Archives of Disease in Childhood, 1973, 48, 159.

Paediatric Pathology Society
Proceedings of the Seventeenth Annual Meeting

The Seventeenth Annual Meeting was held in Göteborg, Sweden on 15 and 16 October 1971.

Scientific communications
The survey was done in a rural area of nearly 300,000 inhabitants. In 6 years 768 cases of perinatal death were examined; 615 of these had a birthweight of 1000 g or more. The overall necropsy rate was 84%, rising to more than 95% in the last years of the survey.
The survey consisted of necropsy, pathological examination of the placenta, bacteriological and serological examination, collection of all available clinical data, and discussion of every case by the work group on perinatal mortality in Zeeland. In addition, a control group of 1076 cases and a high-risk group of 1282 cases were studied.
Some special aspects of this survey were the collection of all clinical data by a midwife, the accent on bacteriological examinations on a large scale, and the multidisciplinary assessment of every case.
Of the 615 babies with a weight of 1000 g or more, 38 died from infection. The relative frequency of infection was highest in the late neonatal deaths and in premature babies.
Five cases of generalized listeriosis were discussed in detail. In the same period 3 cases of listeriosis in living babies were diagnosed.

Child mortality in Malmö. Görel Ostberg. Department of Pathology, Malmö General Hospital, Malmö, Sweden.
Malmö is the third largest town in Sweden and has about 258,000 inhabitants. As there is only one hospital and the necropsy frequency is high, there is a good opportunity to do epidemiological work in different fields in the Department of Pathology of the hospital.
In this investigation all childhood deaths in Malmö during the years 1957 to 1970 were reviewed. Necropsy reports were available in 96% of the cases. Death certificate diagnoses were used for the remainder.
49,760 children were born alive, of whom 521 died during the first week of life. There were 546 stillbirths.
Perinatal mortality was 2.1%. Malformations were found in 19% of the stillbirths. Of the first week deaths 30% had hyaline membranes in the lungs, 10% had massive intraventricular haemorrhages, and 21% showed various types of malformations.

255 children died between the ages of 1 week and 1 year. 60% had major malformations. Of the malformations, 60% involved the heart and 25% the CNS. Abnormalities in more than one system were found in one-third of malformed children.
209 children were 1 to 14 years old at death, half of them under 4 years of age. 26% died after accidents, 25% of malignancy, including leukaemia. 19% had major malformations as the cause of death and 17% died of acute or chronic infection.

Value of polygenically determined animal defect in teratological research. C. L. Berry. Department of Morbid Anatomy, Guy's Hospital Medical School, St. Thomas's Street, London S.E.1.
The third molar tooth of the CBA strain of mice is absent in a variable percentage of animals. This 'absence' is determined polygenically and satisfies the criteria for multifactorial inheritance. In this it resembles the majority of malformations found in man, and exhibits threshold characteristics. Experimental systems permit the adjustment of this threshold, and both periods of inhibition of DNA synthesis and virus injections have been studied for their effects on the number of teeth missing. The pathogenesis of the defect has been clarified, and useful information concerning the value of such defects in the screening of potentially teratogenic agents has been considered.

A series of 107 teratomas in children has been reviewed. Most of the teratomas were sacroccygeal. The next most common sites of origin were the ovary and the testis. Other sites included the thyroid, mediastinum, stomach, and retroperitoneal tissue. There was a preponderance of females (70%). Of the 71 patients with sacroccygeal tumours, 57 presented in the first year of life. 51 tumours were excised completely and only 4 children subsequently developed recurrence. 15 were incompletely excised. 7 children in this group eventually showed recurrences. 3 tumours were inoperable and 2 patients died before operation.
Histological examination showed that 44 tumours were differentiated: only 2 of these children subsequently showed recurrences. 19 showed a mixture of differentiated and embryonic tissue: metastases ultimately occurred in 5 of these patients. 8 tumours were histologically malignant when first seen.
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.


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(myelocle 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 medulloccervical dislocation are not related to the site of the open meningo(myelocle, 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 medulloccervical deformity in children with meningo(myelocle 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 oligaemic 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 fructosaeinmia 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
Teratomas in children.

A E Claireaux and J W Keeling

Arch Dis Child 1973 48: 159-160
doi: 10.1136/adc.48.2.159-d

Updated information and services can be found at:
http://adc.bmj.com/content/48/2/159.4.citation

Email alerting service

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/