Collapse of the trachea may be suspected when the newborn child makes a series of respiratory movements without effect. Adequate treatment is, of course, to induce air flow into the trachea by pulling the tongue forwards, by mouth-to-mouth breathing, or by intubation.

A Case of Congenital Lymphoid Hyperplasia. W. L. DONOHUE (Toronto).

Polycystic Disease of the Liver andKidneys in Childhood. B. G. OCKENDEN and H. BLYTH (The Hospital for Sick Children, Great Ormond Street, London). A combined genetic and morbid anatomical study has been made of children with cystic malformation of both renal tubules and hepatic bile-ducts (polycystic disease of liver and kidney). 27 families have been studied in which the diagnosis has been confirmed histologically. Dominant inheritance is likely in 1 family and 6 others fall into this group on histological grounds. In 20 families an autosomal recessive pattern of inheritance is indicated. On clinico-pathological grounds the latter fall into four contiguous subgroups. Within individual families the type of disorder appears to breed true, therefore these subgroups are likely to be due to different recessive genes.

Relationship in Anencephaly between the Size of the Adrenal Gland and Length of Gestation. K. M. LAURENCE, A. B. N. ANDERSON, and A. C. TURNBULL (Welsh National School of Medicine, Cardiff). Evidence in the human fetus that the pituitary-adrenal axis is concerned in regulating the onset of labour may be obtained indirectly from observations in anencephaly.

A retrospective study was made of the 8 cases of anencephaly delivered at the Cardiff Maternity Hospital between 1959 and 1967, which were not complicated by hydramnios, and where the length of gestation was not in doubt and labour started spontaneously or was induced more than 4 weeks past term. In only one case did labour begin before term and in 5 of the remainder gestation was prolonged. In the 2 with the longest gestations, even to induce labour was difficult. Adrenal weights varied from 2·4 g. in the one with the shortest gestation, by even progression to 0·2 g. in the one with the longest gestation. The fetal (X) zone in the former was of almost normal dimensions and appearance. In the smaller glands there was a progressive diminution in the size of the zone. A histologically normal though small anterior pituitary gland was identified in all but one case; no posterior pituitary gland or hypophysis was found in any case.

A varying anterior pituitary stimulus is implied by the size of the adrenal, and it is suggested that a low pituitary-adrenal activity is associated with prolonged pregnancy and that the fetus to some extent determines the time of onset of labour.

Adrenal Hypoplasia during the Perinatal Period. C. B. F. DAAMEN (Rotterdam). In 88% of our 61 cases of anencephaly (2·9% of our material) there was coexistent hypoplasia of the adrenals, with more pronounced decrease of the fetal cortex.

Hypoplasia of the adrenals may be less pronounced in anencephaly before the 28th week of pregnancy.

In disorders of the whole primitive brain as well as disorders of the distal part of the primitive brain there exists a preponderance of females. In disorders of the anterior part of the primitive brain there exists a preponderance of males (Hamersma, 1966). In anencephaly postmaturity is frequent (18%). During birth mortality is high. Congenital hypoplasia of the adrenals without developmental disorders in the primitive brain was present in 2·5% of our perinatal mortality cases (2739 cases). In these the histological structure of the adrenals was similar to that found in anencephaly.

There was a preponderance of males and postmaturity. Advanced maternal age was frequent. All our 7 cases of 'isolated' adrenal hypoplasia died on the first day of life, and cyanosis was an obvious clinical feature.

References

Functional Activity of the Fetal and Neonatal Rat Pituitary and Adrenal Cortex. A. SCHABEER (Department of Pathology, University of Leiden, Netherlands). Making use of the organ culture technique, three groups of experiments were performed. Anterior pituitaries of 1-week-old rats were cultured for a period of 3 weeks and then transplanted to the sella turcica and under the renal capsule of 6-month-old hypophysectomized rats. At the time of transplantation the cultured explants had lost their specific functional activity, and consisted of completely degranulated cells. Five months after the transplantation the surviving animals were killed. Sections were made of the sites of transplantation. The corticotrophic activity of the transplanted cultures was assessed by measuring the difference in weight between the left adrenal, removed 10 days before the animal was killed and the right adrenal at the time of death. In 4 of the 5 surviving animals vascularized masses of well-differentiated chromophobes and chromophobe cells were found in the sella turcica. The implants under the capsule of the kidney consisted entirely of undifferentiated cells. 3 of the 4 animals with viable sella turcica implants showed compensatory hypertrophy; such a hypertrophy was not observed in rats with implants under the renal capsule. These experiments suggest that the anterior pituitary can only maintain its activity for a short time if there is no contact with the neural hypophysis and hypotalamic area. The activity is reinitated if the contact is restored. The adrenal cortex reacts to the reinitiated activity.

The fragments of the adrenal cortex of young rats were cultured in contact with fragments of the anterior pituitary of newborn and fet al rats. The amount of corticosteroids released over a period of 3 days was measured. The results showed that 19-day-old fetal rat pituitaries can produce corticotropic hormones.
Fragments of the anterior pituitary of young rats were cultured in contact with fragments of the adrenal cortex of newborn and fetal rats, and the corticosteroids present in the medium were estimated. The results of these experiments showed that the 17-day-old fetal rat adrenal can react to corticotropic hormones.

Effects of Chromosomal Anomalies on Fetal Development. D. I. Rushton (Department of Pathology, University of Birmingham).


Solitary Rhabdomyoma of the Heart. J. Hakosalo, O. Rasanen, and F. Steenback (Oulu, Finland). The patient, a boy, had slight postnatal asphyxia, but made normal progress until at 7 months of age there was a 7-day episode of convulsions. At that time the first chest x-rays and ECCs were taken. Left ventricular enlargement with an abnormal, egg-shaped, cardiac contour was seen. The ECC showed left bundle-branch block, which was a constant finding for the rest of the patient’s life. Physical examination of the heart, and physical and mental development were normal. At the age of 4-7 years the boy contracted pneumonia, followed by convulsions and respiratory arrest. Some days later he died, after a cardiac arrest. At necropsy no tuberous sclerosis or other malformations were found. In the lateral wall of the left ventricle, at the apex of the heart, a spherical, pale, spongy, and sharply bordered tumour was found, measuring 3 × 3 × 4 cm. The histological sections presented a typical picture of a rhabdomyoma of the heart. In this case three unusual features of the disease were evident: the tumour was single, the disease was not associated with tuberous sclerosis, and the life span of the patient was quite long. A triad of an abnormal ECC, an abnormal cardiac contour, and convulsions should always arouse suspicions of a heart tumour.

Three Cases of Testicular Adenocarcinoma of Infancy. T. E. Parry (Cardiff). The following cases were reported. Case 1. A soft grey translucent tumour was removed from the scrotum of a 14-month-old boy. It had been present for 11 months. A recurrence was excised 3 months later. The child died at the age of 19 months; at necropsy the pelvis was filled with greyish translucent brain-like tissue which extended into the abdomen. There were extensive peritoneal metastases, and small secondary nodules were present in the left lung. Case 2. A 5-year-old boy was admitted in April 1960 with enlargement of the left testis which had been noticed for 6 weeks. He was otherwise well. A testicular tumour measuring 5 × 4 × 3-5 cm. and consisting of soft friable greyish material was removed. Recovery was uneventful. In June 1960 he was readmitted with acute appendicitis and an acutely inflamed appendix was removed. No evidence of metastasis was seen. He was alive and well 6 years later. Case 3. A boy aged 2-5 years presented with a swelling of the left testis in August 1967. Apart from some anaemia (Hb 9-6 g./100 ml.), nothing abnormal was found on clinical examination and the chest x-ray was normal. The tumour was excised. Examination under anaesthesia 2 months later was normal. He remained well until Christmas 1967, when he complained of pain in the abdomen and legs, and began to lose weight. By January 1968 a large mass was present in the left hypochondrium. Treatment with 'provera'—a progesterone compound—was given, but the patient died one month later. At necropsy a large soft retroperitoneal mass was found overlying the left kidney. On section the tumour presented a greyish translucent appearance resembling neonat al brain. Secondary deposits were present in the para-aortic glands and in both lungs.

All three tumours presented similar microscopical appearances. Each showed an adenocarcinomatous pattern, with well-marked tubular and glandular areas as well as solid sheets of undifferentiated cells, many of which were vacuolated. The cells were rich in glycogen, mucin was present in many of the acinar spaces, and fat stains were weakly positive. The macroscopical features of soft homogeneous greyish translucent brain-like tissue appear to be sufficiently characteristic to suggest the correct diagnosis.

Case 1 was included—as their Case 4—in the series reported by Teoh, Steward, and Willis (1960).

REFERENCES


Lymphangioma of Bone. H. B. Marsden (Royal Manchester Children’s Hospital, Manchester).

Blast Cell Proliferation in Children with Untreated Acute Lymphoid Leukaemia. H. P. Wagner (Berne).

A Case of Dys-ß-globulinaemia. D. I. K. Evans (Booth Hall Children’s Hospital, Manchester). A girl now aged 10 years first developed atopic eczema at 3 months of age. She had had recurrent infections including three attacks of generalized herpes simplex, four attacks of pneumonia, and five attacks of pneumococcal meningitis. ß-globulin treatment was ineffectual.

The tonsils and lymph nodes were small. Circulating lymphocytes averaged 1000/cu. mm. Platelets were normal. Bone-marrow lymphocytes and plasma cells were normal. Immunoglobulins showed high IgG, high IgA, and low IgM values (e.g. IgG 3900, IgA 400 and IgM 27 mg./100 ml. serum). Anti-A titre 1/16, anti-B titre 1/8. There was no response to 0-1 mg. DNCB after a 1 mg. sensitizing dose, and no delayed hypersensitivity to candida antigen.

Two injections of TAB produced antibody titres to S. typhi H of 1/250 and S. paratyphi B H of 1/125,