The Fifth Annual Meeting of the Paediatric Pathology Club was held at Bristol on October 23 and 24, 1959. The meeting on October 23 was held at the Children's Hospital, St. Michael's Hill, when the chair was taken by Dr. Norman Brown. The meeting on October 24 was held at Southmead Hospital, when Dr. Frank Lewis was chairman. Dinner was taken at the Ashton Court Country Club. Forty-five members and guests attended the meeting.

Scientific Communications

Edward Hall (Liverpool). 'Primary Hyperoxaluria.' Eight cases of this condition occurring in three families were described and the histology illustrated. The familial nature of the condition was briefly discussed. (Further details of these families are to be published in this journal shortly.)

J. F. Boyd (Introduced by A. M. McDonnell, Glasgow). 'Adrenal Hypoplasia in Siblings.' The first child of healthy parents was admitted when 3 weeks old with a history of abdominal pain and vomiting. Intestinal obstruction was diagnosed and at laparotomy a volvulus of the small intestine was found and was undone successfully. Post-operatively vomiting continued; the patient's condition deteriorated and he died. Autopsy revealed hypoplastic adrenal glands.

The second child born four years later was also a boy and he was admitted when 9 days old with a history of persistent vomiting. No ante-mortem diagnosis was made, but the child was treated on the basis of an overwhelming infection with hydrocortisone and antibiotics. In spite of this his condition deteriorated and he died within 24 hours.

Autopsy revealed hypoplastic adrenal glands.

The paper described and compared the post-mortem and histological features in the two cases. The evidence suggested that in each case this was a primary failure of the adrenal glands rather than a pituitary failure.

Avinash Mithal (Introduced by John L. Emery, Sheffield). 'Postnatal Development of the Lung.' A study of the postnatal development of the terminal respiratory unit had been carried out which indicated that there is an increase in the number of alveoli following birth. There is a rapid increase in the production of alveoli during the first year after birth and a steady increase in the formation of alveoli throughout the whole of childhood.

Differential counts had been done on the respiratory tubes in the lung which suggested that there is an increase after birth in the number of small bronchioles and that slow progressive cartilaginization of these bronchioles occurs during later childhood.

P. O. Yates (Introduced by F. A. Langley, Manchester). 'Birth Injury to the Neck.' This paper was published in full in this journal (Archives of Disease in Childhood, 34, 436.)

Keith Rogers (Birmingham). 'Group F Haemolytic Streptococci as Pathogens in Children.' This group of streptococci is of importance in lesions connected with the whole gastro-intestinal tract, but the organisms demand a slight increase of CO₂ in the atmosphere in which they are grown, and a simple technical procedure was described to provide this.

Edith Faierman (Introduced by Hugh Cameron, Birmingham). 'The Significance of One Umbilical Artery.' Eleven cases of single umbilical artery were encountered among 411 routine autopsies on stillbirths and live born babies of 8 weeks of age or less (an incidence of 2.7%).

Severe associated malformations were found in 81% and in 23% of controls. The commonest malformation was of the lower urinary tract. There was a high incidence of stillbirths (45%) and of twinning (18%). In only one case was there an associated abnormality of the placenta. No correlation was found between the condition and maternal factors.

Absence of an umbilical artery can be diagnosed at birth by simple examination of the cord. In the absence of severe external malformations, such an infant has a 2:1 risk of internal malformations, which may require early surgical treatment.

G. S. Anderson (Newcastle). 'Pituitary Lesions Associated with Congenital Adrenal Hypoplasia.' Pituitary malformations in the absence of severe lethal malformations of the head are rare and comprise four published cases, all showing adrenal hypoplasia.

Three further cases were briefly described although one showed a cleft palate and other incidental abnormalities. The three infants died unexpectedly at 18, 23 and 33 hours and in all the adrenals were hypoplastic, resembling those in anencephaly. In one, a male, the pituitary grossly appeared absent but may have been ectopically placed in the sphenoid bone. The testes
were undescended and showed marked tubular and interstitial cell hypoplasia.

In the other two cases, both females, a small bean-shaped nodule of tissue was found in a small sella. Serial sections in both cases showed it to consist solely of pars anterior. In both cases there was a small rounded blunt knob of tissue in the region of the tuber cinereum which histologically was composed of malformed neurohypophyseal tissue anterior to which was a minute island of anterior lobe cells. In one case these cells apparently formed an attenuated connexion with the anterior lobe tissue in the sella.

It was suggested that the fundamental lesion in these cases was the pituitary malformation and that the adrenal hypoplasia was but one effect of this, although probably the most important.

H. B. Marsden (Manchester). 'Herpes Encephalitis following Fracture of the Skull.' A girl, aged 4 years, sustained a compound depressed fracture of the skull. There was excellent recovery for one week but on the eighth day pyrexia developed with convulsions and progressive coma. The child died 11 days later and at autopsy an inflammatory reaction was found in the brain involving the meninges and, in particular, the pons and basal ganglia. Perivascular cuffing and foci of inflammatory cells were prominent. An abundant growth of herpes virus in HeLa culture was obtained from the basal ganglia, the lung being sterile.

Evidence was produced to suggest that this was a case of herpes encephalitis, the virus probably entering at the site of the fracture.

The distribution and nature of the lesions were predominantly basal and cellular rather than destructive and unusual. Comment was made on the absence of Lipschütz bodies or type-A inclusions.

K. M. Laurence (Cardiff). 'The Pathology of Hydrocephalus.' The material seen between 1955 and 1958 at the Hospital for Sick Children, Great Ormond Street, London, was presented.

On the basis of the series, under-absorption was thought to be unproven, and over-production of C.S.F., seen in cases of choroid plexus papilloma, though interesting, was uncommon, while obstruction to the C.S.F. pathway was both pathologically and numerically the most important. Examples of aqueduct malformation were shown, most of which were found in association with other C.N.S. anomalies, notably spina bifida cystica and the Arnold-Chiari malformation. It was stressed that in cases of spina bifida cystica, the often associated hydrocephalus, although frequently due to malformation, was in many cases aggravated by, if not entirely resulting from, post-inflammatory lesions caused by ascending infection from the myeloeoe.

Aqueduct gliosis, although rare, presented problems in aetiology. Because of its association with neurofibromatosis, it was suggested that, like neurofibromatosis, it might be hamartomatous in origin.

The purely inflammatory lesions formed about half the series. The various pathological changes, such as aqueduct ependymitis leading to aqueduct block, and occlusive fibrosis of the arachnoid resulting in fourth ventricle exit foramen, or basal cistern block occurring singly or in combination, were illustrated by cases.

It was stressed that in over 60% of the group bleeding into the C.S.F. pathway at the time of delivery was regarded as the aetiological inflammatory agent, though in some cases little evidence of bleeding could be found at the time of the autopsy. (In discussion several members said that this series is a highly selected group and does not represent the hydrocephalus seen generally in the country.)

H. Urich (London). 'Some Remarks on the Neuronal Lipidoses.' All neuronal lipidoses share the characteristic feature of accumulation of lipids in the cytoplasm of nerve cells in addition to storage in other organs. Six diseases of this group were studied in Dr. R. M. Norman's laboratory in Bristol (see Table). Contrary to some of the older teaching based on superficial similarities, these diseases constitute separate entities distinguishable both histologically and by chemical analysis.

The conditions most commonly confused are Tay-Sachs' disease and Niemann-Pick's disease, despite the differences in the chemical composition of the brain in the two conditions. Examination of a case of Tay-Sachs' disease with exceptionally severe visceral involvement also revealed histological differences. The coarse vacuoles found in this case in reticulo-endothelial and glandular epithelial cells were apparently empty in formalin-fixed material, while in Niemann-Pick's disease the stored lipid could be easily stained with haematoxylin lakes.

Batten's disease (juvenile amaurotic idiocy) differs from Tay-Sachs' disease in the absence of an excess of ganglioside in the brain. The lipid stored in reticulo-endothelial cells of some cases is histochemically similar to that found in the neurones. It has not been identified chemically.

Gargoylism can be distinguished from the amaurotic idiocies by the involvement of connective tissues including the meninges and the adventitia of cerebral blood vessels.

In infantile Gaucher's disease only a few nerve cells show unequivocal signs of storage while others undergo degeneration without preceding storage. This phenomenon also occurs in other neuronal lipidoses and may be illustrated by the atrophy of the granular layer of the cerebellar cortex in Batten's disease.

The inclusion of metachromatic leuco-encephalopathy in this group is based on the finding of storage of sulphatide in numerous neurones of the basal ganglia, brain stem, spinal cord and posterior root ganglia. Severe demyelination, which is the most striking feature of this condition, may also be found in other neuronal lipidoses, particularly in cases of Tay-Sachs' disease with a protracted clinical course.

Neuronal storage, degeneration of nerve cells and loss of myelin sheaths may all be interpreted as results of the various disorders of lipid metabolism, presumably enzyme deficiencies, characteristic of these diseases.
R. M. Norman (Bristol). 'Intracerebral Calcifications.' The histological evidence pointing to a vascular factor in the pathogenesis of cerebral calcification was illustrated by reference to cases of the Sturge-Weber syndrome and of symmetrical cerebral calcification. In the former condition the calcifications tend to be localized in the outer layers of the cortex, that is, near the meningeal angiomatosis, but they are also seen in certain deeper laminae, notably the fourth layer of the calcarine cortex. In symmetrical cerebral calcification, whether associated with parathyroid deficiency or with familial microcephaly, the cortical calcifications have a predilection for the deeper parts of the gyral walls, especially in the boundary zones between major arterial territories. The fourth layer of the calcarine cortex may also be selectively affected. In the basal ganglia the localization of the calcifications is very similar to that found in status marmoratus following birth injury, though the lesions are otherwise dissimilar. The common factor appears to be a selective vulnerability of the capillaries of the affected areas.

The following short papers and cases were also presented:

Kenneth Rhaney (Dundee). 'Hypertrophy of the Choroid Plexus.' A full-term female foetus with gross hydrocephalus was delivered with difficulty after craniotomy. The trunk and limbs were normally developed and nothing of interest was found in the thoracic and abdominal viscera.

The head was greatly enlarged although the bones of the vault showed premature synostosis and the fontanelles were not enlarged. Reconstruction of the damaged brain showed that the septum lucidum, fornix and most of the corpus callosum were absent and that the lateral and third ventricles formed a single chamber. The lateral ventricles were greatly dilated; they were enclosed by a thin shell of cortex and atrophic white matter and their ependymal surface was coarsely nodular. Posterior to the genu the corpus callosum was replaced by a thin membrane which lined a large sac. This sac lay between the cerebral hemispheres posteriorly and formed part of the common ventricle.

The whole choroid plexus in the common ventricle formed a large tumour with a rough irregular surface. On histological examination it showed the classical structure of a simple choroid papilloma.

The brain stem and cerebellum showed no abnormality. The aqueduct was of normal size, and the foramina of the fourth ventricle were patent. The leptomeninges showed recent haemorrhage but no other macroscopic abnormality.

The papillomatous choroid plexus appeared to be responsible for ventricular dilatation. It may well have produced an excess of cerebrospinal fluid and also given rise to obstruction.

Leslie White (Manchester). 'Cushing Syndrome associated with Suprasellar Tumour in an Infant.' A male infant of 3 months presented with typical Cushing syndrome. There was sudden deterioration after 17 days with death. Autopsy revealed a large suprasellar tumour and bilateral adrenal hypoplasia with terminal thrombosis of the galenic veins. The tumour, which was continuous with a normal size pituitary gland, had an undifferentiated lobular epithelial structure. Some evidence of basophil cell origin was obtained.

A. M. McDonald (Glasgow). 'Krabbe Type Diffuse Cerebral Sclerosis.' A female, aged 9 months, was admitted the day before death severely ill with bronchopneumonia; she had been brownish-yellow at birth and for the following two months. At 7 months of age she had not sat up; she was a fat, mentally deficient child and her eyes did not follow light. Skull circumference was 43 cm., and she had a large anterior fontanelle.

At autopsy a Krabbe type of diffuse cerebral sclerosis, bronchopneumonia, gastric ulcers and oesophagitis were found.

A. M. McDonald (Glasgow). 'Congenital Absence of Biliary Ducts.' A premature male baby with complete harelip and cleft palate developed jaundice and white stools at the age of 7 weeks. Liver function tests suggested an obstructive type of jaundice.

A cholangiogram showed free passage of contrast medium into the duodenum through the common bile duct. The hepatic duct was faintly outlined and small in calibre.
At biopsy the liver was olive green. Histological examination showed that biliary ducts were either absent or in very small numbers. Biliary thrombi were also in small numbers. Early fibrosis was equivocal. The lesion was considered to be a bile duct atresia at the ductal level and it was thought that cirrhosis would follow.

The child is now 10 months old, is very well and not jaundiced. The cleft palate is about to be repaired.

E. HALL (Liverpool). 'Ganglioneuroblastoma. An Unusual Presentation.' A case of extra-adrenal abdominal ganglioneuroblastoma in a 24-year-old boy who presented with severe diarrhoea and some vomiting. There was a striking cessation of symptoms after removal of the tumour. Attention was drawn to a small number of similar cases recorded in the literature, and the possibility that a minority of these tumours may have internal secretory effects was discussed.

IAN DAWSON (London). 'Adenocarcinoma of the Testis in a Child of 11 months.' A Jewish child of 11 months developed a gradual, apparently painless swelling in the left testis over a period of two months. Both testes were present in the scrotum. A left orchidectomy was performed.

The tests measured $3 \times 2 \times 1.5$ cm. and appeared completely replaced by yellowish, rather gelatinous tissue. The epididymis appeared to be intact.

The histological appearances were uniform throughout all the blocks. The tumour consisted of irregularly arranged gland acini which varied in size from small solid elements to irregular spaces, into some of which papillary processes projected. The epithelium was in places cubical, in places low columnar. The acini were separated by loose oedematous stroma.

This tumour falls into the group of those described by Magner (1956) and may be called an adenocarcinoma of infant testis. The source of the growth remains uncertain but may be rete or junctional tubules. (Several members of the club had seen similar tumours.)

R. L. HOLMAN (Introduced by G. OSBORN). 'Persistence and Overgrowth of Immature Blasema in Both Kidneys of a Premature Infant.' A female infant born of 32 weeks gestation who survived 13 hours. Death was due to respiratory insufficiency; the lungs were only partially expanded and haemorrhagic.

Both kidneys were greatly enlarged; each weighed 70 g. and measured $7.0 \times 3.5 \times 3.5$ cm.; the cut surfaces showed replacement of normal structure by irregular cream and brown areas. The capsular surface showed a moderate degree of lobulation.

Microscopically both kidneys showed widespread persistence and overgrowth of immature blasema tissue interspersed and mingled with smaller zones of normally differentiated renal tissue. Immature blasema showed all gradations from undifferentiated deeply basophilic round cells through developing tubules and glomeruli to moderately well defined structures. A moderate number of mitoses were present in many such areas. There were many large straight tubules, probably all collecting tubules, extending radially from the medulla through the immature cortical zone almost to the capsule. Many of these were dilated, especially where they branched. Some collecting tubules ended in relation to zones of immature blasema tissue. Terminal collecting ducts were largely of normal appearance and lined by transitional cells distended with glycogen. No heterotopic tissues such as skeletal muscle and cartilage were found.

The diffuse nature of the anomaly in these kidneys suggests some generalized failure of organization of normal development rather than a localized overgrowth of metanephrinic blasema, such as occurs in a nephroblastoma.

HUGH CAMERON (Birmingham). 'Angioma of Larynx in Laryngeal Stridor.' This paper concerned three infants with severe persistent laryngeal stridor which did not respond to medical treatment or tracheotomy. All showed severe laryngo-tracheitis at autopsy and a small submucosal capillary haemangioma of the sub-glottis. These angiomas may be missed unless particularly careful examination is carried out and they appear to be more common in intractable laryngeal stridor than is usually recognized.

DOUGLAS Bain (Edinburgh). 'Congenital Leukaemia.' Two cases of congenital leukaemia were described, one of which was presented at birth with nodules in the skin. There was no history of mongolism or of radiation during pregnancy.

JOHN L. EMERY (Sheffield). 'Chronic Rectal Ulcer in Boy of 14.' A chronic indurated bleeding ulcer situated in the anterior wall of the rectum which was probably basically an angioma.

G. S. ANDERSON (Newcastle). 'Pinhole Perforation of the Foetal Colon.' A child with meconium peritonitis due to perforation of the colon, apparently due to a local defect in musculature.

A. M. MCDONALD (Glasgow). 'Umbilical Polyp.' An 11-month-old male infant presenting with an umbilical polyp. Since the cord separated a small black lump had been present in the umbilicus. Histology showed the features of a simple pigmented naevus, the colour being due to excessive melanin.