Conclusion In children and adolescents with T1DM DPN is highly prevalent, but in the majority of patients it is subclinical. Sensitivity and negative predictive values of the clinical neurological exam are low. Therefore, routine NCV measurement for the assessment of DPN appears warranted in these patients.

1513 EAST OR SESAME SYNDROME: A CASE SERIES

doi:10.1136/archdischild-2012-302724.1513

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Introduction Molecular and genetic advances have changed the way we look at associations of signs and symptoms or miscellaneous syndromes. A recent report has hypothesized that KCNJ10 mutations, affecting potassium channels present in the brain, ear and the kidneys, are responsible for the constellation of epilepsy, ataxia, sensorineural deafness. and tubulopathy (EAST or SeSAME syndrome). We present six patients belonging to three families with similar findings.
Case Series We describe three Asian siblings, two Caribbean siblings, and one Caucasian child who have epilepsy, ataxia, sensorineural hearing loss, and tubulopathy. Consanguinity was present only in the Asian family. Seizures were a presenting symptom in four of the cases with onset as early as 3–7 months of age. Development delay and learning difficulties were present in all of the cases. Ataxia was evident from early on. Sensorineural hearing loss was identified at different ages and in some cases was asymptomatic. In some cases, tubulopathy was an incidental finding. Two of the children were being followed up by nephrologists and neurologists before a unifying diagnosis was determined. Five children had previously been extensively investigated with metabolic and mitochondrial investigations, magnetic resonance images, and electroencephalograms all normal. All six children had biochemical evidence of a tubulopathy with hypokalaemia, hypomagnesaemia, and alkalosis. KCNJ10 DNA mutations have been identified in all the children.
Conclusion Recent advances in genetics have enabled us to determine the likely unifying cause for hitherto puzzling signs and symptoms in six children under our care.

1514 DEMOGRAPHIC, ETIOLOGICAL AND CLINICAL CHARACTERISTICS OF OUR CEREBRAL PALSY CASES

doi:10.1136/archdischild-2012-302724.1514

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Aim The aim of this study is to reveal demographic, etiological and clinical characteristics of our cerebral palsy cases and to underline differences peculiar to Thrace region of Turkey.
Methods One hundred and thirty five cerebral palsy cases, followed by Medical Faculty of Trakya University, Department of Pediatrics, Division Pediatric Neurology, were evaluated retrospectively. Data were obtained through out patient clinic files and patient discharge forms. Interviews by the phone were carried on in order to complete the necessary information. Demographic, etiological, laboratory and clinical characteristics of the cases were recorded.
Results The mean age of the cases was 112.65±47.02 months (2–18 years) and boy/girl ratio was 1.8. The majority of the case etiologies were perinatal risk factors accounted for 61.5%. Forty per cent of the cases were term AGA. Spastic type cerebral palsy constituted 91.9% of all cases with cerebral palsy while 46% of them were quadriplegic. Quadriplegic type was encountered most (46.2%) in term deliveries, while diplegic type was the most common form in preterms (47.4%). Speech problems (77.8%) and mental retardation (75.6%) were the most accompanying problems. Epilepsy accompanied 72.6% of the cases. An increasing rate of malnutrition was detected parallel to increasing age groups.
Conclusions Cerebral palsy cases showed certain differences in terms of demographic, etiological and clinical characteristics in Thrace region comparing to other regions.

1515 INFANTILE SPASM IN CHILDREN: CLINICAL FEATURES AND OUTCOME

doi:10.1136/archdischild-2012-302724.1515

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Objective Clinical features and outcome of children with infantile spasms.
Study Design Interventional and observational study.
Place and Duration of Study The Department of Neurology, Children's Hospital, Lahore, Pakistan, from January 2010 to December 2011.
Methodology Children aged <2 years presented with history of infantile spasms were assessed. Clinical presentation, EEG findings and response of anti-epileptic drugs was analyzed.
Results A total of 51,370 children visited Neurology outpatient department of Children Hospital, Lahore. Out of them, 450 infants had infantile spasms at their first presentation. Mean age at presentation was 6.6 + 2.5 months. Out of 450 children, 76% children presented at age < 6 month, 72% presented due to infantile spasms and 18% because of global developmental delay. Spasm types were mixed (38%), flexors (44%), extensor (16%) and asymmetric (2%). Symptomatic seizures were seen in 72% and cryptogenic in 28%. Hypsarrhythmia (67%) was the predominant EEG finding followed by modified hypsarrhythmia (24%) and other forms of epileptic discharges in 9% children. Majority of children were receiving oral Phenobarbitone, Carbamzaepine or Valproate sodium. We initiate the management with oral Prednisolone followed by Clonazepam or valproate acid. ACTH therapy was administered in only 5 children.
Conclusion Infantile spasms are one of the refractory epilepsy in children. Abnormal EEG findings predominantly the hypsarrhythmia or modified hypsarrhythmia are the hallmark. Majority of children received conventional AED with poor response. Oral prednisolone is proved to be the most effective AED. These children should be referred to the tertiary care paediatric neurology centers.

1516 ELECTROPHYSIOLOGICAL STUDY OF PERIPHERAL NERVES IN CHILDREN WITH ACUTE LYMPHOBLASTIC LEUKEMIA

doi:10.1136/archdischild-2012-302724.1516

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Introduction Acute lymphoblastic leukemia (ALL) is a malignant disorder of lymphoid progenitor cells, affects both children and adults. Chemotherapy induced peripheral neurotoxicity is an important side-effect of several chemotherapeutic agents.
The Aim of study was to detect the presence and types of peripheral neuropathy in newly diagnosed children with acute lymphoblastic leukemia, before and after the phase of induction of remission.
Patients and Methods 24 patients newly diagnosed as ALL were enrolled into the present study (group I); they were subjected to clinical and electrophysiological assessments which assess the integrity of the peripheral nervous system through conducting nerve conduction study and electromyography before and after receiving the chemotherapeutic agents. Ten apparently healthy children served as controls (group II).
Results 16 cases (67%) out of the 24 examined ALL patients were proved electrophysiologically to be free from PN, while 8 cases

(33%) were found to have PN and they were distributed as follows: 2 patients (8.3%) were proved to have chemotherapy induced PN, 4 patients (16.6%) showed subclinical chemotherapy induced PN and 2 patients (8.3%) showed subclinical PN due to ALL itself. 5 cases (62.5%) were found to have axonal pattern of PN, 1 case (12.5%) was found to have demyelinating pattern of PN and 2 cases (25%) were found to have mixed axonal demyelinating pattern of affection. Motor nerves affection was greater than sensory nerve.

Conclusion Chemotherapeutic agents used in phase I (induction of remission) proved to have a neurotoxic effects on peripheral nerves. In most of the patients, the peripheral neuropathy was subclinical.

1517 THE CLINICAL FEATURES AND OUTCOMES OF MOYAMOYA DISEASE IN A MEDICAL CENTER IN TAIWAN

doi:10.1136/archdischild-2012-302724.1517

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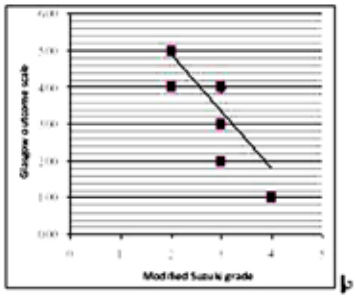
Background and Aims To describe the clinical features and outcomes of patients with moyamoya disease (MMD) who received surgical or medical treatment at a single institute in Taiwan.

Methods From August 2004 to September 2010, medical charts of patients with MMD (ICD-9 code: 437.5) from a medical institute in Taiwan were reviewed. Demographic and clinical characteristics, cerebral imaging files and follow-up information, and outcome were analyzed.

Results There were total 46 patients with MMD enrolled in this study. Male versus female ratio was 21 versus 25 (=1:1.2). Ages ranged from 1 to 84 year-old with the peak incidence in the 31–40 year age group (12 cases). The incidence of cerebral ischemic infarction was 75.0% (6/8) in the pediatric group, and 60.5% (23/38) in adult group; haemorrhagic stroke was 1.3% (1/8) in pediatric group and 26.3% (10/38) in adult group. Symptoms, included paralysis (76.1%; 35/46), consciousness change (34.8%; 16/46), headache (17.4%; 8/46), numbness (17.4%; 8/46), and seizure (17.4%; 8/46). A regression analysis showed that the outcome of patients with MMD was negatively related to a modified Suzuki's score ($p<0.05$).

Case	Sex/Age	Suzuki grade	Modified Suzuki grade	Outcome
1	F/84	6	4	1
2	F/64	3	3	2
3	M/49	3	3	3
4	F/43	3	3	4
5	M/34	3	3	4
6	F/42	3	3	4
7	F/40	3	3	4
8	M/21	3	3	4
9	M/36	2	2	4
10	M/34	2	2	5



Abstract 1517 Figure 1 Glasgow outcome scale v.s modified Suzuki's score

Abstract 1517 Table 1 Image modality and stroke type in a medical center

Image	
MRA	73.91% (34/46)
CTA	30.43% (14/46)
Angiography	21.74% (10/46)
Stroke type	
Ischemic type	63.04% (29/46)
Haemorrhagic type	23.91% (11/46)

Abstract 1517 Table 2 Surgical rate and Glasgow outcome scale

Surgical type	Percentage (N=46)
Neovascularisation	15.22% (7/46)
Removal of hematoma	19.57% (9/46)
None	65.22% (30/46)
Glasgow outcome scale	Percentage (N=40)
5	40% (16/40)
4	42.5% (17/40)
3	2.5% (3/40)
2	2.5% (1/40)
1	5% (2/40)

Conclusions MMD is commonly found in the Asian area, including Japan, Korea and Taiwan. However, the outcomes of patients with MMD are unpredictable. In this study, we found that the severity of MMD might be correlated with the scores of modified Suzuki's grading system. Therefore, the more the scores patients with MMD acquire, the higher risks of infarction will possibly occur in them.

1518 EPIDEMIOLOGICAL ASPECTS IN FEBRILE SEIZURES

doi:10.1136/archdischild-2012-302724.1518

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Background and Aims Febrile seizures are a common health problem in pediatric practice, knowledge of clinical manifestations and their evolution is important for correct therapeutic approach.

Assessment of clinical and evolutional features of febrile seizures (FS), with emphasis on risk factors such as age of onset, sex, FS category, familial history, etc., on the appearance of FS and on the risk of their recurrence is the aim of this work.

Method The authors conducted a retrospective study including 127 children aged 6 months to 5 years, hospitalized for FS between January 2008-March 2009 in our clinic. The role of risk factors in the development and recurrences of FS was analyzed.

Results FS appearance correlates with high fever (78%), male (54%), age 1–2 years (50%), but most do not associate familial history (20%), the global presence of at least one risk factor is found in 93% of cases.

Recurrences appear in 43% of cases of FS and 98% of cases correlate with the presence of at least one risk factor for recurrences (familial history 16% cases, onset of FS under the age of 1 year 29% cases, 47% complex FS).

Conclusions FS appearance correlates with high fever, male gender and age 1–2 years; appearance of relapses associates with complex FS, familial history and age under 1 year at onset.

1519 CLINICAL SPECTRUM OF CEREBRAL PALSY IN SOUTH JORDAN; ANALYSIS OF 122 CASES

doi:10.1136/archdischild-2012-302724.1519

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