

Paediatric Pathology Society

Proceedings of the Fifteenth Annual Meeting

The Fifteenth Annual Meeting was held in Newcastle on 24 and 25 October 1969, at the Post-Graduate Medical Centre, Newcastle General Hospital, and the Royal Victoria Infirmary.

The President of the Meeting was Dr. W. Aherne, and the chair at the scientific sessions was taken by Drs. W. Aherne, T. Bird, and C. B. F. Daamen.

The annual dinner was held on 24 October, when the principal guests of the Society were Professor and Mrs. A. G. Heppleston, Dr. and Mrs. G. W. Pearce, Dr. G. Dale, Mr. and Mrs. J. E. S. Scott, and Dr. and Mrs. B. Tomlinson.

Nineteen papers were given, and there were 9 demonstrations. Some 45 member and guests attended the scientific programme.

The next meeting of the Society will be held in Belfast on 16 and 17 October 1970, when Dr. J. E. Morison will be the President.

Scientific Communications

Germ Cells and Genital Ridges. D. I. Rushton (*Department of Pathology, University of Birmingham*). The lack of knowledge of the interrelationships between germ cells and genital ridge cells is indicated. The knowledge at present available is applied to the problem of gonadal dysgenesis, in particular Turner's syndrome and pure gonadal dysgenesis, and to dysgenetic gonadoblastoma. It is suggested that the factors responsible for the aberrant lines of development probably act at about the end of the first trimester and reflect disturbances in the function of the genital ridge cells. The importance of two functional sex chromosomes in the germ cells is uncertain but may be significant. The need to study the gonads of early embryos and fetuses, particularly where the karyotype is known, is emphasized.

Enzyme Histochemistry of the Developing Ovary and Endometrium. J. Pryse-Davies (*Queen Charlotte's Hospital, Goldhawk Road, London W.6*). The ovaries and fundus uteri were obtained from 11 fetuses, 11 stillbirths, 16 neonatal deaths, and 5 infants: 8 enzymes were studied.

In the developing ovary, alkaline phosphatase was present in pregranulosa and germ cells, absent from superficial epithelium and primordial follicles, but reappeared in Graafian follicles. Leucine aminopeptidase was only active in theca interna cells. Acid phosphatase, succinate and glucose-6-phosphate dehydrogenases and NAD diaphorase were found in all epithelial structures. The dehydrogenases and NAD diaphorase

showed marked theca interna activity, while acid phosphatase appeared heaviest in granulosa during antrum formation. Absence of dehydrogenase and NAD diaphorase in scattered primordial follicles probably corresponded to atresia. The steroid- 3β -ol dehydrogenase method proved unreliable, due to control staining in the absence of substrate.

The phosphatases and monoamine oxidase showed selectively heavier activity in developing endometrial glands of stillbirths and early neonatal deaths, resembling the findings in adult secretory endometrium. These enzyme patterns were also seen in a 26-week abortion following hydroxyprogesterone administration to the mother, and were associated with endometrial vacuolation and menstruation praecox, but were not found after the neonatal period. Leucine aminopeptidase showed slight activity in superficial stroma in cases with marked secretory changes. The dehydrogenases and NAD diaphorase showed non-selective endometrial activity at all stages, providing an index of necropsy tissue viability.

Leucocyte Hydrolytic Enzymes in the Diagnosis of Some Neurological Disorders. D. N. Raine (*Biochemistry Department, The Children's Hospital, Ladywood Road, Birmingham 16*). The chemistry and metabolic relationships of gangliosides and the related globosides and sulphatides have recently been well enough defined to allow rational discussion (Raine, 1969a). The main value to pathology is that ill-understood variants of amaurotic family idiocy can now be recognized as separate nosological entities. Moreover chemical techniques, though specialized, are available for the precise diagnosis of a particular patient suffering from GM1 gangliosidosis (generalized gangliosidosis), GM2 gangliosidosis (infantile Tay-Sachs disease), GM3 gangliosidosis, globoside storage disease, metachromatic leucodystrophy, and some others.

A number of inherited metabolic diseases can be diagnosed by demonstrating the associated enzyme deficiency in leucocytes. This has been done for metachromatic leucodystrophy (Langelaan, 1969) and for GM1 gangliosidosis and for globoside storage disease (Raine, 1969b).

Clinical, chemical, and enzymic studies were reported on cases of metachromatic leucodystrophy, GM1 gangliosidosis, and globoside storage disease.

REFERENCES

- Langelaan, D. E. (1969). Leucocyte arylsulphatase in metachromatic leucodystrophy. *Biochemical Journal*, **112**, 26P.
Raine, D. N. (1969a). Biochemical classification of the sphingolipidoses. In *Some Inherited Disorders of Brain and Muscle*, p. 89. Ed. by J. D. Allan and D. N. Raine. Livingstone, Edinburgh.
Raine, D. N. (1969b). Tay, Sachs *et al.* *Lancet*, **2**, 959.