partly resistant. Causes of the resistance to therapy and possibilities of side-effects were investigated. A scoring method for the assessment of resistance to therapy in children with epilepsy is proposed.

PROGNOSTIC VALUE OF THE EEG IN PERINATAL HYPOXIC-ISCHAEMIC ENCEPHALOPATHY OF THE TERM INFANT

Poster

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Numerous studies have previously demonstrated that EEG recording is of definite value for evaluating the neurological outcome of term newborn infants with perinatal hypoxic and/or ischaemic encephalopathy (HIE). Newborn infants with very abnormal EEGs have a severe neurological evolution and, conversely, a normal EEG background activity in the first days is correlated with a favourable outcome. Many concerns remain for HIE infants whose EEGs display mild or moderate abnormalities. A multicentre retrospective study (1985 to 1991) including 239 term newborn infants with HIE was undertaken to evaluate the prognostic value of EEG in these 'intermediate' cases. According to Dreyfus and Monod's recommendations, the EEG was a multichannel—either continuous or serial—recording of long duration, performed during both the first 48 hours and the period between the third and the seventh day of age. EEG analysis focused on both the characteristics of the seizures pattern and the background activity, defined according to Pezzani's criteria. Careful neurological follow-up was performed by each centre at a minimum age of 12 months to evaluate the prevalence of subsequent developmental disabilities, including mental retardation, motor and sensorial deficits and epilepsy, among the survivors. Overall, the incidence of ictal discharges on EEG incidence was 36.5 per cent. Among this group, numerous discharges, the characteristic feature of status epilepticus, were observed in 72.4 per cent. Seizures appeared before 12 hours of age for 58.4 per cent of the infants and ictal discharges were recorded for more than 48 hours in 48.2 per cent. Background activity was categorised as severely abnormal (inactive or paroxysmal) in 24.3 per cent; moderately altered (continuous or discontinuous) in 20.1 per cent; or normal in 55.6 per cent.

The authors conclude the following: that status epilepticus is not systematically associated with a poor prognosis, since 18 per cent of the infants in this group had a subsequent normal outcome; that favourable neurological outcome was almost constantly observed (98 per cent) in the group exhibiting a normal EEG background activity before 48 hours of age; and that among the 'intermediate' group (42 infants), improvement of background activity with appearance of sleep organisation before seven days of age indicated a good prognosis (15/18), whereas persistent or worsened abnormal activity was strongly associated with a severe outcome (23/24).

WHEN TO DO A GENETIC-METABOLIC ASSESSMENT IN PATIENTS WITH A SUSPECTED MITOCHONDRIAL RESPIRATORY CHAIN DISORDER

Invited Lecture

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Exercise intolerance, (cardio-)myopathy, encephalopathy or a multisystem disorder in combination with lactic acidosis and abnormal findings in the muscle and/or nerve make it likely that the patient has a mitochondrial disorder. A positive family history provides strong support for such a diagnosis. Based on the clinical presentation and the availability of different tissues, a strategy can be devised for further laboratory investigations. Often analysis of mitochondrial DNA (mtDNA) isolated from white blood cells (WBC) is indicated as a first step in the laboratory diagnosis. Point mutations and deletions in WBC mtDNA can be analysed by PCR techniques. If these studies are negative, muscle biopsy is indicated. Material of this biopsy can be used for the analysis of the

metabolic pathways involved in ATP production, for activity measurements of the enzymes of the respiratory chain, for light and electron microscopy studies of muscle morphology, or for analysis of mtDNA. Storage of tissues, cell lines and/or DNA samples for future analysis and genetic counselling are also important. Enzyme analysis provides information about the function of the four complexes of the respiratory chain. The presence of ragged red fibres and COX-negative fibres in a muscle biopsy is suggestive of mitochondrial involvement. Ultrastructural analysis often shows structurally abnormal mitochondria with paracrystalline inclusions. As many mtDNA mutations are heteroplasmic in nature, it may be possible that mtDNA mutations are not present in all tissues. For most cases the PCR technique is sensitive enough to detect also a very small number of mutated mtDNA molecules. However, in case of deletions or depletions of the mtDNA, analysis of muscle tissues has to be recommended.

THE ONSET OF WALKING INDEPENDENTLY: QUALITY DIFFERENCES BETWEEN PRETERM AND TERM INFANTS

Oral Presentation

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There are marked individual differences in the ages at which children acquire the ability to walk without support. Children who are late in acquiring this ability have a higher risk for developmental disorders. Less attention has been paid to the quality of movement of first attempts at walking alone. Qualitative assessments of early walking may provide a sensitive means for detecting those infants with developmental impairments. The aim of this study was to devise a standardised instrument for the early walking in a free field situation. To control for differences in walking experience, all subjects were assessed 14 days after being able to walk five metres independently. The study group consisted of 52 children, of whom 33 were born preterm (further distinguished in terms of small (SGA) or appropriate (AGA) for gestational age) and 19 at term. Walking performance was judged to be optimal, near optimal, near poor or poor, based on a number of criteria including items on symmetrical co-ordination and tone regulation. The onset of walking was significantly later among preterm children, even after being corrected for gestational age. The optimal criteria were met by 12 preterm AGA infants and one preterm SGA infant. There were two term, one preterm AGA and five preterm SGA infants in the 'near poor' group. Eight preterm AGA and six preterm SGA but no term children showed poor walking quality. Those who were SGA were over-represented in the 'near poor' and 'poor' categories.

AUTOSOMAL RECESSIVE INHERITANCE IN LISSENCEPHALIC-LIKE DISORDER

Poster

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Lissencephaly is a known disturbance of neuronal migration leading to the absence or partial absence of gyral formation[1]. The authors describe a consanguineous family with a brother and a sister both having bilateral pachygyria and polymicrogyria. Detailed DNA analysis excluded a deletion on chromosome 17p. Until now the occurrence of isolated lissencephaly has been considered a sporadic event, and only a few families with more than one child affected have been found. The occurrence of consanguinity between parents of a patient with isolated lissencephaly was mentioned by Dobyns[1], who suggested the possibility of an autosomal recessive transmission of this trait. More family studies with adequate chromosome and DNA analysis will be needed to obtain a judgement about the risks of recurrence of isolated lissencephaly in one family.

Reference

1. Dobyns, W. B. (1987) 'Developmental aspects of lissencephaly and the lissencephaly syndromes.' *Birth Defects*, **23**, 225–241.