even if no specific diagnosis is suspected. Unique statistical characteristics of LRs allow for a wide application in different clinical settings.

EFFECT OF DURATION OF STATUS CONVULSION ON NEURONAL APOPTOSIS AND EARLY APOPTOTIC EVENTS IN HIPPOCAMPUS OF RATS

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OBJECTIVE: Our goal was to explore the influence of duration of status epilepticus on neuronal apoptosis, mitochondrial membrane potential, and cytochrome c release in hippocampus in Wistar rats after status epilepticus (SE).
METHODS: SE that lasted for 30 minutes or 3 hours was induced by intraperitoneal injection with lithium chloride and pilocarpine. Rats were killed at different time points. The apoptosis, mitochondrial membrane potential, and intracellular cytochrome c level were investigated by flow cytometry.
RESULTS: The proportion of apoptotic cells, the decrease of mitochondrial membrane potential, and the release of intracellular cytochrome c significantly changed 30 minutes after 30-minute SE. The peak level was at the 12th hour after SE and 6th hour after SE in apoptosis and the 2 early apoptotic events, respectively. Compared with the same time point after 30-minute SE, the levels of apoptosis and the 2 early apoptotic events after 3-hour SE were much higher. The neuronal apoptosis and the 2 early apoptotic events in hippocampus after SE had a positive correlation with the duration of SE in partial correlation analysis.
CONCLUSIONS: Severe seizures could induce the changes of neuronal apoptosis and the early apoptotic events in hippocampus after SE; the longer the duration of SE, the more serious the change of apoptosis and early apoptotic events were.

PROTEOLIPID PROTEIN 1 GENE MUTATION IN CHINESE PATIENTS WITH PELIZAEUS-MERZBACHER DISEASE

Submitted by Yuwu Jiang
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INTRODUCTION: Pelizaeus-Merzbacher disease (PMD) is a rare X-linked recessive disorder that presents with nystagmus, impaired motor development, ataxia, and progressive spasticity.
OBJECTIVE: The objective of this study was to analyze the proteolipid protein 1 (*PLP1*) gene in 6 Chinese patients with PMD.
METHODS: Six unrelated Chinese patients had PMD (P1–P6), and 14 individuals were from family P2. Of these 6 patients, 3 had transitional, 2 had classical, and 1 had connate PMD according to the clinical and MRI features. Genomic DNA was extracted from peripheral blood samples. Gene dosage was determined by multiplex ligation-dependent probe amplification. All 7 exons and exon-intron boundaries of *PLP1* gene were amplified and analyzed by direct DNA sequencing.
RESULTS: *PLP1* duplications were identified in patients 1 through 4 with PMD. Their mothers were *PLP1* duplication carriers. Both duplication carriers and normal genotypes of *PLP1* were identified in the family members of patient P2. A c.517C → T (p. P173S) hemizygous missense mutation in exon 4 was found in patient 5, and his mother was a heterozygote of this mutation.
CONCLUSIONS: We identified 4 gene duplications and 1 missense mutation (p. P173S) of *PLP1* gene in 5 Chinese patients with PMD. This is the first report about *PLP1* mutations in patients with PMD in China.

IRON-DEFICIENCY ANEMIA IS ASSOCIATED WITH CEREBRAL SINOVENOUS THROMBOSIS: A CASE SERIES

Submitted by Fotini D. Kavadas
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INTRODUCTION: Iron-deficiency anemia is a relatively common but preventable condition in children that may have significant adverse implications on children's growth and development. Iron-deficiency anemia has also been sporadically reported in association with cerebral sinovenous thrombosis (CSVT).
OBJECTIVE: The objective of this study was to describe the largest case series to date of iron-deficiency anemia in association with CSVT as an advocacy measure for its prevention in children.
METHODS: Patients were identified through the Canadian Pediatric Ischemic Stroke Registry database (Toronto site). Included were patients who were older than 1 month to 18 years, met criteria for iron-deficiency anemia, and had radiographically confirmed CSVT.

RESULTS: Thirty-six patients who had CSVT and were older than 1 month to 18 years presented to the Hospital for Sick Children, Toronto, from January 2004 to December 2005. Six (17%) of these patients had iron-deficiency anemia; 2 were female adolescents, and 4 were male toddlers. All patients had historical and laboratory evidence of iron-deficiency anemia. Other risk factors, such as dehydration or inflammatory disorders, were present in all patients, but we did not detect significant inherited hypercoagulable disorders or acquired thrombophilia in any of them.
CONCLUSIONS: This report strengthens the evidence of an association between iron-deficiency anemia and CSVT. We suggest that children who have CSVT should be screened for iron deficiency. Advocacy to prevent iron deficiency is important, because this condition may have serious long-term and irreversible developmental consequences.

PREDICTION OF NEURODEVELOPMENTAL OUTCOME AFTER PERINATAL ASPHYXIA VIA TRANSCRANIAL CEREBRAL ARTERY DOPPLER ULTRASONOGRAPHY

Submitted by Haung-Chi Lin
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INTRODUCTION: Perinatal asphyxia is the most common cause of neurologic injury to the neonate and may lead to significant morbidity and mortality. Cerebral artery Doppler ultrasonography has been used for prediction of outcome after perinatal asphyxia in recent decades.
OBJECTIVE: We investigated whether cerebral artery Doppler ultrasonography is 1 of earliest markers of neurodevelopmental outcome.
METHODS: Cerebral artery Doppler ultrasonography was performed serially via temporal window during the first week of life in 24 asphyxiated neonates in our NICU from 1999 to 2007. The resistive indexes (RI) and blood flow velocities in the cerebral arteries of the circle of Willis were obtained. Neurodevelopmental outcomes were evaluated during follow-up.
RESULTS: Four patterns of cerebral artery Doppler ultrasonography were identified: pattern 1, normal RI with normal cerebral blood flow (6 neonates); pattern 2, normal RI with high cerebral blood flow (4 neonates); pattern 3, high RI after 48 hours of life (8 neonates); and pattern 4, low RI with high cerebral blood flow (6 neonates). Patterns 3 and 4 demonstrated grave prognosis, with severe cerebral palsy or mortality reported in more than 3 of 4 of neonates with either pattern, whereas all neonates with pattern 1 had favorable outcomes, free of delayed development or neurologic sequelae.
CONCLUSIONS: Cerebral artery Doppler ultrasonography is an early useful tool for predicting neurodevelopmental outcome after perinatal asphyxia. The outcomes turned out to be grave once abnormal RIs were detected, either low or high, indicating severe neurologic damage after asphyxia.

FOCAL AREAS OF HIGH-SIGNAL INTENSITY ON BRAIN T2-WEIGHTED MAGNETIC RESONANCE IMAGING SCANS ARE SIGNIFICANT FOR THE DIAGNOSIS OF NEUROFIBROMATOSIS VON RECKLINGHAUSEN TYPE 1

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INTRODUCTION: Neurofibromatosis von Recklinghausen type 1 (NF1) is characterized by the following National Institutes of Health (NIH) diagnostic criteria: café au lait spots, freckling, neurofibromas, Lisch nodules, optic glioma, distinct osseous lesions, and first-degree relative with NF1. Focal areas of high-signal intensity (FASI) in white matter and deep gray matter are typical brain MRI findings in children with NF1.
OBJECTIVE: This study evaluated the frequency of FASI and the possibility of using FASI as a diagnostic criterion.
METHODS: In a group of 160 children, the diagnosis of NF1 was confirmed in keeping with the NIH criteria. All children had MRI examination of the brain. The MRI findings of FASI in the children with NF1 were compared both with the brain MRI findings of the control group of 160 children with different diagnoses and with frequencies of the NIH diagnostic criteria.
RESULTS: In 137 (86%) patients with NF1, ≥1 FASI were found. The difference between frequency of FASI in the NF1 group and in the control group (14 [9%]) is highly significant. The frequencies of the diagnostic criteria were as follows: café au lait spots: 157 (98%); freckling: 123 (77%); neurofibromas: 112 (70%); NF1 relatives: 89 (56%); Lisch nodules: 71 (44%); optic glioma: 45 (28%); and osseous lesions: 15 (9%; only partial examination of the group).
CONCLUSIONS: The findings of FASI in T2-weighted images of the brain MRI are significantly frequent in children with NF1. Frequency of FASI is comparable with frequency of NIH diagnostic criteria. FASI could be proposed as an additional or new criterion for the NF1, mainly in childhood.

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