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.

657

WHEN SHOULD WE REQUEST NEUROIMAGING IN CHILDREN?

M. Ahmed, N. Dlamini

*Paediatrics, Burton Hospitals NHS Foundation
Trust, Burton Upon Trent, UK*

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.

658

SOME CLINICO-LABORATORY STUDIES OF INTRACTABLE EPILEPSY

A.A. Soliman¹, M.A.A. Abdo²

*¹Assiut University, ²Pediatrics, Assiut University,
Assiut, Egypt*

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

contributing factor. The use of prednisone could be an effective add on therapy in intractable epilepsy.

659

NEWBORN CONVULSIONS 2004-2009

M. Fejes¹, I. Velkey², T. Megyeri¹, J. Cservenyák², K. Váradi¹

¹Perinatal Intensive Centre, ²Childneurology, Borsod County Teaching Hospital, Miskolc, Hungary

The aim of the study was to introduce epidemiologic data of newborn convulsions and its outcome of our county's population.

Population and method: There were investigated 44 274 lifeborn newborns from 1st of January 2004 to 31st of December 2009 of Borsod County in Hungary. 47 of them had newborn convulsions (C). Therapeutic protocol was: 1st step: Diazepam; 2nd step: Loading phenobarbitone (20 mg/kg/day); 3rd step: Phenytoin (3x10 mg/kg/ 1st day) in some cases chloral-hydrate (25-75 mg/kg/doses maximum 2x/day) then the doses were decreased for maintenance doses.

Follow up examinations were done regularly 1-2 monthly. Statistical methods were mathematical mean, standard deviation and calculation of percentage.

Results: Characteristics of ill patients: gestational age 37,56±1,18 week, birth weight 3008,7±420,23 gram, Apgar 1' 8,19±1,13, Apgar 5' 9,1±0,88, mature/premature rate 36/11, male/female rate 24/23. The etiology was in a wide range: 13 asphyxia, 6 intrauterine infection, 5 familial, 4 patient 4th-5th day convulsions etc.

C incidence was 1,06/1000 lifeborn newborns. Status epilepticus (SE) was developed at 10 patients (inc.: 0,23/1000). 46,80% of all C was in the first five days. 60% of SE was observed on the 1st-2nd days. 3 babies died but not in the early period neither as a consequence of C and SE.

EEG abnormality was observed one third of C patients. Every case of SE had disorders of EEG. 11 epilepsies were observed, 7 of them had been transient, 2 turned into therapy resistant epilepsy.

Conclusion: There were less convulsions and status epilepticus than in the literature. Classical therapeutic protocol was useful in the most cases.

660

PREVENTABLE NEUROREGRESSION-CASE REPORT AND LITERATURE REVIEW OF VITAMIN B12 DEFICIENCY

D.K. Gandhi, R. Jayatunga

Paediatrics, Sandwell and West Birmingham Hospitals NHS Trust, Birmingham, UK

Background: Vitamin B12 deficiency in infancy is a cause of a preventable neuroregression and complex neurological manifestations.

Case report: An 18 month male infant was admitted from an OP Community Paediatrician's clinic for gross pallor. The parents reported concerns regarding the darkening of his skin, losing motor skills and apathy going back 3-4 months. He was still exclusively breast fed although weaning was attempted on various occasions. He had gross pale, puffy hands and feet, hyper-pigmentation, tremors, apathy, variable tone and was developmentally behind. Investigations revealed pancytopenia, low vitamin B12, normal ferritin and folate and hypersegmented neutrophils and grossly abnormal MRI. His mother was vegetarian and had low B12 yet normal haematology. He had a very stormy course during treatment, developed severe autonomic dysfunction with bradycardia, hypertension, drooling and unsafe swallow on initiating treatment and had to be transferred to PICU. He eventually demonstrated increasing attention and motor abilities and discharged with follow up in clinic and CDC.

Discussion: There are various reports in literature which describe the many different neurologic manifestations in deficiency and during treatment in an exclusively breast fed infant with maternal B12 deficiency although not all of these are described in a single case.

Conclusions: Infantile B12 deficiency from maternal deficiency can cause permanent neurological damage. There should be better awareness during the antenatal period and in the prime care to avoid neonatal and infantile B12 deficiency. Can selective neonatal screening prevent the catastrophe? Treatment can result in paradoxical deterioration and therefore needs close monitoring.