DIASTEMATOMYELIA

FIG. 3.—Artist's drawing: view from behind showing fused vertebrae, widespread spina bifida occulta. The large bony buttress was prolonged downwards by a cartilaginous spur. The stylized halves of the spinal cord are shown without any roots of the cauda equina for simplicity. The theca extends to the lower end of the sacrum.

(1957), Seaman and Schwartz (1958), Meacham (1967), English and Maltby (1967), Matson (1969), and James and Lassman (1972). None of these authors has described a single patient presenting so many unusual features. The most important of these features were the numerous skeletal abnormalities and an outstanding massive buttress of bone instead of the usual spur. This sessile mass of central bone split the spinal cord in two halves, which did not unite (Fig. 2, 3). Each half was tethered by fibrous bands to the dural sac which extended to the lowermost segment of the sacrum (each of which had its own motor and sensory roots). All these abnormalities produced rather surprisingly only a moderate degree of neurological disability, mostly of the left lower limb, and only a mild disturbance of bladder control.

One further unusual feature in this child was the complete absence of external (cutaneous) dysraphism.

It is suggested that the child, aged 22 months, was operated upon in time to prevent the development of major and probably permanent neurological deficit.

Summary

A 22-month-old boy was operated upon for massive diastematomyelia. His left leg was weak and his feet deformed. A large buttress of bone was found arising from the fused bodies of the 4th and 5th lumbar vertebrae. The cord was split and remained divided. The tethered halves of the cord were freed. It is concluded that this early operation prevented the development of major, irreversible neurological disability and has already resulted in a substantial recovery.

We thank Miss R. Smithies for her drawings (Fig. 2 and 3).

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Tubular reabsorption of amino acids in vitamin C deficiency

In 1957 Huisman and Jonxis described in this journal the increased amino acid excretion in the urine of children suffering from uncomplicated vitamin C deficiency. The excretion of several amino acids, including phenylalanine and tyrosine, was found to be considerably increased, whereas the plasma amino acid levels were found to be normal (Jonxis and Huisman, 1954). This points to the renal origin of the aminoaciduria in these children, and Turner and Campbell (1961) confirmed these findings. Isolated vitamin C deficiency in children is, however, uncommon. In most cases other deficiencies are found at the same time, especially that of vitamin D. Vitamin D deficiencies also cause aminoaciduria of renal origin, and it is therefore seldom possible to study

the tubular reabsorption of the different amino acids in children with a deficiency of vitamin C alone. Recently a child with isolated vitamin C deficiency was admitted to our hospital, and since to our knowledge plasma amino acid levels combined with tubular reabsorption values in vitamin C deficiency have not been reported, we are presenting the results obtained in this patient to add to our earlier observations.

Patient

A boy aged 8 months, fourth child of healthy parents, was admitted to our hospital with symptoms of irritability and loss of appetite, and he was suffering from a persistent common cold. For several months his diet had consisted almost entirely of a home-made formula of buttermilk, flour, and sugar. The buttermilk had been pasteurized before the mother cooked it again to prepare the pap. The boy had received a daily stew of meat, potatoes, and vegetables, which had been simmering for hours on an oil stove. The child had not been given any fruit or fruit juices. The vitamin C content of his diet must therefore, have been extremely low. He had been given vitamin A and D drops, and the vitamin D intake must have been approximately 600 IU/day.

The boy was thin. His length was at the 10th centile of the normal Dutch child of his age, and his weight at the 3rd centile. The infection of the upper respiratory tract was not severe. On examination a distinct rosary was found, which might have been of scorbutic or rachitic origin. The blood calcium-phosphate and alkaline-phosphatase levels were found to be normal. The x-ray of the wrist showed no signs of rickets. In the x-ray of the legs early signs of scurvy

in the form of a white line were visible on the end of the shafts of the tibia. There were, however, no subperiosteal haemorrhages.

The haemoglobin level was 12.1 g/100 ml. The vitamin C content of the blood was found to be very low. Three determinations showed values of less than 0.1 mg vitamin C/100 ml.

Vitamin C (150 mg/day) was given and 4 days later the concentration of vitamin C in the blood had risen to 1 mg/100 ml; 9 days later it had reached 1.4 mg/100 ml. The general condition of the child improved rapidly. His appetite increased and his irritability disappeared. He gained weight rapidly.

In view of the history, the low vitamin C blood levels, and the improvement after vitamin C administration on one hand and the normal vitamin D intake, the normal calcium, phosphate, and alkaline phosphatase levels of the blood on the other, the diagnosis of vitamin C deficiency, uncomplicated by hypovitaminosis D, was made.

Method for measuring tubular reabsorption of amino acids

Tubular reabsorption of amino acids was measured on two occasions: first before the daily administration of 150 mg vitamin C was started, and again after 4 weeks of treatment.

The percentage of reabsorption of each amino acid was calculated from the equation

$$100 \times \left(1 - \frac{\text{clearance of one amino acid}}{\text{clearance of creatinine}} \right)$$

The clearance studies were performed over a 6-hour period in the fasting patient with liberal allowance of fluids. The creatinine clearance was assumed to be proportional to the glomerular filtration rate. Amino

TABLE

Percentage tubular reabsorption of different amino acids before and 4 weeks after vitamin C administration

Amino acid	Before vitamin C			4 weeks after vitamin C		
	Concentration in plasma (μmol/ml)	Concentration in urine (μmol/ml)	Tubular reabsorption (%)	Concentration in plasma (μmol/ml)	Concentration in urine (μmol/ml)	Tubular reabsorption (%)
Ornithine	0.04	0.02	97.5	0.04	0.02	99.6
Lysine	0.10	0.73	63.5	0.09	0.22	97.9
Arginine	0.05	0.01	99.0	0.05	—	—
Histidine	0.06	1.93	<50.0	0.08	0.85	91.1
Threonine	0.07	0.13	90.7	0.04	0.29	93.9
Serine	0.09	0.86	52.2	0.10	0.15	98.7
Asparagine-glutamine	0.39	1.71	78.1	0.41	0.70	98.6
Proline	0.24	0.11	97.7	0.28	0.00	—
Glycine	0.13	3.33	<50.0	0.19	2.54	88.7
Alanine	0.14	0.51	81.7	0.41	0.72	98.5
Valine	0.26	0.14	97.3	0.12	0.04	99.7
Methionine	0.01	0.12	<50.0	0.02	0.06	97.5
Isoleucine	0.08	0.15	90.6	0.04	0.02	99.6
Leucine	0.13	0.10	96.2	0.07	0.10	98.8
Tyrosine	0.06	0.34	71.7	0.07	0.20	97.6
Phenylalanine	0.05	0.27	73.0	0.07	0.15	98.2
Glutamic acid	0.09	0.08	95.6	0.15	0.73	95.9

acids in plasma and urine were estimated by the photoelectric ninhydrin method of Moore and Stein (1954) with the use of an automatic Spinco Beckman Amino Acid Analyzer model 120B. Plasma was deproteinized with picric acid (Stein and Moore, 1954). Creatinine in plasma and urine was determined spectrophotometrically. After treatment with franconit KL suspension, the absorbance in the presence of picrate was measured at 500 nm (Gorter and De Graaff, 1955).

Results

The concentrations of amino acids in plasma and urine and their tubular reabsorption before and after treatment are set out in the Table.

The Table makes it clear that on admission, before vitamin C, there was conspicuous decreased reabsorption of various amino acids, particularly of histidine, methionine, lysine, threonine, isoleucine, phenylalanine, tyrosine, glycine, serine, alanine, and asparagine + glutamine. There was only a slight decrease in reabsorption of arginine, valine, leucine, and glutamic acid. Small amounts of proline in free form were detectable in the urine.

Four weeks after the start of the vitamin C administration, tubular reabsorption of most of the amino acids had reached normal values (Table). That of histidine, threonine, glycine, phenylalanine, and tyrosine had not completely returned to normal.

Discussion

It is clear from the results obtained on the present study that in vitamin C deficiency the tubular reabsorption of the aromatic amino acids, phenylalanine and tyrosine, is reduced, whereas in vitamin D deficiency it is not (Huisman, 1954). As in our earlier investigations it was found that it takes many weeks, even with ample vitamin C administration, before the tubular reabsorption of all amino acids reaches normal values.

Summary

Tubular reabsorption of amino acids has been measured in a boy aged 8 months suffering from uncomplicated vitamin C deficiency, both before and after the administration of vitamin C.

Before treatment there was a decreased reabsorption of a number of amino acids, particularly of histidine, glycine, methionine, and also of tyrosine and phenylalanine.

Four weeks after the start of the treatment with 150 mg vitamin C per day, tubular reabsorption of most of the amino acids had reached normal values, but that of threonine, glycine, and tyrosine had not completely returned to normal.

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Apnoeic attacks in the newborn treated with aminophylline

It has been postulated that a significant cause of brain damage in surviving premature babies is hypoxia secondary to recurrent apnoea (McDonald, 1963; Bacola *et al.*, 1966; Daily, Klaus, and Meyer, 1969). Present methods of managing such episodes consist of external stimulation, administration of oxygen, and assisted ventilation. However successful such measures are they do not prevent further apnoeic attacks which may be serious to the baby. It was this thought that suggested that aminophylline, by virtue of its direct action on the respiratory and vasomotor centres and on the myocardium, might be useful in the treatment and prevention of prolonged apnoeic attacks in the small baby.

The present preliminary paper describes clinical observations in 10 babies.

Material and methods

Ten preterm babies suffering from idiopathic respiratory distress syndrome (RDS) were studied. Criteria for diagnosis were those of Reynolds, Robertson, and Wigglesworth (1968). An apnoeic attack was defined as a period of nonbreathing, usually lasting more than 30 seconds, during which cyanosis and slowing of the heart rate occurred. Hourly heart and respiratory rates were recorded. Routine care of babies consisted