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PROLONGED VIDEO-EEG NECESSARY TO IDENTIFY NON-CONVULSIVE STATUS EPILEPTICUS IN INFANTS ADMITTED TO ICUS

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Electrographic seizures are electrical events that can be detected by EEG but do not have clear clinical correlation. Despite the fact that these are found in over 40% of unconscious patients in intensive care units, many physicians are not aware of it. In the recent years it became clear that children and infants also suffer from it. Moreover, non-convulsive status epilepticus as a cause of decreased mental status is prevalent in children more than in adults. We present 2 cases of infants who were admitted to the NICU and PICU, were unconscious and their routine EEG did not show epileptic activity. Only prolonged Video EEG demonstrated non-convulsive status epilepticus and the treatment was changed accordingly.

Electrographic seizures in unconscious infants are prevalent. Though the long term consequences of these events are yet to be determined, in adults the delay in diagnosis and treatment is associated with poor prognosis. The closest entity in pediatrics that maybe used as a model is electrical status epilepticus in sleep. This is a pediatric disorder associated with cognitive and behavioral regression and carries a poor prognosis. It is unclear whether the seizures themselves are the offending cause but early diagnosis and treatment increase the probability of recovery.

A prolonged Video-EEG monitoring is a safe, available and reliable test to diagnose this. The literature and these two patients demonstrate the benefit of early use of the test and the immediate implications on the treatment.

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NEURO DEGENERATION IN CHILDREN: DEVELOPING COUNTRY PERSPECTIVES

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Back ground and aims: To find out the spectrum of neurodegenerative disorders of childhood, its diagnosis and role of paediatrician in the

management. It is a descriptive study conducted at Department of Neurology Children's Hospital, Lahore from June 1, 2005 to May 31, 2009.

Methods: A total of 2273 patients were admitted in the Neurology department with the diagnosis of DBD.

Results: Male to female ratio was 1.4:1. Age range was one to fifteen years. Metachromatic leukodystrophy was the predominant type (21%) followed by adrenoleukodystrophy (16%), SSPE (12%), Wilson Disease 10%, Alexander disease 3%, Hurler's disease 2% and Canavan's leukodystrophy in 1.5%.

Conclusion: Degenerative brain diseases are not uncommon entity in paediatric population. Commonest presentation is regression of milestones though it may be variable. Physicians must look into it when dealing with children having regression of milestones to diagnose them earlier. As in majority of patients there is yet no curative therapy available, so role of palliative care is very important. Collaboration should be established between the regional institutes to promote further research in future.

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CONGENITAL MYOTONIC DYSTROPHY: SMOOTH MUSCLE INVOLVEMENT OF THE URINARY TRACT IN A 2 YEAR OLD CHILD

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We report a child with Congenital Myotonic Dystrophy (CMD1) who presented at the age of 28 months with marked smooth muscle involvement of the urinary tract. Congenital Myotonic Dystrophy is a multi-system disorder affecting skeletal, smooth and cardiac muscles. Smooth muscle involvement predominantly affects the gastrointestinal tract. Involvement of the urogenital smooth muscles is extremely rare and has so far only reported in adult cases. There is little published on the effects of myotonia on the detrusor muscle.

The index case was a female infant who was the first child of non-consanguineous Caucasian parents. The antenatal ultrasound scan at 31 weeks showed evidence of polyhydramnios and moderate right renal pelvic dilatation. She was born at 33 weeks gestation by spontaneous vaginal delivery. Her mother had

clinical features of myotonic dystrophy. The baby was hypotonic requiring prolonged duration of ventilator support. The diagnosis of CDM1 in both mother and the baby was confirmed genetically.

A renal ultrasound on day 2 of life showed pelvicalyceal dilatation of the right kidney. At 28 months of age she had urinary tract infection. A renal ultrasound showed bilateral hydronephrosis with hydroureters. A micturating cystogram revealed dilated, tortuous ureters with marked vesicoureteric reflux bilaterally. The bladder was full, smooth and non-trabeculated. A Mercaptoacetyltriglycine (MAG3) scan showed bilaterally delayed and reduced excretion with a minimal response to diuretic on either side. Urodynamic studies done subsequently confirmed the diagnosis of myotonia of detrusor muscle. An MRI of the spine undertaken to rule out any spinal lesion was normal.

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WHEN SHOULD WE REQUEST NEUROIMAGING IN CHILDREN?

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Background: Effective communication between paediatricians and radiologists is crucial to allow timely, appropriate and useful diagnostic neuroimaging. Where its overuse may result in unnecessary radiation (e.g. CT scan) and waste of National Health Service resources, its underutilization may result in delay of potentially serious and life threatening disorders.

Aim: To evaluate the clinical basis for requesting neuroimaging in children and to investigate the impact of findings on clinical management.

Methods: A list of children aged ≤ 16 years, who underwent CT or MRI brain as requested by any specialty or department was obtained from the radiology department.

Results: The indication for neuroimaging was based on symptoms from history in 32 children, positive examination findings in 63 and outcome of preceding investigations in five. 54 children underwent CT and 46 MRI of the brain. Of 37 children who had no positive clinical findings on examination, only one showed an abnormality on scan which was deemed to be clinically insignificant. 73% scans revealed no abnormality. For these children with normal scan

results, the main benefit of imaging was judged to be the provision of reassurance only.

Conclusion: Nearly three quarters of all scans were normal and none of the children with normal clinical examination had clinically significant findings on brain scan. Our data demonstrates the need for careful consideration of the perceived benefits of neuroimaging, especially when there are no positive findings on examination as the risk for clinically significant abnormal neuroimaging result is appreciably lower in such cases.

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SOME CLINICO-LABORATORY STUDIES OF INTRACTABLE EPILEPSY

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Objectives: Is to detect some auto-antibodies that may be present in intractable epilepsy (IE).

Also the role of steroid as add on therapy in treatment and improving the quality of life of patients with IE.

Methods: Serum was obtained from sixty children with IE, and also from ten healthy children as control group. The above antibodies were estimated in both groups. Furthermore, the patients were divided into two groups: thirty of them were given steroid therapy for 18 wk in addition to their regular antiepileptic medication, the other 30 patients were given placebo drug for a similar period.

Results: There is an increased titer and significant difference in the levels of antiphospholipids in 43/60 patients (71%); ANA in 9/60 (15%); antidouble stranded DNA in 7/60 (11.7%) in comparison with controls. As regard anti cardiolipin IgM, was non significantly elevated in 20/60 (33.3%).

Thirteen children on prednisone therapy showed reduction in seizure frequency: 25% reduction in 11 children; 50% in one child; and only one patient with generalized tonic-clonic IE became entirely seizure free. A second serum sample was taken from the sixty children with IE. there was a significant reducing level of the studied auto-antibodies in those who received steroids in comparison to those who received placebo.

Conclusion: Intractability in epilepsy is multifactorial and the presence of auto-antibodies may be a