disease progresses could represent regression of testicular androgen synthesis to the fetal type.

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Congenital Malformations and Neoplasia. Jean Keeling (introduced by Colin Berry). Published in full—Berry, C. L., Keeling, J., and Hilton, C. (1970). *Archives of Disease in Childhood*, **45**, 229. Coincidence of congenital malformation and embryonic tumours of childhood.

Diminished Activity of Platelet Monoamine Oxidase in Down's Syndrome. Philip F. Benson and Jennifer Southgate (*Paediatric Research Unit, Guy's Hospital, London S.E.1*). Subjects with primary trisomic Down's syndrome have low concentrations of blood serotonin (5-hydroxytryptamine) (5-HT). The reason for this is unknown. In Down's syndrome the binding of 5-HT to platelets has been found to be normal. In theory low blood 5-HT may be due either to a diminished rate of synthesis or to an accelerated breakdown.

We report a study on platelet monoamine oxidase (MAO) activities in subjects with Down's syndrome and in controls. While the plasma enzyme is practically inactive with 5-HT, platelet MAO appears to be very similar to the liver mitochondrial enzyme which is the main reservoir of MAO in the body.

Platelet MAO activity (measured fluorimetrically using kynuramine as substrate and expressed as μg . 4-hydroxyquinoline/mg. protein/30 minutes) was significantly lower in children with Down's syndrome (22 subjects, mean 0.81 SD 0.38) than in controls matched for age, sex, and domicile (22 subjects, mean 1.24, SD 0.70) $p = 0.01 - 0.02$. This does not support the view that low concentration of blood 5-HT is due to an accelerated rate of 5-HT breakdown. Moreover, a decrease in MAO activity is compatible with the observation that urinary excretion of 5-hydroxy-indoleacetic acid (5-HIAA) is lower in subjects with Down's syndrome than in controls. If accelerated degradation of 5-HT cannot be incriminated as the cause of low blood 5-HT in Down's syndrome, it is reasonable to suggest that there may be a diminished rate of 5-HT synthesis. This might be due to inefficient intestinal absorption of tryptophan or to diminished activity of enzymes involved in 5-HT synthesis. Theoretically the trisomic chromosome 21 might carry a regulatory gene for MAO, leading to increased synthesis of repressor and therefore to a decreased rate of MAO synthesis.

Effect of Cytotoxic Drugs on Immature Rat's Gonads and Subsequent Reproductive Performance. Colin Berry (*Institute of Child Health, London W.C.1*). (To be published in full elsewhere.)

Comparison of Effect of Isoprenaline and Salbutamol Aerosols on Airways Obstruction and Pulse Rate of Children with Asthma. G. Hambleton (introduced by Malcolm Segall) (*Department of Paediatrics, St. Thomas's Hospital S.E.1*). (To be published in full in this journal.)

Total Faecal Bile Acid Excretion in Children. Clive Leyland (introduced by June Lloyd) (*Institute of Child Health, London W.C.1*). Few studies have been made on total faecal bile acid excretion and no data are available for children.

Using gas-liquid chromatography (Grundy, Ahrens, and Miettinen, 1965) faecal bile acids have been estimated on pooled 3-day stool collections. In 16 children (aged 2 months to 14 years) without evidence of fat malabsorption, the mean daily excretion varied between 10 and 85 mg. and appeared to correlate with body weight.

Very low amounts ($< 3 \text{ mg./day}$) were found in 5 children who had biliary atresia. Increased excretion was found in a child with familial hypercholesterolaemia receiving cholestyramine and in 2 children with resection of the terminal ileum.

Raised excretion was found in 2 children with cystic fibrosis who had steatorrhoea, whereas in 2 other children with cystic fibrosis, in whom steatorrhoea was controlled by a low fat diet, faecal bile acid excretion was normal.

In 5 children with other diseases causing fat malabsorption steatorrhoea had been controlled by dietary fat reduction. 4 had also received supplementary medium-chain triglyceride. In all these children faecal bile acid excretion was normal.

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Sugar Absorption in Rats with Intestinal Blind Loops. Michael Gracey, Valerie Burke, and Ademole Oshin (introduced by Graham Chance) (*Institute of Child Health, University of Birmingham*). Temporary monosaccharide malabsorption in young babies may be associated with abnormal small intestinal flora and deconjugated bile salts (Gracey, Burke, and Anderson, 1969). To investigate this association we have studied the uptake of an actively transported, non-metabolized analogue of glucose (Arbutin) in rats with intestinal blind loops.

Blind loops, 8–10 cm. in length, were constructed in the jejunum of adult Wistar rats. One to two months after operation, 5-day stool collections demonstrated steatorrhoea in all operated animals. In 6 animals, using a recently described *in vitro* technique (Semenza, 1969), and using randomized segments of intestine, we have shown inhibition of Arbutin uptake in the blind loops and in the intestine adjacent to the loops, both proximally and distally. Inhibition was most obvious in the blind loops and was least severe in the proximal segments. In 3 animals the degree of inhibition was

dependent on the distance of the segment studied from the blind loop.

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Serum Phospholipid Levels in Growth Hormone Deficient Children. C. G. Theodoridis, P. H. W. Rayner, E. C. Albutt, and G. W. Chance. (*Department of Paediatrics and Institute of Child Health, University of Birmingham*). Serum total phospholipids were measured by Bartlett's (1959) method in 10 growth hormone deficient children aged 3–18 years and in 21 short stature children of similar age-group who were endocrinologically and metabolically normal.

The mean serum total phospholipid levels obtained from the HGH deficient children— 273 ± 20 mg./100 ml. (range 250–315 mg./100 ml.)—were much higher than the mean obtained from the non-growth hormone deficient children— 205.4 ± 22 mg./100 ml. (range 160–250 mg./100 ml.). This difference was statistically significant ($p < 0.001$). This difference could not be accounted for by thyroid deficiency as only 3 HGH deficient patients had associated TSH deficiency.

Despite raised serum total phospholipids the percentage distribution of serum phospholipid phosphorus between the 4 phospholipid fractions (separation was done by thin layer chromatography) was similar to that of the controls (Table).

TABLE

	GH Deficient Children	Control Children
Lysophosphatidyl choline	9.4 ± 1.5	9.5 ± 1.2
Sphingomyelin	20 ± 0.4	20.4 ± 0.5
Phosphatidyl choline	65.5 ± 3	65.1 ± 0.9
Phosphatidyl ethanolamine	5.1 ± 1.9	5.0 ± 0.7

A 20% fall in serum total phospholipid was observed in 3 children with isolated growth hormone deficiency,

who received growth hormone treatment for 3 months.

In summary, raised total phospholipid levels are demonstrable in children with HGH deficiency. The administration of exogenous HGH leads to a fall of serum total phospholipids. There is no change in the percentage distribution of serum phospholipid phosphorus between the 4 phospholipid fractions in these children.

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- Bartlett, G. R. (1959). Phosphorus assay in column chromatography. *Journal of Biological Chemistry*, **234**, 466.

Vitamin E Therapy in A- β -lipoproteinaemia. D. R. P. Muller, J. T. Harries, and June K. Lloyd (*Institute of Child Health, London*). A- β -lipoproteinaemia is a rare inborn error of metabolism characterized by the absence of β -lipoprotein from the blood, acanthocytosis of the red cells, and steatorrhoea (present from birth), and an ataxic neuropathy and pigmentary retinopathy which develop later and are slowly progressive.

We have investigated the vitamin E status of 6 children with a- β -lipoproteinaemia by estimating serum concentrations of vitamin E and red cell haemolysis. Initially serum vitamin E was undetectable in all of the children, and haemolysis raised ($> 6\%$) in the 5 in whom it was estimated. Oral administration of large doses of vitamin E resulted in rapid correction of the abnormal haemolysis. Serum vitamin E remained undetectable for approximately 6 months in 4 children who received doses varying between 25 and 75 mg./kg. per day, whereas in 2 children who received even larger doses (100 mg./kg. per day) serum vitamin E was detectable after 1 and 3 months, respectively. The maximum level achieved by oral therapy was 0.24 mg./100 ml. (normal — 0.44 mg./100 ml.) which approached the peak levels of 0.3 mg./100 ml. obtained in 2 children who received a large, single intramuscular load.

In 4 children oral therapy has now been given for longer than 3 years, and the retinal and neurological abnormalities present in 2 children have been studied sequentially by tests of motor nerve conduction and retinal function; one child has shown no deterioration and the other a definite improvement. As the natural history of the condition is towards progressive deterioration, and as no other form of therapy was introduced during administration of vitamin E, it seems likely that this vitamin has contributed to the improvement.