INTENSIVE TEACHING AND THERAPY PROGRAMMES FOR DISABLED CHILDREN

Invited Lecture

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Education, therapy and health services for disabled young children in the UK are the responsibility of different statutory authorities, hence the co-ordination of the help that children require may be difficult. The problems of co-ordinating these services are compounded by the lack of data confirming the effects of therapy. In the regional Child Development Centre of Liverpool, UK, the authors elected to evaluate, monitor and treat the difficulties of a group of children who present with moderate impairments of motor, linguistic and cognitive functioning in a multidisciplinary setting. In addition to offering intensive teaching and therapy services, there was close liaison with parents and other community services. A minority of treated children now cope satisfactorily in mainstream school without additional assistance. Factors that determine the likelihood of this satisfactory outcome will be presented, together with an analysis of why most children, even with the availability of early and intensive help for what are considered to be moderate difficulties, nevertheless continue to show evidence of developmental disorder in the school years.

USEFULNESS OF THE ECHO-ENCEPHALOGRAPHIC VENTRICULAR–CRANIAL INDEX

Poster

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In the follow-up of high-risk infants in the neonatal intensive-care unit (NICU), transfontanellar echo-encephalography is the preferential imaging method to detect structural abnormalities and/or ventriculomegaly. To quantify changes in ventricular size, the ventricular–cranial index (VCI) (distance from the lateral wall of the body of the lateral ventricle to the falx) and the modified Evans ratio are widely used. In this study, the authors investigated the accuracy of these measures. Sets of seven coronary and sagittal echo-encephalographic images were used as a 'gold standard' for the conclusion: ventricular dilatation, change of ventricular size and ventricular asymmetry. The images were visually analysed by two experienced child neurologists. Finally, the VCI was measured from the coronal scan across the foramen of Monro. 55 infants in the NICU were chosen randomly in whom sequential (2 to 12) echo-encephalographic series of images were obtained. Using *t* tests, the VCI did not discriminate significantly between: (1) normal and dilated ventricles, (2) changing ventricular size, and (3) a/symmetry. The VCI is not a useful measure for ventriculomegaly, ventricular asymmetry or changes in ventricular size.

CHILDHOOD SPINAL MUSCULAR ATROPHY IN RUSSIAN POPULATIONS

Poster

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Spinal muscular atrophy (SMA) was investigated during the course of population studies of monogenic nervous and muscular disorders in six European Russian regions varying in geography and in genetic structure. Childhood-onset (before 18 years) SMA was discovered in all populations studied (total sample 26 patients/22 families; 16 males, 10 females, including a single case of distal SMA). In most of the populations, SMA was less common than childhood-onset motor and sensory neuropathy, and X-linked and limb-girdle muscular dystrophy. Prevalence of SMA types I to III was 1.45 to 2.50 per 100,000, differences insignificant; gene frequencies 0.0039 to 0.0057. The indices were compatible with data in other populations, though they may be underestimated due to 'loss' of undiagnosed SMA type I with early death. In the sample, SMA type II predominated. All families were compatible with autosomal recessive inheritance, though familial cases were