Fat-laden Macrophages in Cerebrospinal Fluid as an Indication of Brain Damage in Children. D. C. Chester, J. L. Emery, and S. R. Penny (Department of Pathology, The Children's Hospital, Western Bank, Sheffield 10). The occurrence of fat-laden cells in areas of degenerating brain is well known and such cells can escape into the cerebrospinal fluid. The appearance of these cells in stained smears from cerebrospinal fluid was described.

Over a period of one year, all cerebrospinal fluids cultured in the laboratories of the Sheffield Children's Hospital were examined for fat-laden cells. Differential counts were done on positive specimens.

Of 867 fluids examined, fat-laden cells were seen in 336, the majority showing only small numbers of these cells. Correlation of clinical information and laboratory findings suggested the following.

(a) When the cerebrospinal fluid contained less than 10% fat-laden cells, most of the children recovered with no obvious brain damage.

(b) When more than 30% of the cells in the cerebrospinal fluid contained fat droplets, most of the children died and survivors showed evidence of severe brain damage. When intermediate levels of fat-laden macrophages were found, the clinical picture was variable but most of the surviving children showed cerebral symptoms at a later stage.

Examination of cerebrospinal fluid for fat-laden cells is a simple, inexpensive procedure, and may have prognostic significance.

Pseudomonas aeruginosa Bronchopneumonia. A. J. Barson (University Department of Pathology, Williamson Building, Brunswick Street, Manchester 13). Published in Archives of Disease in Childhood, under the title 'Fatal Pseudomonas aeruginosa Bronchopneumonia in a Child's Hospital' (1971, 46, 55).

Is Respirator Lung a Distinct Syndrome? D. G. Fagan (Department of Pathology, The University, Dundee).

Insulin Secretion and Islet Cell Morphology of Human Fetal Pancreas. L. E. Olding (University of Uppsala, Dag Hammarskjolds vag. 17 Uppsala, Sweden).

Paediatric Pathology in the Children's Hospital, Saigon, 1969–70. D. A. Stanley (Royal Liverpool Children's Hospital, Myrtle Street, Liverpool 7).

Familial Dyschondroplasia with Visceral Involvement. A. H. Cameron (Department of Pathology, The Children's Hospital, Ladywood Middleway, Birmingham 16).

Granulomatous Disease with Acid-fast Bacilli. H. B. Marsden (Royal Manchester Children's Hospital, Pendlebury, Manchester M27 1HAO). The paper described two children of Indian stock born in the United Kingdom, a boy aged 3 years 10 months and his sister aged 2 years 6 months. The boy had a large left tonsillar swelling which did no trespand to treatment with PAS and INAH. Generalized lymphadenopathy and necrosis of tibia, femora, and the right clavicle developed together with pyrexia, high neutrophil leucocytosis, and a rash. Investigations for immunological abnormality and leucocyte function were negative. Gland biopsy from the neck showed fibrosis and plasma cell reaction with small polymorph foci. Culture of the gland yielded a branching acid-fast, as yet unidentified, bacillus sensitive to tetracycline and gentamicin.

The sister showed a similar picture of lymphadenopathy and rash without bone disease. Treatment with tetracycline produced a dramatic improvement in both children although glandular enlargement responding to gentamicin recurred in the boy after three months. Antibody was detected in high concentration to the acid-fast bacillus in both children by FA and agglutination of a formalized suspension. 15 controls including the parents were negative.

Squamous Epithelium in the Respiratory Tract of Children with Tracheo-oesophageal Fistula, and 'Retention Lung'. J. L. Emery and A. J. Haddadin (Department of Pathology, The Children's Hospital, Western Bank, Sheffield 10). Serial blocks from 35 children with tracheo-oesophageal fistula showed that 25 had extensive areas of squamous epithelium in the trachea.

The squamous change occurred principally in the muscular segment of the trachea but in a considerable number of children extended throughout the whole length of the trachea and into the bronchi and around the whole perimeter of the trachea.

A detailed survey of the cause of death in 50 children with tracheo-oesophageal fistula showed that many of the deaths previously ascribed to pneumonia were apparently due to the lack of ciliated epithelium in the bronchial air passages and the retention within the lungs of cellular debris and inhaled mucus. This appeared to be the major cause of death in children with isolated tracheo-oesophageal fistula.

The histological appearance of retention lung was discussed and it was pointed out that this change is nonspecific and possibly forms one of the facets of respirator lung. The condition is important to recognize clinically as the most rational treatment would appear to be pulmonary lavage.

Disseminated Ectopic Calcification in a Newborn Infant. F. A. Langley (Department of Pathology, St. Mary's Hospital for Women and Children, Whitworth Park, Manchester 13). This infant was born to a mother who was aged 20 and had been suffering from systemic lupus erythematosus for 3 years. Treatment by aspirin and chloroquine was stopped when she became...
pregnant. At about 8 months gestation she developed lupus nephritis and was treated with prednisolone and furosemide. She delivered a male infant spontaneously at 36 weeks.

The infant was ‘small-for-dates’, developed hypoglycaemia, and died on the fourth day. At necropsy extensive pulmonary haemorrhage was found. There was calcification of the fat in the neck, the intima of the pulmonary arteries, the aorta, and the circle of Willis. There was arborizing calcification of the kidney. There was hyperplasia of the islands of Langerhans of the pancreas and also of the adult cortex of the adrenal. The kidney showed fibrosis of the subcapsular zone, destruction of some tubules, and dilatation of others. Large foci of calcification were seen. The bones showed depressed osteogenesis.

Liver Damage after Treatment with THAM via Umbilical Catheter. J. D. Elema (Pathologisch-Anatomisch Laboratorium, Oostersingel 63, Groningen, Holland).

Symposium on the Early Detection of Phenylketonuria

The Organization of a Large-scale Guthrie Screening Programme. S. F. Cahalane (Department of Pathology, The Children’s Hospital, Temple Street, Dublin, Eire).

Early Detection of Phenylketonuria and Other Aminoacidopathies in a Large City Using Plasma Chromatography. D. N. Raine (Department of Biochemistry, The Children’s Hospital, Birmingham B16 BET). Capillary blood has been collected from infants, mainly at 6 to 9 days of age, born in the City of Birmingham (population 1 million; 20,000 births per year), for the past 18 months and the plasma subjected to paper chromatography as described by Scrivener, Davies, and Cullen (1964). Midwives take the samples to one of 12 collecting centres and all are delivered to the laboratory by midday. All subsequent work is done by one person whose only laboratory training has been in this procedure. The interpretation is checked by a biochemist.

Studies have established that the results on specimens posted to the laboratory, although different from those delivered immediately, can be interpreted satisfactorily. The optimum age for repeating initial tests which show only increased tyrosine has been established as six weeks, irrespective of whether the infant is premature or not. Administration of ascorbic acid has not shortened this period by a useful amount.

In 18 months the diagnoses that have been made include phenylketonuria 3, histidinaemia 3, prolinaemia 2, and hyperlipidaemia 1. There have been 14 infants showing hypermethioninaemia, mostly transient, but four required admission and one died with a possible diagnosis of ‘tyrosinosis’.

The initial test needed to be repeated in 6% of all cases. Three-quarters of the repeats were required because the initial pattern was abnormal, and 66% of the abnormal patterns were due to tyrosine. The remaining one-quarter were retested for technical reasons (haemolysis, specimen collected before 6 days, and laboratory mishaps).

Management of Hyperphenylalaninaemia (HPA) in Northern Ireland. Nina A. J. Carson (Nuffield Department of Child Health, The Queen’s University of Belfast, Grosvenor Road, Belfast BT12 6BJ). For a period of 1 year and 9 months, from 1 January 1969 to 30 September 1970, 56,257 newborn infants were screened in Northern Ireland for phenylketonuria (PKU) by the use of the Guthrie inhibition assay.

Specimens were collected on the 7th day of life by midwives, health visitors, or hospital staff and all tests were carried out in one centre in Belfast under the direction of the Consultant Bacteriologist of the Royal Victoria Hospital, Dr. George Gibson.

In all babies with phenylalanine levels of 4 mg/100 ml and above, repeat tests were requested and those with rising levels were investigated further. In these infants venous blood was examined for aminoacids by the use of an automatic aminoacid analyser, and a sample of urine examined for aminoacids and phenylketones. 17 infants showed levels persistently above 4 mg/ml. 8 infants were found to have classical PKU with phenylalanine blood levels in excess of 25 mg/100 ml, and phenylketones in the urine. A daily intake of phenylalanine of less than 300 mg was required to keep their blood phenylalanine levels between 2–8 mg/100 ml. 9 infants had persistently high blood levels varying from 12–23 mg/100 ml. On initial testing, urinary phenylketones were not found in this group. All were started on treatment, those with lower levels being given a low natural-protein diet; those with levels nearer 20 mg/100 ml, a hydrolysate mixture as in classical PKU.

Up to the time of writing, three of these children could tolerate a normal diet and the remaining six were still

Reference


Screening for Phenylketonuria on a District Hospital Scale. L. R. Davis (Department of Pathology, The Belgrave Hospital for Children, Clapham Road, London S.W.9). A report was presented on the experience gained in the first year’s work on screening infants born in the King’s College Group of Hospitals and the neighbouring Borough of Southwark. The reasons for undertaking small scale screening were given. Over the year 3500 Guthrie tests had been performed. The procedures for minimizing the clerical and technical work were described. Technical difficulties had been confined to allowing quality control of the results because of the variation in the amount of growth with different batches of medium and on different plates. Ways of allowing for this variation were suggested. Only two positive tests were discovered, but about 0.5% of tests gave results about the upper limit of normal and required repeating. No true cases of phenylketonuria were detected during the year.

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Disseminated ectopic calcification in a newborn infant.

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