(Zellweger) syndrome and neonatal adrenoleukodystrophy. Similarities in phenotype and accumulation of very long-chain fatty acids. *Johns Hopkins Med. J.* 151 (1982) 344–351

Hanson, R. F., Isenberg, J. N., Williams, J. C., Hachey, D.,
Szczepanik, P., Klein, P. D. and Sharp, H. L. The metabolism of 3α,7α,12α-trihydroxy-5β-cholestan-26-oic acid in two siblings with cholestasis due to intrahepatic bile duct abnormalities. J. Clin. Invest. 56 (1975) 577–587

Hanson, R. F., Szczepanik-Van Leeuwen, P., Williams, G. C., Grabowski, G. and Sharp, H. L. Defects of bile acid synthesis in Zellweger's syndrome. *Science* 203 (1979) 1107–1108

Kase, F., Bjorkhem, I. and Pedersen, J. I. Formation of cholic acid from 3α,7α,12α-trihydroxy-5β-cholestan-26-oic acid by rat liver peroxisomes. *J. Lipid Res.* 24 (1983) 1560–1567

Mathis, R. K., Watkins, J. B., Szczepanik-Van Leeuwen, P. and Lott, I. T. Liver in the cerebro-hepato-renal syndrome: defective bile acid synthesis and abnormal mitochondria. *Gastroenterology* 79 (1980) 1311-1317

Monnens, L., Bakkeren, J., Parmentier, G., Janssen, G., van Haelst, U., Trijbels, F. and Eyssen, H. Disturbances in bile acid metabolism of infants with the Zellweger syndrome. *Eur. J. Paediatr.* 133 (1980) 31–35

Oftebro, H., Bjorkhem, I., Skrede, S., Schreiner, A. and Pedersen, J. I. Cerebrotendinous xanthomatosis. A defect in mitochondrial 26-hydroxylation required for normal biosynthesis of cholic acid. J. Clin. Invest. 65 (1980) 1418-1430

Poulos, A., Pollard, A. C., Mitchell, J. D., Wise, G. and Mortimer, G. Patterns of Refsum's disease. Arch. Dis. Child. 59 (1984) 222–229

Poulos, A. and Sharp, P. Plasma and skin fibroblast C₂₆ fatty acids in infantile Refsum's disease. *Neurology* (1984) (In press)

Salen, G. and Shefer, S. Bile acid synthesis. *Ann. Rev. Physiol.* 45 (1983) 679–685

Scotto, J. M., Hadchouel, M., Odievre, M., Laudat, M. H., Saudubray, J. M., Dulac, O., Beucler, I. and Beaune, P. Infantile phytanic acid storage disease, a possible variant of Refsum's disease: Three cases, including ultrastructural studies of the liver. *J. Inher. Metab. Dis.* 5 (1982) 83–90

Trijbels, J. M., Monnens, L. A., Bakkeren, J. A., Van Raay-Selten, A. H. and Corstiaensen, J. M. Biochemical studies in the cerebro-hepato-renal syndrome of Zellweger: a disturbance in the metabolism of pipecolic acid. *J. Inher. Metab. Dis.* 2 (1979) 39-43

Whiting, M. J. and Watts, J. McK. Prediction of the bile acid composition of bile from serum bile acid analysis during gallstone dissolution therapy. *Gastroenterology* 78 (1980) 220–225

Book Review

Congenital Adrenal Hyperplasia, M. I. New and L. S. Levine, 1984 (Monographs in endocrinology 26), Springer-Verlag, Berlin, DM72, US\$ 26.90.

Congenital adrenal hyperplasia is relatively common with an incidence in newborns of about 1 in 10 000, but general awareness of this situation is recent. It is therefore timely that this monograph should appear. This book is about clinical endocrinology but necessarily involves steroid biochemistry as its basis. The different biological activities of many steroids determine that the chemistry and biology are inseparable. This close linkage has the advantage that clinical findings can direct one with great accuracy to a diagnosis and to its relevant investigation. The knowledge available in this monograph is therefore necessary to all concerned in the clinical investigation and treatment of these patients.

The contents of the monograph are: Chapter 1 Steroidogenesis and enzymatic conversions of adrenal steroid hormones, Chapter 2 Fetal sexual development, Chapter 3 Enzyme defects, Chapter 4 Recent advances: the fasiculata and glomerulosa as distinct glands, Chapter 5 Treatment, Chapter 6 Pubertal development, Chapter 7 Genetics, Chapter 8 Prenatal diagnosis. There is a short subject index and a 16-page list of selected references.

The contents therefore include a useful summary of recent findings on the genetic linkage with the HLA complex. The consideration of heterozygote detection is consistent with evidence in other areas like galactosaemia that individuals can carry two expressed mutant alleles to give a spectrum of changes ranging from gross disease to

apparent normality. It should also be noted that isoenzymes under independent genetic control are well recognized. In other words some greater breadth of view might have strengthened some of the arguments developed in a detailed consideration of this group's work. There is justifiably only one page on prenatal diagnosis. Since it was mentioned that low maternal oestrogen levels in blood and urine could indicate cholesterol desmolase deficiency, it should also be pointed out to readers using this book for differential diagnosis that similar low levels might be expected in 17α hydroxylase and 17-20 lyase deficiencies. Although Maria New must be given credit for her pioneering work on screening for this disease greater caution on the clinical benefits of screening might have been derived from experience in parallel areas. The accumulated experience of screening for this defect is due to be reviewed in 1985 at Liverpool in a Workshop attached to the SSIEM meeting and it may be inadvisable to anticipate the conclusions of such a review. Those with the 1983 edition of Stanbury and colleagues, Metabolic Basis of Inherited Disease, and with some other publications already have similar information from the same author(s).

In summary, this is a good monograph which achieves useful clarity on a common condition. It should be excellent for clinical endocrinologists in training or those requiring updating. Its appearance is timely since we are in the research and development phase of screening for this condition.