

REVIEWS

EMBRYOLOGY

By M. B. L. Craigmyle and R. Presley

Bailliere Tindall 1975 2nd edition 271 pp. £ 2.50

This is the second edition of a small book in the series of Concise Medical Textbooks published by Bailliere Tindall London. The backcover claims that "this extensively revised and re-illustrated edition now contains all the embryology required for 2nd M.B. and 2nd B.D.S. examinations". Anatomists who have been brought up on the larger tomes of yesteryear may wish to challenge that claim but with the increased curtailment of time allotted to purely morphological studies this book will fill a need of preclinical medical and dental students preparing for their respective examinations.

A glance at the contents shows that of 15 chapters, two thirds are devoted to organ and tissue development. This is consistent with the vocational objective of the book. Developmental anomalies are clearly and fearlessly emphasized *in situ* and not relegated to a separate chapter or in small print at the end of a chapter as an afterthought. To give an example from the chapter on the circulatory system, the description of atrial septal and related defects follows logically the account of its normal development and precedes the description of the formation of the ventricular septum and the anomalies that

may arise therefrom. Such an arrangement emphasises the need to understand normal development in order to appreciate its aberrations.

Notwithstanding, it is refreshing to see appended lists of suggested further reading at the end of the chapters, but the omission of references after the chapter on the lymphatic system is puzzling, especially since the ontogeny of immunological competence is such a current topic. Many of the references point the student to original sources. It is to be hoped that these will be referred to by the more curious and scholarly so that the pursuit of embryology may be seen not only as a prelude to the study of abnormal development but also as a no mean science to be cultivated in its own right.

Apart from small typographical errors, particularly in the index, the book as a whole has been well written and can be recommended to preclinical medical and dental students.

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INHERITED BIOCHEMICAL DISORDERS AND URIC ACID METABOLISM

By David S. Newcombe, M.D.

HM + M Publishers, Aylesbury, England, 1975.

Price: £ 12.

Described in 1964 the Lesch Nyhan Syndrome has probably done more to foster research on purine metabolism over the last decade than gout itself. The time is ripe for a book like Newcombe's.

Within 36 succinct chapters, remarkably up-to-date, he covers 30 inheritable diseases associated mainly with raised serum acid levels. They range from the common (e.g. diabetes mellitus, familial hyperlipoproteinaemias) to the rare (e.g. hepatic fructose 1,6-diphosphatase deficiency, hyperammonaemia). The hypouricaemic conditions are the diseases associated with the Fanconi Syndrome, xanthinuria and the newly described syndrome of Sperline (1974) in which hypouricaemia occurs with hypercalciuria and decreased bone density. This is the kind of critical, discursive work that one would expect of the Director of the Rheumatology Unit at the University of Vermont College of Medicine.

The pathogenesis of hyperuricaemia in a large

number of the diseases stem from some degree of organic aciduria which presumably act via the renal tubular transport system for uric acid. Thus the resistance to probenecid of the hyperuricaemias of glyco-gen storage diseases and alcoholic ingestion arise in most instances from increased blood lactic acid concentrations. Allopurinol, however, is effective in lowering these uric acid levels as it inhibits the biosynthesis of uric acid but does not alter its renal handling.

When one comes to gout biochemical heterogeneity has replaced any simple unitary hypothesis. There are experimental data to incriminate any of the following: (a) a partial deficiency of hypoxanthine-guanine phosphoribosyltransferase, (b) altered sensitivity of purine feedback mechanisms, (c) increased phosphoribosylpyrophosphate synthetase activity, (d) defective glutamine metabolism augmenting (c) and (e) (believe it or not) increased purine

catabolism in a patient! In one vital area, the association of hyperuricaemia with lipid disorders, theories are hard to come by because of unresolved questions on the relationship of purine and lipid metabolism.

Entrance into the rarefied atmosphere of purine nucleotide research would have been made somewhat easier if there were clear accurate diagrams. Here I regret having to point out some errors in an otherwise delightful volume.

Figs. 1 and 2 are apt to mislead as the enzymes, represented by numbers, are not always next to the arrows indicating the reactions they catalyse. Between Fig. 4 and Table VII the switch from ordinary to Roman numerals should not have been allowed. "Dihydroxyacetone" of Fig. 5 should be "dihydroxyacetone phosphate". Fig. 7 is misleading in that carbamyl phosphate formed for urea synthesis is mitochondrial in location and is separate from

the cytoplasmic pool which caters for pyrimidine synthesis. More discussion could have been given to the synthesis of phosphoribosylpyrophosphate (PRPP) in an introductory chapter as this is a key intermediate in purine regulatory pathways.

Since the review was written the following paper adds weight to the argument that an increase in PRPP synthetase activity is the cause of the abnormally high cellular PRPP content and possibly of the purine overproduction described in the Lesch Nyhan Syndrome. "Phosphoribosylpyrophosphate overproduction, a new metabolic abnormality in the Lesch Nyhan Syndrome. G. H. Reem (1975) *Science*. 190, 1098—1099."

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"MOTOR DEVELOPMENT IN THE DIFFERENT TYPES OF CEREBRAL PALSY"

By Bobath, B and K. William Heinemann

London 1975

£1.85

This little book by the Bobaths is the result of 20 years of experience dealing with cerebral palsied children, distilled and compacted within 100 small pages. Their thesis is that cerebral palsied children cannot be assessed simply by comparing with children with normal development, because each type of cerebral palsy changes the whole tempo of development peculiar to its own type of lesion. A totally wrong assessment will result if this is not taken into consideration. They first describe normal development, and on a background of this pattern, they then describe the altered developmental patterns of the different forms of cerebral palsy. This is illustrated by 67 photographs for clarity.

This small book can be recommended wholeheartedly to all paediatricians especially those taking care of cerebral palsied children, to medical students as well as other professional personnel such as physiotherapists, social workers, etc.

It can also be recommended to parents of children with cerebral palsy, as the descriptions and explanations are in simple language. In these days of escalating prices for medical books, £1.85 for the price of this book is an excellent investment.

Professor Wong Hock Boon

Gout is an inherited disorder of purine metabolism that causes hyperuricemia in humans, particularly men. The term "gout" in general use refers to a form of arthritis. Typically, elevated blood uric acid levels (serum-urate levels >7 mg/dl in men and > 6 mg/dl in women) cause monosodium urate crystals to deposit in joints, bones, and subcutaneous tissues. Inflammatory reactions to these crystals give rise to severe recurrent bouts of acute arthritis. Tophaceous gout is a disorder of purine metabolism or renal excretion of uric acid. Monosodium urate precipitates, leaving deposits (tophi) throughout the body. One of the more common sites of gouty tophi is the helix of the ear. In this location, tophi are nonpainful, firm nodules. Gout may occur as an inherited or an acquired disease.

Inheritance. Disorders of amino acid metabolism. Urea cycle defects. Amino acid transport disorders. Organic acidemias. Disorders of carbohydrate metabolism. Galactose and fructose disorders. Glycogen storage disorders. Congenital disorders of glycosylation. The metabolism of the carbohydrates galactose, fructose, and glucose is intricately linked through interactions between different enzymatic pathways, and disorders that affect these pathways may have symptoms ranging from mild to severe or even life-threatening. Clinical features include various combinations of hypoglycemia (low blood sugar), liver enlargement, and muscle pain. Most of these disorders can be treated, or at least controlled, with specific dietary interventions. Galactose and fructose disorders.