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PROCEEDINGS OF THE NUTRITION SOCIETY

ABSTRACTS OF COMMUNICATIONS

The Three Hundred and Thirty-third Meeting of the Nutrition Society for the presentation of original communications was held in the Kennedy Lecture Theatre, Institute of Child Health, Guilford Street, London on Wednesday, 19 *September,* 1979, *when the following papers were read:*

https://doi.org/10.1079/PNS19790067 Published online by Cambridge University Press

Branched chain amino acid metabolism in low birth weight babies and the effects of dietary protein quality. By P. H. SCOTT, H. M. BERGER and B. A. WHARTON, *Biochemistry Department, Selly Oak Hospital and Sorrento Maternity Hospital, Birmingham*

In the development of 'humanized' infant feeding formulae the quality of the protein component has been altered from the predominantly curd cow's milk type to the more balanced curd and whey combination of proteins. The effects of this on growth and general metabolism have recently been studied (Berger *et al.* 1979). However, the associated changes in the metabolism of amino acids as reflected in their plasma concentrations has not been reported.

Fifty-five preterm or term light-for-dates babies, birth weight 1750–2260 g, were studied. They were fed either a curd formula (based on SMA) or a curd and whey formula (based on Gold Cap SMA-S26; John Wyeth, Maidenhead, Berks.). Nitrogen balance studies were performed on a smaller group of these babies during the third week.

Infants fed the curd and whey formula grew better over the period from day 7 to day 21. Plasma amino acid levels on day 11 were generally higher in those babies fed the curd formula. From day 11 to day 21 the levels fell so that by day 21 those differences which remained were only present in the preterm group. The plasma branched chain amino acid levels on day 21 correlated inversely with growth over the period of study, and also correlated inversely with nitrogen retention.

It is probable that peripheral utilization of branched chain amino acids by muscle is an important determinant of their plasma levels. Those babies which grow faster utilize a greater proportion of available branched chain amino acid and therefore have lower plasma levels. Poorer growth and therefore poorer utilization results in loss of nitrogen as urinary urea. Indeed, urinary valine excretion correlated inversely with nitrogen retention. Lower levels of branched chain amino acids found in those infants who grew better may indicate some inadequacy of protein intake. The valine:glycine value is lower in the preterm curd and whey group on both days 11 and 21, suggesting a poorer nutritional status.

Plasma branched chain amino acid levels in low birth weight babies fed a curd whey protein formula are lower than in a similar group fed a predominantly curd protein formula. This is probably associated with their higher rate of growth and may indicate some dietary inadequacy of protein.

Berger, H. M., Scott, P. H., Kenward, C., Scott, P. & Wharton, B. A. (1979). *Archs Dis. Childh.* **54**, 98.

0029–6651/79/3803–333A $00.35

Cell size and cell number: a reconsideration of organ growth and catch-up potential. By J. DOBBING, JEAN SANDS and CHARMAINE A. GRATRIX, *Department of Child Health, University of Manchester, The Medical School, Manchester M*13 9*PT*

The last decade has seen a widespread acceptance of the doctrine (Winick & Noble, 1965) that tissues and organs pass through three phases of growth and development. The first mainly consists of cell multiplication (hyperplasia); the last, of growth in cell size (hypertrophy); and a middle phase during which the cessation of cell multiplication overlaps with the beginning of growth in cell size. Cell number and size in this context have been measured indirectly by total organ DNA; and weight or protein content per unit of DNA respectively. The importance of this doctrine has been due to the suggestion (Winick & Noble, 1966) that nutritional growth restriction during the first phase, but not during the last, is followed by a resistance to 'catch-up' on rehabilitation.

We have re-examined the basic premise in the liver, heart, kidney and gastrocnemius of 392 developing rats, ranging in age from 18 d of gestation to 112 d of postnatal life, and have found that the sequence is very different from that which has been described. Cell multiplication, instead of ceasing early with an adult number of cells, continues throughout the entire period of growth in organ weight. Growth in mean cell size begins in the early stages of the period studied, not later, and is complete long before the end of cell multiplication or growth in organ weight.

It is therefore difficult to accept the earlier hypothesis regarding catch-up growth; and the commonly derived, inverted hypothesis relating later obesity to an early period of excessive adipocyte multiplication may be similarly threatened. Other aspects of the earlier description of growth will also be criticized, including the use of general expressions of cell number and cell size in tissues with cell populations which are heterogeneous, not only in type, but also in their sequence of multiplication.

More realistic descriptions of characteristics determining catch-up potential are being sought.

Winick, M. & Noble, A. (1965). *Dev. Biol.* **12**, 451.
Winick, M. & Noble, A. (1966). *J. Nutr.* **89**, 300.

0029–6651/79/3803–333A $00.35

The effect of a diet rich in linoleic acid on fetal brain lipids. By T. A. B. SANDERS and D. J. NAISMITH, *Department of Nutrition, Queen Elizabeth College, London W8 7AH*

The substitution of vegetable oils, and margarines rich in linoleic acid (18:2ω6) for animal fats and butter in the British diet has been advocated in the belief that this change may reduce the incidence of coronary heart disease (Royal College of Physicians/British Cardiac Society, 1976). Yet little is known about the effects of a high maternal consumption of linoleic acid on fetal development, in particular on the composition of brain lipids. A high intake of linoleic acid has been shown to inhibit the conversion of ∝-linolenic acid (18:3ω3) to docosahexaenoic acid (22:6ω3) in the liver of the rat (Rahm & Holman, 1964). Since docosahexaenoic acid is a major component fatty acid of neuronal phosphoglycerides, it seemed important to determine whether the fetal brain was also susceptible to the influence of a high concentration of linoleic acid on linolenic acid metabolism.

Eight rats were mated, and transferred from a stock diet to an experimental diet containing 200 g margarine/kg. Eight litter-mates were given a similar diet in which the margarine was replaced with a 1:1 (w/w) mixture of butter and lard. Linoleic and ∝-linolenic acid accounted for 425 mg/g and 5 mg/g of the total fatty acids respectively in the margarine diet, and 61 mg/g and 12 mg/g in the butter–lard diet. On day 22 of pregnancy the rats were killed and their fetuses were dissected. The mean number of pups/litter, and the weight of the pups and placentas were similar in the two groups.

In comparison with pups from the group fed on the butter–lard diet, pups from the margarine group showed a marked reduction in the proportion of docosahexaenoic acid (22:6ω3) in the brain phosphoglycerides ($57\pm2\cdot7$ mg/g compared with $92\pm2\cdot4$ mg/g; $P<0\cdot01$). There was, however, a corresponding increase in the proportion of docosapentaenoic acid (22:5ω6) derived from linoleic acid ($67\pm3\cdot3$ mg/g compared with $28\pm1\cdot4$ mg/g; $P<0\cdot01$). Whether this difference in the nature of the major long-chain polyunsaturated fatty acid in the brain lipids is of physiological importance remains to be determined.

We are grateful to the Rank Prize Funds for a research grant.

Rahm, J. J. & Holman, R. T. (1964). *J. Nutr.* **84**, 15.
Royal College of Physicians/British Cardiac Society (1976). *J. Roy. Coll. Phycns* **10**, 213.

0029-6651/79/3803-333A $00.35

Aspects of sulphur metabolism in low birth weight babies. By H. M. BERGER, P. H. SCOTT and B. A. WHARTON, *The Infant Development Unit, Queen Elizabeth Medical Centre, Edgbaston, Birmingham B15 2TG*

Sulphur is an essential part of certain mucopolysaccharides and forms salts with important steroid molecules such as bile acids and vitamin D. The source of S is mainly cysteine either present in the diet or formed from dietary methionine.

Aspects of S metabolism have been studied in low birth weight babies receiving varying amounts of cysteine. Babies receiving the higher cysteine intake from a cow's milk formula excreted less urea, more sulphate, and had a lower urinary cystathionine:cysteine value. Unlike babies receiving breast milk, however, duodenal bile acids were conjugated predominantly with glycine rather than taurine.

These observations are compatible with the known immaturity of the transulphuration pathway but show that this immaturity is still evident at 3 weeks of age in low birth weight babies. These observations may have implications for the advisable diet of the newborn.

0029–6651/79/3803–333A $00.35

Nutritional anaemia in childhood coeliac disease. By D. STEVENS, *Department of Paediatrics, Gloucestershire Royal Hospital, Gloucester*

Thirty-four children out of ninety-one (37%) (aged 2 months to 9 years) with coeliac disease were anaemic at diagnosis. The mean haemoglobin was lower and incidence of anaemia significantly greater ($P<0.05$) in children of 2 years and above. The duration of the illness prior to diagnosis might account for the difference in incidence of anaemia with age because iron stores have to be totally depleted before erythropoiesis is affected. The mean duration of symptoms prior to diagnosis was much longer in the older children (184·4 weeks compared to 16 weeks).

Red cell folate was low (below 160 ng/ml) in 50% of the estimations (34). The incidence (7) of low erythrocyte folate estimations (9) was greater in children above the age of 5 years. However, there was little evidence that folic acid deficiency played a part in the pathogenesis of anaemia. Half the erythrocyte folate estimations were normal in anaemic children and only three children had macrocytic erythrocytes on blood film examination and none had hypersegmented neutrophils. Erythrocyte folate is a poor screening test for coeliac disease because half the estimations were normal in children with coeliac disease.

Children with coeliac disease, whether they are anaemic or not, are likely to have evidence of Fe deficiency. Serum Fe was below 12 μmol/l in 87% and percentage saturation of transferrin below 16% in 93% of estimations. All anaemic children had hypochromic blood films.

Less than half (37%) of children with coeliac disease in the first 10 years of life were anaemic at diagnosis. The incidence of anaemia was significantly greater in children at least 2 years old. All anaemic children had evidence of Fe deficiency. There was little evidence that folic acid deficiency played a part in the pathogenesis of anaemia. Erythrocyte folate is a poor screening test for coeliac disease because half the estimations were normal in children with coeliac disease at diagnosis.

0029–6651/79/3803–333A $00.35

The nutrition of the Asian immigrant mother in relation to birth weight.

By J. G. BISSENDEN, P. H. SCOTT and B. A. WHARTON, *Institute of Child Health, Department of Biochemistry, Selly Oak Hospital and Sorrento Maternity Hospital, Birmingham*

Evidence that undernutrition of the pregnant mother compromises birth weight was based largely on war-time studies such as the siege of Leningrad (Antonov, 1947) and the Dutch famine (Smith, 1947). However, current work in countries such as Guatemala has shown that improving the nutrition of the mother results in larger babies (Lechtig *et al.* 1975) which when later assessed had a better intellectual status than babies from non-supplemented control pregnancies (Klein *et al.* 1976).

We have performed a prospective study on pregnant Asian mothers in Birmingham, relating their plane of nutrition to birth weight. Two techniques were used. Firstly, the anthropometry of the mother was measured including weight, biceps, triceps, and subscapular skinfold thicknesses, and mid upper-arm muscle circumference. At the same time, biochemical estimations of the mother's nutritional status were measured. These included urinary nitrogen and hydroxyproline excretion, plasma valine:glycine value and serum alkaline ribonuclease.

The results showed that compared to mothers producing well grown babies (above the 10th centile using the correction of Thomson *et al.* 1968), mothers producing babies under the 10th centile, put on less weight and fat in the second trimester. At this stage of pregnancy they also had biochemical signs of a lower nutritional plane. (Higher serum alkaline ribonuclease and reduced excretion of peptide hydroxyproline). Significantly, out of eighteen pregnancies producing poorly grown babies, only six were complicated by hypertension.

It is clear from this study that the nutrition of the pregnant Asian mother is important. We suggest that those mothers showing anthropometric and biochemical signs of undernutrition in the second trimester should receive a food supplement in the third.

Antonov, A. N. (1947). *J. Pediat.* **30**, 250.
Klein, R. E., Arenales, P., Delgado, H., Engle, P. L., Gusmian, G., Irwin, M., Lasky, R., Lechtig, A., Martorell, R., Mej'ia Pivaral, V., Russell, P. & Yarbrough, C. (1976). *Bull. Pan Am. Hlth Org.* **10**, 301.
Lechtig, A., Delgado, H., Lesky, R., Yarbrough, C., Klein, R. E., Habicht, J-P. & Behar, M. (1975). *Am. J. Dis. Child.* **129**, 553.
Smith, C. A. (1947). *Am. J. Obstet. Gynec.* **53**, 559.
Thomson, A. M., Billewicz, W. Z. & Hytten, F. E. (1968). *J. Obstet. Gynaec. Br. Commonw.* **75**, 903.

0029–6651/79/3803–333A $00.35

Effect on vitamin D metabolism of reconstructive surgery in children with tracheo-oesophageal fistulae. By L. S. TAITZ, L. SPITZ and R. R. G. RUSSELL, *Department of Pediatrics, Children's Hospital, Western Bank, Sheffield S10 2TH*

Two children are described who developed hypocalcaemic seizures several years after undergoing major alimentary tract surgery following failure of early reconstructive attempt to correct tracheo–oesophageal fistula and atresia.

In both children there was evident exclusion of gastric mucosa from the digestive system. The investigations revealed that the hypocalcaemia resulted from inadequate availability of vitamin D and was corrected by modest amounts of this vitamin. This conclusion was supported by the presence of secondary hypoparathyroidism.

The possible mechanism for this vitamin D deficiency or inadequate absorption will be discussed.

0029–6651/79/3803–333A $00.35

Taurine in breast milk: a role in fat utilization. By D. J. NAISMITH and KAREN N. CASHEL, *Department of Nutrition, Queen Elizabeth College, London, W8 7AH*

The free taurine content of breast milk is reported to be ten times that of cow's milk (Ghanidi & Pecora, 1963), and the proteins of breast milk contain twice as much cysteine from which taurine may be synthesized (Department of Health & Social Security, 1977). In early infancy, however, the capacity to form cysteine from methionine, and taurine from cysteine is severely limited (Gaull *et al.* 1977).

In the neonate, the digestibility of the fat in cow's milk formulae is substantially below that in breast milk, although it gradually improves with time (Widdowson, 1974). The digestion and absorption of fat is influenced by a number of factors, including the bile salts. In the young breast-fed infant bile salts are conjugated predominantly with taurine, whereas in infants fed cow's milk formulae, as in older children and adults, glycine-conjugated bile salts predominate (Brueton *et al.* 1978). The poor digestibility of fat in cow's milk formulae might therefore be due, in part, to a difference in the properties of the bile salts.

To test this hypothesis, the effects of different concentrations of taurocholic acid and glycocholic acid, ranging from 2·5 mM to 12·5 mM on the activity of human pancreatic lipase were investigated in vitro. Ostermilk Complete Formula, prepared as for a feed, was used as substrate, and a duodenal aspirate taken from an adult male subject following a Lundh test meal was used as a source of lipase. After the incubation at 37°, total lipids were extracted, and the various glycerides and free fatty acids were separated by thin layer chromatography, charred with sulphuric acid, and quantified with a scanning photodensitometer. The rate of lipolysis was measured by estimating the free fatty acids in the mixture.

Taurocholic acid had a maximal effect on free fatty acid release within the first 10 min of the reaction. At a concentration of 5 mM, a value reported to be the 'critical micellar concentration' in in vitro systems (Morgan & Hoffman, 1971), an increase of 36% was found, and at 7·5 mM, an increase of 113%. In contrast, additions of glycocholic acid had no appreciable effect until a concentration of 12·5 mM was reached, when a stimulation of 21% was observed.

Bile composition may thus be an important determinant of fat utilization in the young infant.

Brueton, M. J., Berger, H. M., Brown, G. A., Ablitt, L., Iyngkaran, M. & Wharton, B. A. (1978). *Gut* **19**, 95.

Department of Health and Social Security (1977). *The Composition of Mature Human Milk*, London: H.M.S.O.

Gaull, G. E., Rassin, D. K., Raiha, N. C. R. & Heinonen, K. (1977). *J. Pediat.* **90**, 348.

Ghanidi, H. & Pecora, P. (1963). *Am. J. clin. Nutr.* **13**, 75.

Morgan, R. C. H. & Hoffman, N. E. (1971). *Biochim. Biophys Acta.* **248**, 143.

Widdowson, E. M. (1974). In *Scientific Foundations of Paediatrics* [J. A. Davis and J. Dobbing, editors]. London: Heinemann.

0029–6651/79/3803–333A $00.35

The role of zinc in childhood diabetes mellitus. By F. W. ALEXANDER, *Newcastle General Hospital, Newcastle upon Tyne*

We have investigated the zinc status of eight insulin dependent diabetics before and after treatment with insulin.

Quarterman *et al.* (1966) observed a serious impairment of blood glucose homoeostasis in Zn deficient rats. They also showed that glucose uptake by adipose tissue is increased by insulin but further increased by the addition of Zn. These results suggest that Zn is necessary for insulin production and that Zn deficiency may be a contributory factor to some diabetic states.

Serum and 6 h urine samples were collected during the initial investigative period and again after stabilization with insulin from children presenting with diabetes mellitus. Zn concentrations were measured by atomic absorption spectrophotometry after suitable preparation. Urine samples were collected from suitable controls for Zn levels.

Zn concentration in the serum of diabetics before treatment was (mean $\pm$SE) $12{\cdot}7\pm3{\cdot}6$ μmol/l which was higher than, but not significantly different from, the level of $9{\cdot}7\pm3{\cdot}8$ μmol/l after 6 weeks treatment. Urinary concentration of this was also higher before than after treatment (513 ± 299 compared with 478 ± 331 μmol/l). Both of these values are greater than that for control subjects at 199 ± 48 μmol/l, but again the levels were not significantly different.

There is thus no evidence to support the hypothesis that Zn deficiency contributes to the onset of diabetes mellitus. There is, however, a suggestion that diabetes causes zincuria and may lead to an over-all loss of Zn as reported by Culebras *et al.* (1977).

Quarterman, J., Mills, C. F. & Humphries, W. R. (1966). *Biochem. Biophys. Res. Commun.* **25**, 354.

Culebras, J. M., Luque, M. J., Ribas, B., Santiago, M. & Dean, M. (1977). T.E.M.A. 3. p. 381.

0029–6651/79/3803–333A $00.35

The role of hyperinsulinaemia in chemically induced obesity. By G. P. WEBB, P. D. ROGERS and S. A. JAGOT, *Department of Paramedical Sciences, North-east London Polytechnic, London E15 4LZ*

There have been several reports of hyperinsulinaemia in animal models of obesity; the demonstration of a high correlation between plasma insulin and the degree of obesity has led to the suggestion that hyperinsulinaemia may be a causative factor of obesity (see Assimacopoulos-Jeannet & Jeanrenaud, 1976). We induced obesity in T/O mice by administration of either gold thioglucose (GTG) or bipiperidyl mustard (BPM) (Rutman *et al.* 1966) and maintained the animals on one or two laboratory diets 41B or FFG(M). Fourteen weeks after injection we determined the total body fat and plasma insulin levels of the mice (Table 1).

Table 1. *Plasma insulin levels (mU human insulin equivalent) and percentage fat of T/O mice*

(Values are means with their standard errors for ten (*nine) determinations)

	41B				FFG(M)			
	Insulin		Fat (%)		Insulin		Fat (%)	
	Mean	SE	Mean	SE	Mean	SE	Mean	SE
Control	43·5	17·8	14·8	2·1	44·9	11·9	13·9	1·3
GTG	27·6	11·2	23·5	3·3	88·0	24·1	25·3	2·8
BPM	11·2	3·2*	15·8	1·5*	113·6	20·9	31·6	3·3

Analysis of variance for insulin: treatment NS; diet $P<0.001$; interaction $P<0.05$.
Analysis of variance for % fat: treatment $P<0.001$; diet $P<0.001$; interaction $P< 0.005$.

Two way analysis of variance showed there was no significant effect of the fattening agents on plasma insulin but animals on FFG(M) were highly significantly hyperinsulinaemic ($P<0.001$) and there was a significant interaction between diet and fattening agent ($P<0.05$). There were also highly significant effects of both diet and fattening agent ($P<0.001$) on percentage fat and a significant interaction ($P<0.005$) between diet and treatment. The 41B fed, GTG treated group were significantly fatter than the controls ($P<0.025$) but were not hyperinsulinaemic. When the coefficient of correlation between plasma insulin and percentage fat was calculated it was significant over-all but not when calculated for the animals on each diet or when only the control animals were considered.

However, when the results for the treated animals only were considered there was a significant correlation both over-all ($P<0.001$) and for each diet considered separately (41B $P<0.01$; FFG(M) $P<0.001$). Further analysis of the correlation between plasma insulin and percentage fat will be discussed.

These results indicate the importance of diet in the chemically induced obesity syndrome in mice and demonstrate that such obesity occurs both in the presence and absence of hyperinsulinaemia.

Assimacopoulos-Jeannet, F. & Jeanrenaud, B. (1976). *Clinics Endocr. Metab.* **5**, 337.
Rutman, R. J., Lewis, F. S. & Bloomer, W. D. (1966). *Science* **153**, 1000.

0029–6651/79/3803–333A $00.35

Lipoprotein lipase and hepatic lipase activities in children with hyperlipidaemia. By T. R. GAMLEN, D. P. R. MULLER and O. H. WOLFF, *Department of Child Health, Institute of Child Health, 30 Guilford Street, London WC1N 1EH*

Chylomicrons and very low density lipoprotein are cleared from the circulation by the action of lipoprotein lipase (LPL) at the capillary endothelium. The resulting particles are removed by the liver and converted into low density lipoprotein, probably by hepatic lipase (HPL). Both LPL and HPL are released into the circulation by low dose intravenous heparin, and we have developed methods for the direct assay of both these lipases in post-heparin plasma.

The technique was validated using rat adipose and liver tissue extracts as sources of LPL and HPL respectively, and their activities were assayed using modifications of the specific LPL and HPL radiolabelled substrates described by Nilsson-Ehle & Eckman (1977). Radiolabelled rat chylomicrons were also used as a specific LPL substrate and the results with this natural substrate correlated well with those using the artificial LPL triolein substrate (r 0·84).

LPL and HPL activities were assayed in post-heparin plasma from healthy adults and children with primary and secondary hyperlipidaemic states. The mean LPL and HPL activities of six healthy males were (mean±SD) 2·2±1·1 and 12·5±3·3 μmol of fatty acid released/ml post-heparin plasma per h respectively. Four patients with hyperchylomicronaemia (type I hyperlipoproteinaemia) had, as expected, reduced LPL (0·4±0·4) and normal HPL (6·1±3·2) activities. Seven children with familial hypercholesterolaemia had normal LPL and HPL activities (3·6±1·6 and 7·1±4·7 respectively). Five children with glycogen storage disease had reduced LPL and HPL activities (0·5±0·3 and 1·8±0·9 respectively). The plasma of patients with poorly controlled glycogen storage disease was found to inhibit normal HPL activity. Preliminary longitudinal studies suggest that with improved metabolic control serum concentrations of triglyceride fall, HPL and LPL activities increase and the inhibitory effect of the plasma on normal HPL activity becomes less pronounced.

Nilsson-Ehle, P. & Eckman, R. (1977). *Artery* **3**, 194.

0029–6651/79/3803–333A $00.35

Parents and children in oral acting and dietary habits. By E. HARJU and E. VÄISÄNEN, *Departments of Physiology and Psychiatry, University of Oulu, Kajaanintie* 52 *A*, 90220 *Oulu* 22, *SF-Finland*

The Finnish diet contains too few vegetal products and too many saturated fat and milk products. To study the factors influencing food habits, a questionnaire survey was carried out among fifteen ice hockey players, eighteen male and twenty female students and twenty female office workers, taken from a group of 1200 people who had their dietary habits previously examined by food records method. The results from the survey were in accordance with those from the food records.

Parents have significant influence on food habits (r 0·661). The mother seems to be the more important model in food practices: 81% of the subjects expressed the opinion that their mother was concerned about their eating habits, whereas the fathers' attitudes varied. A change in mothers' food habits and attitudes is the first step to improve those of the other members of the family. The increase of fathers' interest in nutrition would indicate a new balance in family psycho-dynamics.

The obesity of the mother correlated to the smoking habits of their daughters (r 0·696). The mothers eat when anxious and the girls usually smoke but both have a real oral method of satisfaction. Female office workers who are heavy smokers are likely to have taken their father as a model for developing their smoking habits (r 0·904). The father seems to be the object of introjection for his daughter. The mother shows her daughter the subjective way of reacting and the father shows her the objective way. In female office workers, alcohol consumption correlated with mothers living until their daughters reached the age of sixteen (r 0·688). Thus the mother may be acting as a model for their alcohol consumption.

The appreciation of porridge and tea among those who left home at an early age (r 0·427) may indicate that food habits learnt in early childhood are the most permanent. The appreciation of butter among subjects may have been learnt from parents who have previously lived during a period of food shortage.

Changes in dietary habits often happen through the influence of other members of the family and the most effective way of teaching the essentials of good nutrition lies in personal contacts. The child soon learns and introjects what is acceptable and as an adult has strongly fixed food habits. Thus the parents not only should have good nutritional practices but also their emotional relations should be warm and trusting.

0029–6651/79/3803–333A $00.35

Muscle phosphofructokinase deficiency (Tarui's disease). By M. J. TARLOW, D. A. ELLIS, G. W. PEARCE and M. ANDERSON, *East Birmingham Hospital, and Midland Centre for Neurosurgery and Neurology*

Phosphofructokinase is a key enzyme in glycolysis, and in its absence energy supply to skeletal muscle is grossly compromised.

Deficiency was first described in 1965, and since then seven cases have been described in the world literature. It has not been previously recognized in a child.

A 7 year old boy presented with a history of severe muscle cramps from the age of two, and easy fatiguability became manifest 2 years later. Subjecting the forearm muscles to ischaemic exercise led to no rise in serum lactate or pyruvate (McArdle's test) and a tentative initial diagnosis of muscle phosphorylase deficiency was made. EMG, muscle histology, and electron microscopy were within normal limits, but on enzyme assay of a muscle biopsy, no phosphofructokinase could be detected.

Unlike other cases recorded in the literature, no evidence of haemolytic anaemia could be detected in our patient, and no form of treatment has been effective.

0029-6651/79/3803-333A $00.35

Fructose 1,6 diphosphatase deficiency. By M. J. TARLOW, A. D. PATRICK, D. R. CARLTON and M. E. BARTON, *East Birmingham Hospital and Child Health Institute, University of London*

Fructose 1,6 diphosphatase is a central enzyme in gluconeogenesis, the synthesis of glucose and glycogen from non-carbohydrate precursors.

Deficiency of hepatic fructose 1,6 diphosphatase was first described in 1960, and since then thirteen reports have appeared in the literature.

A 2 year old Pakistani boy, the fourth child of unrelated parents, presented at the age of 11 months with severe dehydration and metabolic acidosis in the course of a mild rotavirus gastroenteritis. At that time, hepatomegaly and transiently elevated transaminases were noted.

He has since had recurrent episodes of severe dehydration and metabolic acidosis, sometimes associated with hypoglycaemia. Aged 19 months, he was readmitted with profound hypoglycaemia and convulsions (glucose 0·3 mmol/l) but only moderate acidosis.

Initial biochemical investigations (including raised serum lactate, triglycerides and uric acid; and a glucose tolerance test) suggested the diagnosis of Type I glycogen storage disease. Enzyme analysis of the liver biopsy excluded all types of glycogen storage disease, but demonstrated complete absence of fructose 1,6 diphosphatase. Treatment with a fructose free diet has proved ineffective. Crises have usually, but not invariably, been precipitated by infection.

0029-6651/79/3803-333A $00.35

Management of hepatic glycogen storage disease with regular oral glucose drinks. By D. B. DUNGER and J. V. LEONARD, *Department of Child Health, Institute of Child Health, London*

Striking improvements in well-being and growth velocity have been achieved in patients with hepatic glycogen storage disease (GSD) when the blood sugar has been maintained within the normal range by frequent glucose drinks by day and naso-gastric infusions at night (Greene *et al.* 1976). However, this approach is not always practical and not without danger (Leonard & Dunger, 1978). In milder cases, such intensive therapy may not be necessary.

Three patients (one patient with type Ia, one with Ib and one with type III) who are relatively tolerant of hypoglycaemia, but who were short with persistent hyperlipidaemia, hyperuricaemia, and lactic acidosis, were treated with regular oral glucose supplements. The glucose was given as a short-chain polymer, hourly by day and 2–3 hourly by night. The families managed the treatment well and the general health of the children improved. There was a marked increase in growth velocity and improvement of the biochemical abnormalities (see Table).

Effect of oral treatment in Type Ib glycogen storage disease

	Age (years)	Stature (m)	Stature SD*	Growth velocity (mm/year)	Growth velocity SD*	Total cholesterol (mmol/l)	Tri-glyceride (mmol/l)	Uric acid (mmol/l)	ALT (IU/l)	AST (IU/l)
Before treatment	5·45	0·85	−5·1	29·2	−3·46	5·47	8·05	0·30	29	80
After treatment	11·26	1·255	−3·0	111·1	+5·53	3·16	2·15	0·27	13	25

*Tanner *et al.* 1965*a,b*.

Two patients with mild proven type I hepatic GSD have grown normally with oral supplements by day alone.

The use of continuous naso-gastric infusion at night remains the optimal treatment for GSD, but we have shown that for some patients frequent oral feeds can be a satisfactory alternative.

Green, H. L., Slonim, A. F., O'Neill, J. A. Jr. & Burr, I. M. (1976). *N. Engl. J. Med.* **294**, 423.
Leonard, J. V. & Dunger, D. B. (1978). *Lancet*, ii, 1203.
Tanner, J. M., Whitehouse, R. H. & Takaishi, N. (1965*a*). *Archs Dis. Childh.* **41**, 454.
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0029–6651/79/3803–333A $00.35

The nursing couple and the anti-infective qualities of fats. By MAVIS GUNTHER, *Ember Lane, Esher, Surrey*

Some free fatty acids have anti-bacterial, anti-viral or anti-fungal activity (Galbraith *et al.* 1971; Welsh & May, 1979). Throughout lactation the human mammary gland synthesizes and secretes lauric and myristic acids and other fats from the diet as triglycerides. Lauric acid at least and some longer chain acids have been found to be active at some concentrations.

A suckled baby, as distinct from one fed human milk by bottle, receives low fat in the first part of the feed but up to 10 per cent as the breast empties. It is suggested that the higher concentration may inhibit microbial growth once it has been acted on by lipase.

It seems worthwhile considering whether the fat of the terminal part of the feed, dallying in the stomach, is made into an anti-infective provision by lipase. Lingual lipase is secreted during the activity of sucking and is active at the pH of the stomach (Hamosh, 1979). It is likely that non-nutritive sucking by the baby, apparently idly staying on the breast, serves a purpose, sending concentrated fat and more abundant lipase to the stomach. In many parts of the world, spoon-fed carbohydrate mushes, highly infected, cause gastroenteritis in infants (Mata, 1979). Possibly sources of fat in a form a baby could hold and chew once he could sit up might offer a safer way of adding to an insufficient milk supply.

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0029-6651/79/3803-333A $00.35

Studies of lipase activity in human milk. By B. HALL, D. P. R. MULLER and J. T. HARRIES, *Department of Child Health, Institute of Child Health, 30 Guilford Street, London WC1N 1EH*

Human breast milk has been shown to contain a lipase which hydrolyses long-chain triglycerides under the conditions found in the small intestine (Hernell, 1972). Pancreatic lipase activity (the principal lipolytic activity in the adult) is reduced in the neonate, increasing approximately tenfold to the adult level by the age of one year (Zoppi *et al.* 1972). Milk lipase is, therefore, likely to have an important physiological role in the hydrolysis and subsequent absorption of lipid in the neonate.

This study reports observations on milk lipase using fresh human breast milk as a source of both the enzyme and substrate rather than using skimmed milk as enzyme source and an artificial substrate as used by others (e.g. Hernell, 1972). Our studies have shown that lipase activity is very low in fresh human milk (usually $<0{\cdot}5$ μmol fatty acid released/ml milk per h), but can be induced by up to fiftyfold by physical procedures such as freezing and thawing, or sonication, or by the addition of bile salts; the glycine conjugates being four times more effective than the taurine conjugates. Purified glyconjugated bile salts were therefore routinely added to the assay system.

The fat content and lipase activity has been followed in a single individual from day 2 to 1 year of lactation. Lipase activity remained unchanged during that period (mean of thirty observations being $18{\cdot}1 \pm 4{\cdot}2$ (1 SD) μmol/ml per h) whereas the fat content rose significantly during the first two weeks of lactation (from 20 to 40 g/l) and then remained constant at 40 g/l. This suggests that the enzyme is secreted independently from the lipid.

Activities of milk lipase have been estimated in over fifty samples from twenty-three different women. Activities fell into two distinct groups with approximately 40% having high activity ($11{\cdot}7$–$27{\cdot}3$ μmol/ml per h) and the remainder low activity ($0{\cdot}4$–$9{\cdot}5$ μmol/ml per h). This corresponded to the hydrolysis of 19–53% and $0{\cdot}5$–10% milk triglyceride respectively in 2 h, which is the estimated transit time between duodenum and ileum (Henderson, 1942). If, however, milks from the low activity group were frozen and thawed or sonicated prior to assay, their activities increased to levels within the high activity group. Studies where milk serum has been separated from the milk fat and then the serum from a high activity milk recombined with the lipid from a low activity milk and vice versa, and also enzyme kinetic results, suggest that a proportion of the enzymatic activity of low activity milks is 'bound up' in some way and thus prevented from hydrolysing the substrate. The two distinct groups of lipase activity appear, therefore, to result from differences in availability of the enzymes rather than to differences in the properties of the membrane surrounding the milk fat globules.

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0029–6651/79/3803–333A $00.35

Relation between storage, cooking time and available lysine in cow peas *(Vigna sinensis).* By KAUSAR ALMAS and A. E. BENDER, *Department of Food Science and Department of Nutrition, Queen Elizabeth College, London W8*

Although the protein quality of legumes is limited by methionine plus cystine and lysine is considered of secondary importance (Hulse *et al.* 1977) the availability of lysine is reduced on cooking and can be considerably reduced on prolonged cooking.

The Table shows a loss of 10–14% of available lysine when cow peas were cooked for the minimum period. Losses are increased to 12–18% after 2 h and 18–29% after 8 h (a common practice in some communities), greater losses being found in legumes with higher content of reducing sugars.

During storage dry legumes harden and require longer cooking. The Table shows that storage at 25° lengthens the cooking time (measured by penetrometer) from 35 to 55 min after 6 months and to 62 min after 12 months. This change does not take place during storage at 4° and is not inhibited by heating (5 min at 120° in a closed container) before storage at 25°.

		Cooking time (min)		Available lysine value* g/16g N		
Storage	Storage time	Without $NaHCO_3$	With $NaHCO_3$	Raw	Cooked without $NaHCO_3$	Cooked with $NaHCO_3$
Fresh	0	35	20	5·8	5·2	5·4
25°	6 months	55	30	5·7	4·9	5·2
	12 months	62	30	5·8	4·9	5·2
4°	6 months	35	20	5·8	5·2	5·4
	12 months	35	20	5·7	5·2	5·4
Pre-heat treated	6 months	55	30	5·8	4·9	5·2
25°	12 months	62	30	5·8	4·9	5·2

*Measured by Dye-binding capacity method (Hurrell & Carpenter, 1975).

The extra time required for cooking the hardened bean results in a further although small loss of available lysine from 5·2 to 4·9 g/16 g N.

The addition of sodium bicarbonate reduced the time required to soften the beans both in freshly harvested samples (from 35 to 20 min) and in samples stored for 12 months (from 62 to 30 min). The shorter cooking time is reflected in a slightly improved available lysine content.

The shorter cooking time of 20 min is adequate to destroy trypsin inhibitors and haemagglutinins.

Since heating before storage does not prevent the hardening process it would appear that this is a chemical rather than enzymic process.

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