Vertebral dissection. The baby was put on anticoagulant agents. MRA (at 6 months) demonstrated no recanalization of the right VA and filiform stenosis of the left. A digital angiography showed bilateral IVA occlusion.

Final diagnosis: Spontaneous recurrent bilateral VA dissection.

Patient 2: A six-year-old boy presented with acute occipital headache, gait ataxia, and strabismus. MRI revealed ischemic lesions in cerebellar hemispheres and thalamus. MRA demonstrated no blood flow signal of the left VA. Second event: two months later, the boy had acute headache, vomiting, and gait ataxia. MRI showed new ischemic strokes. A digital angiography confirmed bilateral VA dissection with pseudoaneurism and intramural thrombus at the left V4 segment. He was put on anticoagulant agents. Common connective-tissue disorders were ruled out in both patients. Other major arteries were studied and were normal.

Conclusion: We believe that underlying structural arterial-wall pathology may be a risk factor for spontaneous recurrent dissection in these patients.

P425

VASCULAR MALFORMATIONS OF THE SPINAL CORD: PRESENTATION OF A CASE SERIES

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INTRODUCTION: Spinal vascular malformations (SVM) are rare and still underdiagnosed entities. If not properly treated, SVM typically lead to progressive spinal-cord symptoms and myelopathy. Depending on the type of vascular malformation, presenting symptoms may vary between acute or progressive myelopathy. The aim of this study was to describe the clinical, imaging features, management, and outcome of nine patients with SVM.

METHODS: We reviewed 14 medical records and MRIs of patients with SVM seen between 2003-2013.

RESULTS: Seven patients had arteriovenous malformations, one a pial, and another duralarteriovenous fistula. Mean age at presentation was 5.9 years. Most-common symptoms were: paresthesia (5), lower-limb pain and weakness (5), sphincter involvement (2), and cervical bruit (2). MRI of patients with arteriovenous malformations showed flow voids and bright intramedullary lesions in T2 according to the region involved. Perimedullary arteriovenous fistulas usually occurred on the spinal-cord surface and were supplied by medullary arteries. Digital angiography confirmed the diagnosis. Endovascular embolization was performed in five and combined embolization and surgery in two. In one patient no treatment was possible. Outcome: After a 6-8 month follow-up, patients had no more paresthesia or pain and showed slow and progressive motor and sensory improvement and sphincter control. Four patients with arteriovenous malformations were monitored with digital subtraction angiography and no recurrences were observed.

CONCLUSIONS: SVM are rare entities to be considered in the face of a variety of neurological symptoms. Clinical suspicion together with MRI and digital angiography allow for diagnosis and early treatment. Surgery and/or embolization are the management of choice.

P427

CLINICAL PRESENTATION OF MOYAMOYA DISEASE AND ITS ASSOCIATION WITH NEUROIMAGING AMONG BANGLADESHI CHILDREN.

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Objectives: To observe the common clinical presentation & its association with neuroimaging abnormality in MoyaMoya disease.

Methods: This cross-sectional study was conducted among children under 18 yrs & moyamoya was diagnosed on the basis of RCMJ criteria. Their clinical and neuroimaging findings were analyzed.

Results: 20 children diagnosed as moyamoya disease were enrolled. The mean age was 7.24 ± 3.34 (3-14) years. Male: Female ratio was 1.2:1. The majority (n=18) had ischaemic stroke. 16 (80%) exhibited hemiparesis and 4 had alternating hemiparesis. The other presentations were dysarthria (n=13), visual impairment and convulsion (5 each), headache, altered sensorium, ataxia, involuntary movements in each of 3 cases, papilloedema and abnormal vital signs in each of 2 cases. Among presenting features, convulsions, altered sensorium, alternating hemiplegia, papilloedema and abnormal vital signs were significantly (p < 0.5) associated with abnormal CT scan. Stroke risk factors or FHO stroke in 5/20 (p<0.3) and previous experience of TIA in 9/20 (p<0.01) were strongly associated with subsequent occurrence of stroke in under and over 5 yrs children respectively. "Puff of smoke" appearance on MRA was present in 19 cases. One case was diagnosed through cerebral DSA. Bilateral ICA and MCA stenosis was most commonly observed abnormality followed by ACA and PCA.

Conclusion: Most cases presented with ischaemic stroke. About half of the cases previously experienced TIA, which foretells the evaluation for moyamoya through angiogram. Other clinical features correlated with the presentations of moyamoya reported elsewhere.

P428

FIBROCARILAGINOUS EMBOLISM AS A CAUSE OF SPINAL CORD INFARCTION A CASE REPORT.

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Introduction: Fibrocartilaginous embolism is a rare cause of spinal cord infarction, caused by an acute vertical disk herniation of the nucleus pulposus material followed by a retrograde embolization to the central artery.

Objective: Describe a case of spinal cord infarction after fibrocartilaginous embolism in a teenager admitted in Hospital das Clinicas, SP.

Case Description: We present a previously healthy 11-year-old girl with sudden onset of back pain and progressive paraparesis within 24 hours after a fall on her coccyx during roller skating. The patient presented with flaccid paraplegia with anesthesia below T12 level, associated with urinary retention and bowel dysfunction, with fecal incontinence. Admission laboratory investigation, cranial TC and cerebrospinal fluid analyses were normal. Spinal magnetic resonance showed a low medular lesion, between T11 level and the medular cone, with features suggesting ischemic injury. There were also signs of degeneration of the nucleus pulposus between T7-8 and T8-9 and Schmorl's nodes in T12 level, suggesting the etiology of fibrocartilaginous embolism. The patient was discharged after investigation to rehabilitation, maintaining the same neurologic exam.

Discussion/Conclusion: The accurate diagnosis of fibrocartilaginous embolic spinal cord infarct is usually post-mortem. The clinical presentation after an innocuous trauma, associated with laboratorial exams and neuroimaging are consistent with spinal cord infarct. The increased axial load during trauma with concomitant Schmorl's nodes may embolize to the vascular supply of the spinal cord causing infarction. In cases of paraparesis with no other evidence than trauma,

pediatricians should consider the clinical diagnosis of fibrocartilaginous spinal cord infarction.

P429**POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME: RETROSPECTIVE ANALYSIS OF A PEDIATRIC INTENSIVE CARE UNITY**

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Introduction: Posterior reversible encephalopathy syndrome (PRES) is a specific clinic and radiologic entity first defined by Hinchey in 1996 and posteriorly refined by Casey in 2000, characterized by the presence of headache, consciousness impairment, seizures and visual abnormalities associated with an often symmetric transient vasogenic white matter edema which predominantly involves the posterior part of the brain. A variety of factors can precipitate PRES. However, studies in the pediatric group are restricted. Here we describe our experience in a pediatric intensive care unit along ten years.

Methods: Retrospective survey of patients with diagnosis of PRES, focusing on etiology and MRI results, from May 2003 to August 2013 in Instituto da Criança do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo.

Results: We found fifteen confirmed cases in ten years. The median age was 10 years old (yo), range from 4 to 17 (yo). All subjects had acute elevation of blood pressure and focal seizures; 20% had visual symptoms and vomit and 33% had headache before presenting with seizure and conscious impairment. None of them went to status epilepticus and 80% had decreased level of consciousness. All underwent brain CT and MRI and 87% had involvement of the posterior part of the brain, 53% of the gray matter and 40% had ischemic complication. Two patients died in other hospitalizations and other two had neurological sequelae (epilepsy and behavior change).

Conclusion: Even with ischemic events the rate of complications is low.

P430**STURGE-WEBER: A RETROSPECTIVE STUDY OF 28 PATIENTS**

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Introduction/Objectives: Evaluate the most common clinical features of children and adolescents with neurocutaneous disease Sturge-Weber.

Methods: The records of 28 patients who were diagnosed with Sturge-Weber disease at CENEP-HC-UFPR in the period 1970 to 2013 and collected data regarding the changes more commonly found in this condition were evaluated.

Results: Of the 28 patients, 12 were male and 16 were female. The mean age at first visit was 63.3 months (1 to 163m). Three patients didn't have the typical hemangioma of the disease but had radiological changes compatible. In addition to injuries in face, lesions in trunk (7), neck (5), MMSS (3), MMII (4), dorso (1) and lumbar (3) were observed. About 28,5% of patients had a diagnosis of glaucoma. Epilepsy was present in 53,5% of patients, and of these, 40% refractory to anticonvulsant treatment. Headache was present in 21,4% of the sample, 33,3% related to seizures. Approximately 53,5% of patients had electrographic changes being 33,3% irritative activity. Cognitive impairment was found in 17,8% of patients and psychiatric disorders in 14,2%.

Conclusion: Besides the skin changes, Sturge-Weber disease can course with comorbidities such as epilepsy, glaucoma, mental retardation and psychiatric disorders, which must be diagnosed and treated early.

P431**STUDY OF STROKE IN CHILDHOOD AND ADOLESCENCE**

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Introduction: The Organização Mundial de Saúde (1978) definem Acidente Vascular Cerebral (AVC), Como UMA síndrome Clínica los Opaco Há UMA Manifestação de Sinais de Disfunção Neurológica Agudos UO focal global, duracao Superiores de 24 Horas, o Opaco PODE UMA causar morte do Paciente, com nenhuma outra Causa Opaco Localidade Localidade: Não vascular . Muito se Fala nenhum Acidente vascular cerebral los adultos e Pequeno Acidente vascular cerebral los crianças e adolescentes com, com sequelas Sepulturas, muitas Vezes Deixando prejuízo de Parágrafo a Vida dividido PODE O Curso Ser Acidente vascular cerebral isquêmico e hemorrágico los

Métodos: Estudo retrospectivo. Pacientes 0-18 Anos com Diagnóstico de Acidente vascular cerebral, Nao TRATADO Hospital Infantil de San Antonio (HCSA)

Resulta: Pacientes aproximado de 91,5% DOS tinham Menos de 1 Ano de idade enquanto 74,5% tinham Opaco ATÉ 1 mês de idade. Em Relação AO peso de nascimento 68,1% tinham Mais de 2.000 ge 31,9% tinham Menos de 2000g. 68,1% DOS PACIENTES los 1 Minuto Apgar abaixo de 7. 42,6% tinham etiologia da anoxia neonatal, Doença Cardíaca Congênita 29,8% e 27,7% OUTRAS Causas de 60% DOS PACIENTES apresentaram paresia UO hemisferio completo. 45% DOS PACIENTES tiveram convulsões focais generalizadas UO.

Conclusão: Tem Sido demonstrado Opaco UMA maioria das Mães apresentavam idade Acima de 19 Anos Localidade: Não Momento do Nascimento. tambem Localidade: Não houve Diferença significativa Segredos los Relação AO Fazer sexo Uma maioria DOS PACIENTES ERAM Mais Jovens do Opaco Ano de Vida e na maioria DELAS ERAM Recém-Nascidos.

P432**RESTING-STATE FUNCTIONAL MAGNETIC RESONANCE IMAGING OF MOTOR NETWORKS IN PERINATAL STROKE**

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Background: Perinatal stroke causes hemiparetic cerebral palsy and lifelong disability. Arterial (AIS) and venous (PVI) lesions damage motor pathways, but differences in timing and location dictate unique developmental plasticity trajectories that determine functional outcome. Emerging models are defining central therapeutic targets, but lack understanding of the integrated network that ultimately determines motor function. We demonstrate that resting state functional magnetic resonance imaging (rs-fMRI) can identify motor neural networks in children with perinatal stroke.

Methods: Ten children (6 males, mean 14.5yrs, 4 AIS, 6 PVI) were included from a population-based cohort with MRI-confirmed unilateral perinatal stroke and mild-to-severe motor disability (Melbourne Assessment: 74-100%; Assisting Hand Assessment: 52-100%). Participants underwent 3T MRI (GE750W) including anatomical imaging and six minutes rs-fMRI. The time-course of rs-fMRI signal in the primary motor cortex (M1) of the non-lesioned hemisphere was extracted and compared to all brain voxels using a General Linear Model. Descriptive analyses assessed relative connectivity between lesioned and non-lesioned M1 and supplementary motor area (SMA) and potential associations with motor function.

Results: Rs-fMRI motor networks from the non-lesioned M1 were obtainable in all subjects. In PVI children, non-lesioned M1 appeared more connected to contralateral, lesioned M1 than to SMA. The opposite trend was suggested for AIS (figure). Motor outcome appeared positively associated with relative connectivity to contralateral M1. Seeding from lesioned M1 demonstrated motor networks but was challenging in AIS.

Conclusions: Rs-fMRI can assess motor networks in children with perinatal stroke. Understanding developmental plasticity motor network patterns could further define central therapeutic targets.

P433**FAMILIAL MOYAMOYA IN 3 SIBLINGS WITH CONFIRMED C.14576G>A VARIANT MUTATION IN RNF213 GENE OF SOUTH ASIAN (BANGLADESHI) DESCENT**

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Introduction: Moyamoya disease (MMD) is a rare idiopathic cerebrovascular disorder characterized by progressive stenosis of the internal carotid arteries (ICA) leading to ischemic or hemorrhagic events. Due to higher incidence of familial occurrence in East Asian populations (Japanese, Korean and Chinese), RNF213 has most recently been identified as a MMD susceptibility gene with the c.14576G>A variant mutation being associated with the highest risk of MMD with odds ratio >250. However, there appears to be a gradient prevalence of this variant even amongst East Asians (> 79% in Japanese and Koreans, only 23% in Chinese), but thus far, no reported linkage in Southeast Asians or Caucasians has been reported. The dosage of the c.14576G>A variant mutation has been purported to predict the severity of clinical phenotype homozygotes having early onset and aggressive disease course presenting before age 4-years with ischemic stroke.

Case Description: We report a South Asian (Bangladeshi) non-consanguineous family with both parents having confirmed heterozygous c.14576G>A polymorphism of RNF213, and all 3 children having homozygous mutations of the same variant. Two of the 3 children presented with ischemic stroke and confirmed MMD at age 5 and 14 years. The youngest child (currently 7 years) is asymptomatic but has suspicious unilateral focal narrowing of the right ICA that could portend early MMD. Both parents in their 40s have reported TIA-like events, but no MMD on neuroimaging. This is the first report of siblings and parents of South Asian descent with confirmed RNF213 variant mutations of different doses.

Conclusions: RNF 213 gene mutation may be more prevalent in MMD in other ethnic groups also in addition to East Asians and testing is merited, particularly in familial cases.

P434

PEDIATRIC ARTERIAL ISCHEMIC STROKE IN A SERIE OF 63 PATIENTS: CLINICAL PROFILE, RISK FACTORS AND SEVERITY

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Introduction: Stroke is a cause of childhood disability. The aim of this study is to describe the clinical profile and risk factors in a serie of patients.

Methods: Children aged between 29 days -18 years were enrolled (1989-2012). Clinical characteristics and risk factors were described by age group. PedNIHSS severity score was estimated and correlated with the presence of risk factors, by ordinal and linear regression models.

Results: From 63 patients (average age 3,52 years) 67% were male. The most common symptoms were motor deficit (78%) and seizures (47%). The main risk factors were arteriopathy (60%) and infection (40%). Prevalence of systemic conditions was higher in children <3 years ($p < 0,05$). An average PedNIHSS score of 7.4 (0-17) was obtained. There is no significant relationship between the number of risk factors and PedNIHSS score. In a linear regression model, the presence of acute disorders of the head and neck was significantly correlated with PedNIHSS score ($p < 0,05$).

Conclusions: We confirmed male predominance and motor deficit at debut as previously described. We found at least one risk factor in all patients with complete information. We confirmed arteriopathy as the main risk factor, but we did not found any case of focal transient arteriopathy, may be because of lack of follow up neuroimaging. Searching for all risk factors is important, because the number of them is not predictable by severity score, but is important for recurrence risk. Acute disorders of the head and neck are predictors of higher PedNIHSS score.

P435

RISK FACTORS FOR PERINATAL ARTERIAL ISCHAEMIC STROKE: A CASE-CONTROL STUDY

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Introduction: Perinatal arterial ischaemic stroke (PAIS) is an important cause of morbidity and mortality. The pathophysiology and associated risk factors are not completely understood.

Objective: Describe the clinical presentations of PAIS and to evaluate associated risk factors.

Materials y methods: Case-control study. We evaluated clinical data of patients with PAIS controlled in our center between 1993 and 2012. Each infant with PAIS was matched to three healthy control. Risk factors were studied using univariate and multivariate conditional logistic regression analysis.

Results: We analyzed 40 patients (66,7% male, 33,3% female). The mean gestational age was 39weeks. 21(52,5%) of cases were confirmed as perinatal arterial ischaemic stroke(PAIS) and 19(47,5%) presumed perinatal arterial ischaemic stroke(PPAIS), the mean age of diagnosis were 6 days and 2 years-3month, respectively. The most frequent clinical presentation were seizures(86%) for PAIS and focal neurologic signs(95%) for PPAIS. Strokes preferentially involved the MCA territory(88%), 95% unilateral, 65% in the left hemisphere. All patients presented some neurologic deficit in the following clinical controls, the most common were hemiparesis and epilepsy. Significant risk factors in the multivariate analysis ($p < 0,05$) were nulliparity (OR 11.74; CI 3.28-42.02), emergency caesarean section (OR 13.79; CI 3.51-54.13) and Apgar score (5 min) ≤ 7 (OR 13.75; CI 1.03-364.03).

Conclusions: The principal clinical profile were seizures in PAIS and focal neurological signs in PPAIS. The prognosis was in general poor, all patients presented neurologic alterations. The risk factors nulliparity, emergency caesarean section, and Apgar score (5 min) ≤ 7 were found to be important risk factors in PAIS.

P436

EVOLUTION AND PROGNOSIS IN PEDIATRIC ARTERIAL ISCHEMIC STROKE IN A SERIE OF 64 PATIENTS

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Background and aims: arterial ischemic stroke is a uncommon but severe cause of pediatric disability. Our aim is evaluate prognosis and evolution in patients was diagnosed with arterial ischemic stroke between 1 month and 18 years on San Borja Arriarán Hospital in Santiago, Chile.

Methods: study design was descriptive-retrospective toward search on clinical records from 1989 to 2012.

Results: were included a number of 64 patients with arterial ischemic stroke between 1 month and 14 years. 55/64 patients was a single episode and 9/64 had recurrence of the event. 3 patient no had sequels. Of the 61 patients with sequels, 65 % (40/61) evolutionate with hemiparesis, 4/61 with tetraparesis and 1/61 diparesis. 6 patients had extrapyramidal symptoms and 4 with cerebellum symptoms. Epilepsy was evident in 23/61 patients, while 17 patients presented cognitive impairment in different grades.

Conclusions: is evident the high degree of sequels in our population, mostly in the motor área. The mortality was low than international reports. Is necessary to find new methods to prevent or ameliorates the impairment of the childrens affected with the disease.

P437

ARE METHYLENETETRAHYDROFOLATE REDUCTASE POLYMORPHISMS A1298C AND C677T GENETIC RISK FACTORS FOR PEDIATRIC STROKE AND IS THERE DIFFERENCE BETWEEN BOYS AND GIRLS?

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Introduction: Boys have been found to have higher risk for pediatric stroke. One of important genetic risk factors for pediatric stroke is considered to be the decreased activity of methylenetetrahydrofolate reductase (*MTHFR*), which is caused by polymorphisms of C677T and A1298C. The aim of the study is to examine the role of two *MTHFR* polymorphisms as pediatric stroke risk factors and investigate possible gender differences.

Method: 96 randomly chosen stroke patients (F=44; M=52), 59 with perinatal and 37 with childhood stroke, participated. Population-based control group was 300 healthy people (M=150, F=150), ages 18-30. DNA testing for *MTHFR* 677C>T, 1298A>C mutations were

performed. *MTHFR* genotypes were determined by PCR using specific primers.

Results: Any mutation in either or both of the loci was found in 86% of patients. c.1298A>C and c.677C>T polymorphisms have higher tendency amongst boys with perinatal stroke than girls, 28.8% vs 23.7% (OR=1.96) and 30.5% vs 23.7% (OR=4.13) respectively. Boys with childhood stroke also have higher tendency for c.677A>C mutation than girls (35.9% vs 27%; OR=5.14), although girls have higher tendency for c.1298A>C (27% vs 21.6%, OR=5.95). Comparison with controls showed significantly higher tendency for c.677C>T polymorphisms in children with stroke (57.3% vs 46%).

Conclusion: Our data support the hypothesis that *MTHFR* polymorphisms represent an important genetic risk factor for pediatric stroke. *MTHFR* polymorphisms were higher in boys, whereas boys had higher polymorphisms for C677T and girls for A1298C. Our findings suggest that higher stroke rate in boys could be caused by higher genetic risk factors.

P438

EPILEPSY AFTER PRESUMED PERINATAL STROKE

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Introduction: The aim of the study was to find out the incidence and predictive factors of epilepsy after presumed perinatal stroke (PPS). Children with PPS present with congenital hemiparesis or focal seizures beyond first month of life and have chronic focal brain lesion with vascular origin on brain imaging.

Methods: Patients were recruited from the Estonian Paediatric Stroke Registry. At least two neuroradiologists confirmed the radiological diagnoses. The study group consisted of 34 children (9 boys, 25 girls). Twenty-two children (61%) had unilateral periventricular venous infarct and 12/34 had stroke in the territory of the medial cerebral artery. Left hemisphere was the prominent side in 26 cases (76%). Mean follow-up time was 9.9 years (range: 2.0 to 7.6 years).

Results: Nine (26%) children developed epilepsy with focal onset seizures. Mean onset of epilepsy was 3.9 years, range: 9 months to 7.2 years. The Kaplan-Meier probability of remaining seizure-free at 4 years was 82%. The onset of epilepsy was not associated with sex, type of the brain damage, size of the arterial stroke, prematurity or severity of the motor deficit ($P > 0.05$). Left side of the damage was associated, but not significantly with onset of epilepsy ($P = 0.077$).

Conclusions: Epilepsy occurs in 26% of children with PPS. No predictive factor for epilepsy was found in this study, probably because of the small study group.

P440

ISCHEMIC STROKE IN CHILDHOOD

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The average incidence of childhood stroke of about 7.8 cases per 100 thousand people per year. Child stroke is considered heterogeneous in etiopathogenesis and clinical course, so the improvement of knowledge in the field of child stroke necessary to provide quality preventive and curative care.

Aims: Identify the frequency and structure of ischemic stroke in children, as well as the causes and clinical features and diagnosis of this pathology.

Results: In the period from 2012 to 2013, 34 children were hospitalized with a diagnosis of ischemic stroke in the Center of Pediatric emergency medicine Almaty. In 32% (11) of children aged stroke to 1 year of life. Complaints on admission were: decreased motor activity in 26 (76%) patients, weakness in 25 (74%), headache in 14 (41%), vomiting in 12 (35%). 10 (29.4%) patients underwent chickenpox, 9 (26.4%) diagnosed with a closed head injury in 14 patients (41.2%) hypertensive syndrome in 2 (5.9%) had an insect bite with allergic reaction. In the neurological status: in 27 (79.4%) patients with a clear conscience, 14 (41.2%) small-swinging nystagmus, 6 (17.7%) right-sided hemiparesis, 7 (20.6%) left-sided hemiparesis, 8 (23.5%) convulsions. All children performed CT scan of the brain: 65% (22) ischemic stroke diagnosed in the middle cerebral

artery in 15 (44.1%) patients diagnosed with calcifications in the basal ganglia.

Discussion: Thus, the most critical period for children of ischemic stroke is up to 1 year of life. The most frequent localization in the middle cerebral artery. Cause of strokes for the region act received a closed head injury and transferred chickenpox.

P441

PREDICTORS OF ARTERIAL ISCHEMIC STROKE IN PEDIATRIC PATIENTS WITH TUBERCULOUS MENINGITIS IN A TERTIARY GOVERNMENT HOSPITAL

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Objectives: To determine the incidence of stroke among patients diagnosed with tuberculous meningitis. To determine the factors predictive of stroke in pediatric patients with tuberculous meningitis. Subjects and

Methods: Patients (28 days to less than nineteen years of age) diagnosed with TB meningitis admitted in a tertiary government hospital from November 2012 to August 2013 were included. Patient's age, sex, duration of illness (TB meningitis), severity of illness (BMRC staging), clinical manifestations, presence of concomitant risk factors and medications given were obtained. Logistic regression analysis was done to determine the predictors of stroke for the study population.

Results: Thirty-five patients were included in the study. Eighteen patients (51.4%) had stroke prior to inclusion in the study, while 5 patients (14.2%) developed stroke after study enrolment. The mean age of patients with stroke is 7.1 ± 1.3 years with male:female ratio of 1:1.08. Statistical analysis revealed that duration of illness is the only significant predictor of stroke. Those who developed stroke are 17 times more likely to have been ill with TB meningitis for more than 7 days (odds ratio = 16.95, $p = 0.040$).

Conclusion: This study showed that a duration of illness of >7 days increases the risk of developing a stroke in pediatric patients with tuberculous meningitis. Therefore, early diagnosis and treatment is crucial to prevent the occurrence of stroke and improve the quality of life of patients with TB meningitis.

P442

PREVENTING STROKE IN SICKLE CELL DISEASE IN PORTUGAL

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Sickle cell disease (SCD) is a group of inherited haemoglobin disorders, of which the most common and severe is Sickle Cell anemia. It affects mainly children of African descent in whom it is the major cause of stroke, contributing to significant mortality and morbidity. The STOP Study (published in 2008) changed the standard practice Guidelines introducing the recommendation of systematic transcranial doppler (TCD) monitoring in order to stratify stroke risk and implement regular blood transfusions to prevent stroke. In a recent study, 590 patients were identified in Portugal, 62% are less than 25 years old and 530 (90%) live in the Lisbon area. About 100 children and adolescents with SCD are currently followed in our Institution. In order to improve the neurological care of these patients, a Pediatric Neurovascular Consultation was created in articulation with the Hematology Unit (where these patients are regularly followed) and the Neurosonology Unit. This organizational effort improved the communication between different specialties, increased the number of patients that systematically perform TCD and reduced the number of stroke events. Between 2009-2011, in the Neurosonology Unit, 97 children with SCD performed regular TCD and among them, six had increased stroke risk and only one suffered an ischemic stroke, after sudden interruption of regular blood transfusions when she immigrated to a different country. After permission, we translated and adapted the Sickle Cell and Stroke Leaflet (from The Stroke Association) that is given to parents and teachers and is a useful resource in educating caregivers regarding Stroke prevention.

P443**TRANSIENT CEREBRAL ARTERIOPATHY IN A CHILD ASSOCIATED WITH CYTOMEGALOVIRUS INFECTION**

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Introduction: Transient cerebral arteriopathy (TCA) is a recently described entity that is increasingly recognized as an important cause of arterial ischaemic stroke in children. Infectious agents associated with TCA include varicella-zoster virus, enterovirus, HIV and *Borrelia burgdorferi*. We report a patient with cytomegalovirus (CMV)-associated TCA.

Case Description & Methods: A previously healthy 30-month-old girl presented with acute onset of left hemiplegia. Brain CT, magnetic resonance imaging (MRI) and angiography (MRA) were arranged. A testing for viral infection was performed and included detection of viral material in the cerebrospinal fluid (CSF) using polymerase chain reaction (PCR) techniques and detection of antibodies (IgG and IgM) in early and late sera.

Results: Cranial MRI and MRA showed proximal stenosis of the right medial cerebral artery and ischemic lesions in the territory of this artery. Intriguingly, "puff-of smoke" network of vessels in the right basal ganglion are also depicted on MRA. Analysis of the CSF showed pleocytosis but normal chemistry profiles and negative bacterial culture. Positive CMV IgG and IgM and detection of CMV-DNA in CSF specimens by PCR suggested active CMV infection. Treatment with ganciclovir and anti-CMV immunoglobulin in addition to prednisolone mediation for 4 months resulted in gradual improvement of clinical symptoms. Intriguingly, the subsequent MRA revealed reversible vascular changes in the previously occluded cerebral artery after 6 months.

Conclusions: To our knowledge, this is the first report of a CMV infection associated with TCA in an immunocompetent child. Our report demonstrates the propensity for CMV to be involved in pediatric cerebral vascular disease.

P444**EPIDEMIOLOGY AND CHARACTERISTICS OF PEDIATRIC STROKE IN TAIWAN: A NATIONWIDE POPULATION BASED STUDY**

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Objectives: This study reported the prevalence and risk factors of pediatric hemorrhagic and ischemic stroke in Taiwan

Methods: We used the total data of Inpatient expenditures by admissions of 2011 of National Health Insurance to estimate the prevalence and incidence of stroke. In NHI database, patients aged less than 20 years old were enrolled. Cases of stroke were identified according to International Classification of Disease 9th edition (ICD-9) for outpatients. Statistic information on age, sex, disease status, and urbanization were used in this study. Risk factors were majorly according to the data of Registry for catastrophic illness patients and associated ICD-9 coding of the admission Claims of 2011.

Results: There were totally 5125,254 children enrolled and 394 children were diagnosed of stroke during the hospital admissions in 2011. The crude prevalence of stroke was 7.7/100,000. Hemorrhagic stroke were 44.4% and 37.6% for ischemic stroke. Boys had a higher prevalence than girls (8.1/100,000 vs. 6.7/100,000). Seizures and alter of consciousness were commonly commitment coding with pediatric strokes. 36 patients (9.1%) died during the admission and 130 patients (33.0%) received rehabilitation in 2011. Age < 2 has significant higher prevalence of stroke than other subgroups (P < 0.05). In addition, children lived in more rural areas were also at higher risk of stroke (OR = 1.31, 95% CI = 1.21-1.42).

Conclusion: Our findings suggest that age younger than 2 years has significant higher risk factor of stroke. Other Risk factors included Living in rural area, vascular disease, Previous diagnosed of cerebral palsy, congenital heart diseases, CNS infection, trauma, Brain tumor and hematologic malignancy.

P445**STROKE OUTCOME PROGNOSTICATION IN THE CRITICAL CARE SETTING**

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Introduction: Prognostication with unilateral large territory anterior circulation arterial ischaemic stroke (LAIS) in the critical care setting is challenging. Perceptions that active medical management is futile present a potential barrier to acute stroke therapies for such children.

Objective: To determine positive outcomes in children requiring critical care unit support following LAIS. To highlight limitations in using diffusion weighted imaging (DWI) findings for LAIS prognostication.

Methods: We report prospectively collected clinical and radiological findings of 5 children admitted Nov 2008-Jan 2010 to critical care following LAIS. Outcome data including video recordings and Paediatric Stroke Outcome measures at follow-up were obtained in all.

Results: Among 5 children with LAIS (2 male; median age 0.58 yrs, range 2 days – 6.33 yrs) arterial territories involved were middle cerebral artery alone (two), middle and anterior cerebral artery (one) and middle and posterior cerebral artery (one). Based on history and LAIS extent critical care staff withdrew active medical support (one), and proposed futility of active treatment or prognostication of profound neurological disability (four). All survived and at median of 8 years (1.0-2.9 years) follow-up. Both previously ambulant children and both neonates attained/regained ambulation by 18 months. The 7 month-old infant at stroke sat independently at two years.

Conclusion: Predicting outcome and decision making following large territory arterial ischaemic stroke in the critical care setting is challenging. We suggest caution in the interpretation of the significance and prognostic value of history and extent of DWI in this setting.

P446**TO REPORT A UNIQUE CASE OF BILATERAL MIDDLE CEREBRAL ARTERY INFARCTION ASSOCIATED WITH SILDENAFIL USE IN A CHILD.**

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Sildenafil is a phosphodiesterase 5 inhibitor that is widely used for the treatment of pulmonary hypertension in children. It is generally well tolerated and its safety data is largely derived from small uncontrolled trials, published reviews and case reports.

We report a case of twenty months old boy with Trisomy 21, who was admitted to our paediatric intensive care unit following a 'stiffening episode' at home followed by sudden onset of breathing difficulty. He had had been taking sildenafil for pulmonary hypertension secondary to bronchiolitis obliterans. During admission he required artificial mechanical ventilation for poor respiratory effort. CT scan of the brain showed extensive bilateral infarction in the middle cerebral artery territory with cerebral oedema and subuncal herniation. He was confirmed brainstem dead after 24 hours of admission. In view of the poor clinical condition and severe brain injury, intensive care was withdrawn after consulting family.

On literature search we found only one case report of a 52-year-old man who developed bilateral middle cerebral artery infarction soon after taking sildenafil and this entity has not been described in a child before to our knowledge.

Conclusion: Cerebral ischaemic event should be considered in a child on sildenafil who presents with altered neurological signs or symptoms. To our knowledge there is no case report of bilateral middle cerebral artery stroke associated with sildenafil in the paediatric population.

P447**RATIONALE AND DESIGN OF AN INTERNATIONAL MATERNAL NEWBORN STROKE REGISTRY :**

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Introduction: The incidence of peripartum maternal stroke is about 34 per 100,000 deliveries and the incidence of perinatal stroke is 1/2800 to 1/5000 live-births. Since pregnancy and delivery confer a highly significant increased risk for both maternal and infant strokes, there are likely to be as yet undiscovered interactions among maternal and infant characteristics in the pathogenesis of both stroke subtypes.

Methods: Women with peripartum stroke and their infants, and newborns with stroke and their mothers will be identified and enrolled retrospectively back as far as 2003 and prospectively at the time of the incident stroke. Control maternal-newborn pairs will be matched by age, race, and sex. All data will be entered into a web-based database. We will analyze predictors of stroke characteristics, risk factors and discharge outcome by using multivariate logistic regression.

Results: The pilot maternal newborn stroke registry at 4 international sites has formed the first collaborative network of pediatric and adult stroke neurologists, neonatologists, and obstetricians. Our progress includes developing the protocol and obtaining IRB approval, and building the data collection form and the REDcap registry module for web-based data entry. Enrollment is ongoing at 3 of the 4 sites.

Conclusion: The primary outcome of this international registry is to provide novel data on the risk factors and outcomes of maternal/newborn strokes. We will collect clinical and imaging data and blood samples for future DNA analysis and biomarker assays to further investigate the pathophysiology of maternal and newborn strokes.

P448**LEPTOMENINGEAL PHOSPHORYLATED ERK EXPRESSION AND URINE VASCULAR BIOMARKERS IN STURGE-WEBER SYNDROME**

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Introduction: Sturge-Weber syndrome is a neurocutaneous vascular malformation consisting of a facial capillary malformation, a choroid angioma of the eye and a leptomeningeal angioma. Affected patients develop seizures, hemiparesis, visual deficits, and cognitive impairments. We recently identified the somatic mosaic mutation in GNAQ which causes both Sturge-Weber syndrome and port-wine birthmarks.

Methods: Subjects (n=38) with Sturge-Weber syndrome and family member controls (n=30) provided urine samples and neurological scores at time 1, a year later and two years later. MMP, VEGF and bFGF levels were quantified in the urine samples. Immunohistochemistry was performed for phosphorylated ERK in cortical brain tissue samples from subjects with SWS and more affected areas compared to less affected areas.

Results: Urinary MMP2 and bFGF were more likely to be seen or to be abnormally high in the urine of subjects with SWS (21/33 versus 7/20, p=0.02; and 19/38 versus 7/29, p=0.015). Furthermore, MMP9 was higher in females than males (p=0.02); this was not seen in the family control data. MMP9 correlated with total neurological clinical score r=0.523, p=0.005 and with hemiparesis score r=0.399, p=0.03. Abnormal leptomeningeal vessels from more affected areas demonstrated increased expression of p-ERK in endothelial cells compared to expression in endothelial cells in less affected brain regions.

Conclusions: The hyperactivating somatic mutation in GNAQ, or alternatively blood stasis with resulting hypoxia-ischemia, may result in increased endothelial cell p-ERK expression in abnormal leptomeningeal vessels. Increased p-ERK expression may drive increased MMP and bFGF release and these angiogenesis factors may prove useful as clinical biomarkers.