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psychiatric and social monitoring of input and outcome of the programme at Great Ormond Street Hospital will be mentioned.

ISCHAEMIC STROKE IN CHILDHOOD

Poster

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Thirteen patients with ischaemic stroke who were diagnosed between 1987 and 1994 were evaluated retrospectively. Their ages ranged between four months and 15 years; eight were female. Lipid profile was examined in all of them. Two patients had elevated triglyceride levels and two had elevated triglyceride and cholesterol levels. The causes of ischaemic stroke were as follows: moyamoya disease (two), type 1 diabetes mellitus plus hyperlipidaemia (one), subacute bacterial endocarditis (one), cavernous sinus thrombosis due to infection (one), hyperlipidaemia (one), trauma (two). In five patients the cause could not be determined. Since the patients with moyamoya disease have a high recurrence risk of stroke, they were given verapamil (4mg/kg/day), and the patient with subacute bacterial endocarditis was started on 100mg/day acetylsalicylic acid. Hemiparesis resolved in three patients; the others had sequelae of differing severity.

DYSTROPHIC MUSCLE CHANGES WITH GIANT LYSOSOMES AND VITAMIN E DEFICIENCY: A CASE REPORT

Poster

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Vitamin E is an antioxidant and protects unsaturated membrane lipids from oxidation by free radicals. Prolonged deficiency of vitamin E causes severe pathological changes in the gonads, heart, liver, blood, central nervous system and skeletal muscle in animals or humans. While in animals the skeletal muscle changes are primarily myopathic or dystrophic (necrosis and regeneration of muscle fibres), the condition in humans is less severe and frequently primarily neurogenic abnormalities in muscle may also be observed. In both conditions, the occurrence of abnormal lysosomal structures—also called giant lysosomes—is a prominent feature. In this report, the authors present the muscle biopsy data of a 19-year-old patient with congenital intrahepatic biliary hypoplasia. The biopsy was taken four years after the start of high-dose water-soluble vitamin E treatment (at age 15), which elevated his plasma vitamin E content (1µmol/L) to normal values (30µmol/L). From this time onwards, a continuous clinical improvement of his muscle complaints took place. The muscle biopsy showed dystrophic characteristics (severe increase of replacing fat cells, slight increase of endorsial collagen, variation in muscle fibre diameter, occasional fibres with internal nuclei and splitting, and some regenerating fibres) without actual necrosis. The most outstanding light- and electron-microscopic change was the presence of scattered so-called giant Lysosomes (membrane-bound structures consisting of mainly finely packed osmiophilic granules). Although many abnormal Lysosomes remain visible, the histological picture is not dominated by necrosis or by an ongoing neurogenic process.