

The best prospect of a permanent cure occurs in those cases seen in the first year of life and diagnosed early, where the tumour can be completely excised and is well differentiated.

Teratomas arising in other situations may be noticed at, or soon after, birth. This is particularly true of those arising in the thyroid, mediastinum, or stomach, and of the nasopharyngeal teratomas (epignathi). Teratomas of the testis or ovary, on the other hand, may present throughout childhood, and their histological structure is usually devoid of embryonic tissue.

**Factors affecting the form of medullocervical dislocation deformity in relation to meningo-myelocele and spina bifida.** J. L. Emery and N. MacKenzie. The Congenital Anomalies Research Unit, University of Sheffield, Thornbury Annexe, Sheffield 10.

Earlier studies indicated that with the Cleland-Arnold-Chiari deformity there is compression of the cranial segments of the cervical cord, a compression that is dispersed by the level of C8.

The present study is based on a complete dissection of 100 cadavers with meningo-myelocele and hydrocephalus. The lesions were first classified into degrees of deformity, and measurements of the deformities were recorded. The extent and type of the deformity was correlated with other anatomical features in the children.

The extent and degree of the medullocervical dislocation are not related to the site of the open meningo-myelocele, but are directly related to its extent. Evidence also suggests that the form of the deformity, i.e. the dorsal spur of 'knickung', and cyst are also related to the laxity of the cranial end of the dentate ligaments.

It would seem most likely that the medullocervical deformity in children with meningo-myelocele is secondary to the open spinal dysraphism.

**Uhl's anomaly.** W. A. Aherne. Department of Pathology, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne NE4 6BE.

The patient was born normally at term, of young and unrelated parents, after an uneventful pregnancy. She was admitted to hospital at the age of 7 weeks with an upper respiratory infection from which she made a good recovery. A chest x-ray at this time showed some enlargement of the heart shadow. She was admitted again at the age of 10 months for investigation of increasing general oedema, restlessness, and irritability. On examination the apex beat was not palpable; the heart rate was 125/minute; the blood pressure 110/70 mmHg. There was considerable enlargement of the liver, which was firm and acutely tender on palpation. X-ray of the chest showed gross enlargement of the heart, especially on the right side, and oligemic lung fields. There was incomplete right bundle-branch block. Cardiac catheterization showed (among other features) only a 3 mm pressure difference between the right atrial A-wave and the right ventricular systolic peak. Cineangiography showed a huge right atrium and ventricle with very poor contractions of the right

ventricle. She deteriorated and died in congestive heart failure.

At necropsy there was evidence generally of congestive heart failure, and, apart from this, the significant findings were in the heart and liver. The heart was greatly enlarged, due mainly to dilatation of the right ventricle and dilatation with hypertrophy of the right atrium. The free wall of the right ventricle appeared pale and fibrous except for a narrow zone, anteriorly and posteriorly, where it joined the interventricular septum. The cavity of the right ventricle was enormously enlarged, and though the anatomical configuration was normal, the papillary muscles in particular were extremely thin.

Sections taken from various parts of the right ventricle showed almost complete absence of myocardium in the greater part of the free wall; there were occasional small bundles of muscle fibres. There was a well-established endocardial fibroelastosis which was in direct contact with the epicardial and subepicardial tissues. At the junction of the right ventricular free wall and the septum there was a quantity of very small and only occasionally striated muscle fibres, which strongly resembled embryonic myocardium. There was no evidence of past myocarditis or of ischaemic damage; the appearances in general suggested rather a failure of myocardial development. Apart from the hypertrophy of the right atrial muscle, the rest of the heart appeared macroscopically and microscopically normal. It was concluded that propulsion of blood through the right heart was due almost entirely to atrial contractions.

Sections of the liver showed a cardiac cirrhosis, presumably due to the backward pulsations from the right atrium.

**Studies on GM2 type 2 gangliosidosis.** A. D. Bain. Department of Pathology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF.

**Necropsy diagnosis of fructosaemia in the newborn.** J. S. Wigglesworth. (Nuffield Neonatal Research Unit, Institute of Child Health, Hammersmith Hospital, Du Cane Road, London W.12.

**Lipid histochemical study in Fabry's disease.** B. Ivemark. Department of Pathology, Karolinska Sjukhuset, Stockholm 60, Sweden.

**Hirschsprung's disease: experience with some enzyme histochemical techniques.** J. D. Elema. Department of Pathology, University of Groningen, Oostersingel 63, Groningen, Netherlands.

Acetylcholinesterase (AChE)-positive nerve fibres are increased in number in the mucosal layer of the bowel of patients suffering from Hirschsprung's disease.

To decide whether rectal suction biopsies stained for AChE could be used in the diagnosis of this condition, 36 patients with constipation were investigated for AChE activity.

29 patients ultimately proved not to be suffering from Hirschsprung's disease; none of the rectal biopsies