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Quantitation of small intestinal biopsies from children. J. W. Keeling and R. A. Risdon. The Hospital for Sick Children, London.

Small intestinal biopsies from children have been analysed quantitatively to obtain indices reflecting both the mucosal volume and surface-to-volume ratio.

In subjectively normal biopsies, correlation of the patients's ages with the surface-to-volume ratios showed the latter to be significantly reduced in younger children and some overlap occurred between these lower values and the range seen in biopsies showing 'partial villous atrophy'. However, in the abnormal biopsies additional mucosal abnormalities were invariably present. It is suggested, therefore, that slight changes in the villous pattern of biopsies from children under 2 years of age should be interpreted with caution and should not be regarded as necessarily pathological in the absence of other mucosal abnormalities. 'Flat' biopsies from children with untreated coeliac disease invariably showed smaller surface-to-volume ratios than controls, but the mucosal volumes of these biopsies were significantly increased. Specimens showing less marked abnormalities ('partial villous atrophy') had surface-to-volume ratios in an intermediate range, but, like the biopsies from untreated coeliacs, mucosal volumes were significantly increased. The most useful application of quantitative analysis is likely to be in following accurately the sequential changes in biopsies from the same patient. We have therefore applied this technique to serial biopsies obtained from a number of children suspected on clinical grounds of having coeliac disease, but in whom this diagnosis had not been confirmed histologically. Biopsies were obtained while the patients were receiving a gluten-free diet and also after a 'gluten challenge'.

Case of hyperammonaemia due to ornithine transcarbamylase deficiency. C. Morley and I. B. Sardharwalla. Willink Biochemical Genetics Laboratory, Royal Manchester Children's Hospital, Pendlebury, Manchester.

A 5-year-old female with normal mental and physical development presented with a 3-month history of episodic vomiting, lethargy, and nocturnal confusion, each lasting for about 48 hours. Spontaneous recovery occurred each time. In the final attack she became ataxic, confused, and lapsed into coma, which led to her admission. Examination revealed that she was deeply unconscious, pyrexial, and hypertonic with hepatomegaly. Investigations showed high blood and CSF ammonia levels ($>1000 \mu\text{g}/100 \text{ ml}$). In addition, the

urine contained increased amounts of pyrimidine derivatives, namely orotic acid, uridine, and uracil. Plasma and urine glutamine concentrations were raised. In an attempt to lower blood and CSF ammonia, peritoneal dialysis was carried out with standard dialysis solution and the ammonia levels returned to almost normal within 48 hours. The effectiveness of dialysis was shown by high levels of ammonia in the dialysate. Unfortunately the child died of cerebral oedema. Enzyme assay in the liver obtained at necropsy within 6 hours of death showed marked deficiency of ornithine transcarbamylase which was 3% of normal at pH 7. The other urea cycle enzymes were normal.

Osteodystrophy associated with prolonged cholestasis. J. F. T. Glasgow and P. S. Thomas. Institute of Clinical Science, Belfast.

Metabolic bone disease was studied in 9 infants (birthweight 1.02–3.53 kg) during the course of prolonged obstructive jaundice beginning in early infancy. 5 had proven biliary atresia and in 2 others cholestasis complicated severe rhesus isoimmunization. The occurrence of bone disease at 2 to 4 months of age was accompanied in only 3 patients by features suggesting this diagnosis, and jaundice was diminishing in those who did not have biliary atresia. Indeed, 2 patients were no longer clinically icteric. At the time of diagnosis each patient had a marked rise in serum alkaline phosphatase concentration and aminoaciduria. Serum calcium and phosphorus values were more variable, the former being closely related to the gestational age of the baby at birth. X-rays in the majority showed classical rickets with bone age retardation. One infant, however, had in addition to rickets of the scapula, severe demineralization of the axial skeleton and multiple rib fractures. The appendicular skeleton was normal. Rapid skeletal healing took place in those whose cholestasis subsided. In biliary atresia distinctly abnormal bones remained and one child of 13 months developed multiple long bone fractures. Various pathogeneses might be contributory to the osteodystrophy, including a shortened gestation.

Effect of B17-V inhalation on hypothalamic-pituitary-adrenal axis and on growth in asthmatic children. M. Friedman and J. Frears. University College Hospital, London.

The effect of the administration of 1600 μg daily of B17-V by aerosol was studied in 15 adult volunteers over a 7-day period. The daily plasma cortisol levels taken at

9 a.m. before and after the administration of B17-V showed no change. The responses to physiological and pharmacological doses of ACTH were within the normal range. After insulin-induced hypoglycaemia, 7 of the 9 subjects studied had a normal maximum level. A total of 53 severely asthmatic children from our clinic have been treated with B17-V by aerosol. There appears to be no suppression of their hypothalamic-pituitary-adrenal axis, as judged by morning plasma cortisol levels, response to insulin-induced hypoglycaemia, and response to pharmacological doses of ACTH. The growth velocity of 24 children treated with B17-V aerosol for longer than a year was found to be normal. Growth hormone secretion after insulin-induced hypoglycaemia was normal in 14 children who had received therapy for longer than 6 months. These findings indicate that there is no significant suppression of the hypothalamic-pituitary-adrenal axis after short- or long-term administration of B17-V under the conditions described, and no observed effect on growth.

Assessment of venous admixture in infants with congenital heart disease using arterial blood gas tensions. J. B. Owen-Thomas, F. Meade, and R. S. Jones. Department of Child Health, University of Liverpool.

Noninvasive method of continuous measurement of gas exchange in spontaneously breathing infants in a neutral thermal environment. F. Meade and J. B. Owen-Thomas. Department of Child Health, University of Liverpool.

Conducting tissue in common ventricle. J. L. Wilkinson, R. Arnold, R. H. Anderson, and R. S. Jones. Royal Liverpool Children's Hospital, Liverpool.

Studies of the normal development of the conducting tissue in the heart have shown that the atrial and ventricular components develop separately. The main bundle and its branches develop *in situ* within the developing ventricles and the atrioventricular node develops from sinus venosus and atrial tissue, there being a complete ring of specialized tissue in the embryo around the atrioventricular valves. In addition to the atrioventricular node itself, small areas of ring tissue may persist and make accessory connexions with the ventricles (e.g. Kent's bundle). In malformed hearts the atrioventricular node may fail to make a connexion in the normal site and abnormal connexions may then form the sole pathway. Conduction via an abnormal anterior route has been shown in congenitally corrected transposition and in single ventricle with outlet chamber. The present study involved 6 univentricular hearts, without outlet chambers. Heart block developed in 2 of 4 cases operated on. The conducting pathways are extremely variable and are difficult to predict from the morphology, which is also inconstant. In one case (the only one to have sinus rhythm postoperatively), the conducting tissue was situated on the right lateral wall of the common ventricle. In two other specimens the conducting tissue took a posterior route, but in neither

did it follow the line which appeared most probable on gross inspection and hence attempts to avoid it surgically would be largely a matter of chance.

T₄ levels in normal neonates. R. H. Davies introduced by R. S. Jones. Alder Hey Children's Hospital, Liverpool.

Experience with use of CPAP in respiratory distress in the newborn. N. R. C. Robertson and J. D. Baum. Department of Paediatrics, John Radcliffe Hospital, Oxford.

Effect of continuous positive airway pressure on the lung mechanics of babies with congenital heart disease. J. J. Cogswell, D. J. Hatch, B. W. Taylor, and A. A. Kerr. The Hospital for Sick Children, London.

The effect of continuous positive airway pressure (CPAP) on the lung mechanics of 14 babies with congenital heart disease was investigated. At the time of study all babies were receiving ventilatory support and were temporarily disconnected from the ventilator to allow measurements to be made during spontaneous breathing. During CPAP there was a small but significant fall in minute ventilation. There was no consistent change in dynamic compliance. There was a significant fall in pulmonary resistance. In those babies with small thoracic gas volumes before the study there was a significant fall in the work of breathing during CPAP. It is suggested that these changes are due to an increase in the functional residual capacity.

Pitfalls in neonatal insufflation. P. M. Dunn, H. Perez-Alzueta, and B. D. Speidel. Department of Child Health, University of Bristol.

Positive pressure insufflation of the lungs is the most effective method of resuscitating the severely depressed newborn infant. Pressures as great as 30 mmHg may be required to achieve initial expansion, though much lower pressures are usually sufficient to maintain ventilation thereafter. As pressures greater than 30 mmHg may cause alveolar rupture, neonatal insufflation equipment is usually designed to avoid the generation of higher pressures, often by the incorporation of a safety device. To test the safety of currently used resuscitation equipment, dynamic pressure recordings within an intubated model lung were made using 7 different methods of insufflation: mouth-to-mouth ventilation, face mask and bag (M.I.E.), the Ambu baby resuscitator (Ambu International), the Cardiff infant inflating bag (Penlon), the Stephenson minuteman resuscitator (B.O.C.), the EPAC resuscitator (B.O.C.), and the Resuscitaire infant trolley (Vickers Ltd.). In each case the equipment was applied by paediatricians experienced in newborn resuscitation who were not permitted to watch the pressure trace during recording nor to practise beforehand. The results of these investigations were reported, drawing attention to the dangers inherent in the use of many of these techniques.