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INVESTIGATION OF CEREBROSPINAL FLUID DYNAMICS IN CHILDREN

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In order to diagnose any disturbance in cerebrospinal fluid (CSF) dynamics in children with macrocephaly and a normal growth rate of head circumference, we chose the lumbar CSF infusion method with a constant flow. By this method of investigation we obtained the most qualitative and quantitative data, giving information on CSF production, CSF absorption, the compliance of the CSF spaces and the pressure in the venous sinus. By relating the measuring data of the continuous infusion to the time function of CSF pressure by a computer model, a quick and accurate determination of the variables was possible.

We were able to confirm that the pressure-volume relationship can be expressed best as a mono-exponential function with a constant. We discuss the significance of the outflow resistance plus the advantage of the pressure-volume index, which together constitute the major parameters of the test.

In our patients with communicating hydrocephalus, CSF absorption was normal, whilst the Pressure Volume Index was elevated. As far as we have been able to ascertain, we are the first in the field to point to a positive correlation between ventricular size and pressure volume index in children with moderately enlarged CSF spaces.

Comparison of the lumbar CSF infusion with radionuclide cisternography revealed a wide distinction in the quantity of disorders. The difference between the results of the radionuclide cisternography and the lumbar CSF infusion can be explained by a different methodological approach. Whereas the absorption of the radionuclid is related to the CSF outflow as well as to the CSF volume, the lumbar CSF infusion depends exclusively on the CSF outflow.

We concluded that the lumbar CSF infusion with a constant flow is a relative simple investigation and the information on CSF dynamics, thus obtained, is easily comparable to that derived from other infusion techniques.

OPTOKINETIC NYSTAGMUS IN CEREBRAL VISUAL DISTURBANCES

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The examination of the optokinetic nystagmus (O.K.N.) has a long history in the Netherlands (Ter Braak and Van Vliet for one[1]). The examination concerned the binocular way of stimulation. Wood et al.[2] found a decrease of the nasotemporal following movement of the horizontal O.K.N. with regards to the temporonasal one in cats after surgical ablation of area 17, 18 and 19, suggesting a cortical loop of mediation of the nasotemporal movement. In 1975 Collewijn[3] and Hoffmann and Schoppmann[4] found, approximately at the same time, evidence of a subcortical route of mediation of the temporonasal component of the O.K.N. via the so-called Nucleus of the Optic Tract.

In 1983 we reported computertomographically demonstrated lesions of the optic tract and the striate area in children with cerebral visual disturbances, due to perinatal asphyxia[5].

We hereby report the first results of the optokinetic nystagmus in three of these children.

Monocularly testing, we demonstrated in two of them a decrease of the nasotemporal following movement, suggesting a lesion of a cortically mediated pathway.

In the third patient, an abundant latent nystagmus hampered the examination.

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CEREBRAL AND GENETIC ASPECTS OF ARTHROGRYPOSIS

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The preliminary results of a study of 55 cases of arthrogryposis (one or more congenital contractures, present at birth) are presented with special emphasis on cerebral and genetic aspects. Arthrogryposis may be caused by:
1) Muscle weakness due to cerebral, medullar or neuromuscular disorders leading to decreased fetal movements,
2) Mechanical restriction due to intra-uterine environmental factors such as oligohydramnios or
3) Prenatal primary connective tissue diseases.
In the subjects arthrogryposis originated from cerebral disorders (in 10 cases), medullar disorders (in 17 cases), congenital neuropathy (1 case), congenital myasthenia (1 case) and from myopathic disorders (5 cases). A primary connective tissue disorder was suspected in one case and oligohydramnios was present in 7 cases. In 13 cases it was not possible to reveal the underlying cause. Genetic investigations showed affected sibs in 8 cases, parental consanguinity in 8 cases, whereas in at least 14 cases hereditary conditions were present, indicating the importance of genetic counseling.

Of 45 cases of non-cerebral origin arthrogryposis, cerebral abnormalities were demonstrated in 21 cases, due either to perinatal complications or concomitant prenatal cerebral disorders.

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DISSOCIATED FOVEAL AND PARAFOVEAL VISUAL EVOKED RESPONSES IN SUBACUTE COMBINED DEGENERATION (SCD)

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In four patients with subacute combined degeneration clinically silent involvement of the visual pathways are demonstrated by VERs following selective foveal and parafoveal stimulation[1]. Whereas in MS patients optic nerve plaques are preferentially located in the macular region resulting in a delay of the small foveal fibres leaving the pattern-evoked response unchanged, the major abnormality in patients with SCD observed was a delay of the checkerboard pattern evoked responses contrasting with a normal or only slightly prolonged small-size square major positivity. During parenteral vitamin B_{12} treatment the latencies of the small-size square evoked responses remained constant or decreased to normal values. In contrast, recovery of the pattern reversal evoked responses was delayed and showed a persistant nerve conduction abnormality even at a 15 months follow-up examination. Thus the characteristic distribution of the lesion in patients with SCD is different from that generally seen in MS patients affecting slowly and fast conducting fibres inversely. It is suspected that impairment of conduction in the larger parafoveal fibres at an early stage of SCD may represent the pathogenetic abnormality responsible for the abnormalities of the VERs as was reported to result from paranodal demyelination in the peripheral nervous system in the rat[2] and in an experimental model of a giant axonal neuropathy produced by the toxin IDPN[3]. This model suggests that secondary paranodal demyelination is due to primary toxic axonal alterations, a mechanism that may well suit the pathogenesis of SCD.

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