Details of five patients on home parenteral nutrition for intestinal failure

Case No	Age (years)	Sex	Disease and nutritional problem	Time on parenteral nutrition (months)	Time at home on parenteral nutrition (months)	Weight at start of treatment (kg)	Weight at end of treatment (kg)
1	18	F	Polyarteritis nodosa. Mesenteric vascular occlusion. Short-bowel syndrome	5½	3	28	39
2	21	M	Intestinal motility disorder. Small and large bowel resection. Short-bowel syndrome	13*	9*	45	56
3	36	M	Crohn's disease, multiple entero-cutaneous fistulae	10	3	25	47
4	49	M	Mesenteric ischaemia, intestinal fistula, short-bowel syndrome	5	1½	44	52.5
5	34	M	Crohn's disease, multiple small bowel resection, short-bowel syndrome	5	1½	45	57

*Parenteral nutrition continuing.

biochemistry whenever feeding by mouth was reintroduced. All five patients were referred to the hospital. Treatment began with a vigorous programme of resuscitation followed by parenteral nutrition, initially through a silicone elastomer catheter inserted into the right atrium by infraclavicular puncture of the subclavian vein. The feeding regimen was calculated in standard fashion from the patient's measured nitrogen losses and estimated energy requirements. As soon as long-term parenteral nutrition appeared to be necessary the subclavian catheter was changed to one more suited for prolonged intermittent feeding. We have found the Broviac catheter (Evergreen Medical Products, USA) the most suitable. It is inserted into the cephalic vein in the deltopectoral groove and the tip positioned in the right atrium. The proximal end of the catheter is tunnelled beneath the skin to emerge below and medial to the ipsilateral nipple, where the hub is easily accessible to the patient. Once nitrogen and energy requirements have stabilised the infusion system is changed to the "big bag" technique. In this the day's nutrients are placed under sterile conditions into a three-litre bag (Viaflex, Travenol Ltd) and given over the whole or part of the day.

The patients were taught to infuse the nutrients themselves and (between infusions) to fill the catheter with heparinised saline (Heparin lock). When home parenteral nutrition was expected to be for only a few weeks the patients were not taught to use an infusion pump. Instead they were taught to rely on overnight gravity infusion from the three-litre bag controlled by a metal gate clamp. When treatment seemed likely to be needed for several months patients were taught to use a constant infusion pump (Imed 922H, Tekmar Medical Ltd). We gave them a manual, written by ourselves, which reminded them of the technique and how to minimise complications. The staff of the departments of medicine and surgery provided 24-hour cover for emergencies on a rota basis. Complications were remarkably few. Catheter sepsis caused septicaemia in two patients. No case of catheter sepsis occurred in a patient at home. Two Broviac catheters that fractured were repaired successfully with the manufacturer's repair kit.

All the patients lived and four of the five returned to feeding by mouth and are in good health. The results are summarised in the table. A period of 15 weeks off parenteral nutrition in one patient was associated with a profound fall in weight and the onset of listlessness. He rapidly improved when home parenteral nutrition was reintroduced. He will probably have to remain on it indefinitely.

Comment

Only a few patients with nutritional problems require this type of treatment, and hardly ever permanently. The main indications are the short-bowel syndrome resulting from extensive resection or severe inflammatory bowel disease, complicated in some cases by fistula. In the former the residual bowel usually compensates enough for feeding by mouth to be resumed, while in the latter the disease often subsides with bowel rest and appropriate medication. Parenteral nutrition at home adds a new dimension to the treatment of these patients. It is analogous to the early days of home renal dialysis for renal failure. Our results show that the treatment is effective. The problem is the cost, which we calculate exceeds £15 000 a year for each patient. Nevertheless, we believe that it should be available in a few centres.

This work was supported in part by grants from the North Western Regional Health Authority and the Sheila Dawber Research Fund of the

Salford Area Health Authority. We thank the consultants who referred their patients and the pharmacy staff of the Park Hospital, Davyhulme, for their services.

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(Accepted 15 February 1980)

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Salivary duct calculi presenting as trismus in a child

The submandibular gland and duct are the commonest sites for salivary calculi, which are usually found in adults with good oral hygiene. The condition is rare in children and thus may be overlooked. The diagnosis can usually be made on the typical history of painful swelling in the submandibular region during eating. I report a case in a young girl with salivary duct calculi who presented with acute trismus, making clinical assessment difficult.

Case report

A 9-year-old Caucasian girl presented with a 24-hour history of pain in the right side of the neck and a visible swelling in the same region. On the day of admission to hospital she had not spoken and had been unable to drink, holding her mouth slightly open and dribbling saliva. She was afebrile and had oedema, without erythema, of the right side of the face and neck. Below the angle of the mandible on the right side a hard, fixed, acutely tender swelling was palpable. Examination of the mouth was extremely difficult but suggested there was a calculus in the region of the right submandibular duct orifice. Haemoglobin concentration was 12.9 g/dl and white cell count $16.9 \times 10^9/l$ ($16\,900/mm^3$) with a 90% polymorphonuclear neutrophil leucocytosis. X-ray films of the submandibular area, taken with great difficulty, showed a small (3 mm diameter) calculus in the region of the right submandibular duct orifice. Examination under anaesthesia confirmed that there was a calculus 5 mm proximal to the right submandibular duct orifice. A direct incision into the duct released the calculus and a small amount of sterile pus. Over the next 48 hours the trismus and oedema resolved completely and the right submandibular gland became palpable.

Comment

Submandibular calculi are very rare in children. Only a handful of cases have been reported since 1895. Doku and Berkman¹ reviewed the first major series of 11 cases in children under the age of 16 years and added one case. Several cases have been described since in otherwise healthy children.²⁻⁴ Stones are thought to form because of deposits of calcium salts around a central nidus, such as desquamated epithelial cells, bacteria, or a foreign body. Perhaps surprisingly, only one case has been reported in a child with cystic fibrosis, where there is apt to be increased calculus formation owing to a high ratio of calcium to phosphorus in the saliva and increased turbidity of the saliva with a tendency to form crystals.⁵

Although the condition is unusual in children, there is often a history of intermittent, submandibular swelling, but the diagnosis is often overlooked.² There have been no reports of children with submandibular calculi presenting acutely with cervicofacial swelling

and trismus. As an acute presentation is unexpected therefore, clinical assessment in this case was difficult.

A diagnosis of salivary duct calculi should always be considered in children when there is submandibular swelling and the appearances are uncharacteristic of suppurating cervical nodes. In this way inappropriate exploration of the cervical swelling will be avoided.

I thank Professor R Y Calne for permission to report this case.

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(Accepted 1 March 1980)

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Porphyria cutanea tarda during maintenance haemodialysis

Skin lesions indistinguishable from those of porphyria cutanea tarda occur in about 1% of patients on maintenance haemodialysis.¹ Usually there is no evidence of abnormal porphyrin metabolism,¹ but three cases in which true porphyria cutanea tarda developed during chronic haemodialysis have been reported.² The primary enzyme defect is a decrease in hepatic uroporphyrinogen decarboxylase activity.³ We describe a patient with an inherited defect of this enzyme who developed the condition while receiving chronic haemodialysis.

Case report

A 16-year-old boy with chronic glomerulonephritis was started on chronic haemodialysis in August 1975. In August 1976 he noticed hyperpigmentation, skin fragility, and multiple bullae on his face and backs of his hands. At this time urine output was very low and the diagnosis of porphyria cutanea tarda was established by measuring porphyrins in plasma (table) and faeces (isocoproporphyrin:coproporphyrin ratio 1.2; normal less than 0.1). There was no family history of porphyria and no evidence of liver disease, and he was teetotal. Since August 1975 he had received aluminium hydroxide, methyl dopa, propranolol, vitamin complex, iron-dextran (100 mg monthly), and six units of blood. From October 1976 dialysis was performed with a Haemodialyzer RP 6 HP using an acrylonitrile membrane (1.03 m²; 30 µm), which reduced the plasma porphyrin concentration (table). The calculated dialysance for uroporphyrin was 79.7 ml/min. Skin fragility persisted and treatment with chloroquine (75 mg weekly for two months), which worsened his hypertension, and cholestyramine was unsuccessful. In January 1979 serum ferritin was 878 µg/l (normal 10-250 µg/l).

The patient's uroporphyrinogen decarboxylase activity, measured in haemolysed whole blood after activation with dithiothreitol, using penta-

carboxylate porphyrinogen III as substrate, was 17.2 pmol coproporphyrin/min/mg haemoglobin (mean and range for 18 normal subjects: 24.8, 20.3-32.6 pmol/min/mg). His sister and mother had similar low enzyme activities, 17.1 and 17.2 pmol/min/mg respectively, whereas those of his brother (35.6 pmol/min/mg) and father (28.6 pmol/min/mg) were not low. Total urinary and faecal porphyrin excretion was normal in these relatives but his sister and mother showed increased urinary excretion of heptacarboxylic porphyrin, indicating subclinical porphyria cutanea tarda.

Comment

The occurrence of porphyria cutanea tarda during chronic haemodialysis for chronic renal failure is rare and the relation between these disorders is uncertain. In previous cases the cause of the uroporphyrinogen decarboxylase defect has not been determined.² In our patient, as in a proportion of other patients with porphyria cutanea tarda,⁴ it appears to have been caused by inheritance of an autosomal dominant gene, the effect of which is expressed in the liver as well as in red cells.³ The enzyme defect, therefore, must have been present before the onset of renal disease. Thus chronic haemodialysis, like alcoholic liver disease with siderosis,³ may be associated with porphyria cutanea tarda because it is capable of making an otherwise occult enzyme defect clinically manifest. Why haemodialysis should have this effect is not clear. The iron overload that is common in patients undergoing haemodialysis⁵ may be a factor in precipitating the disease.^{2,3} Our patient received iron-dextran, and iron overload was certainly present after the onset of the porphyria. Alternatively, patients undergoing haemodialysis may produce or be subjected to compounds that affect hepatic haem synthesis.

Haemodialysis almost halved the concentration of porphyrins in the plasma of our patient, with nearly quantitative recovery in the dialysate and with little change in the porphyrin profile (table). In contrast, Poh-Fitzpatrick *et al*² were unable to lower plasma porphyrins by haemodialysis. This difference may be explained by our use of a high-permeability membrane. Despite regular haemodialysis, plasma concentrations remained high, and effective treatment of porphyria cutanea tarda in these patients is difficult. Our patient reacted adversely to a low dosage of chloroquine and depletion of iron stores by repeated venesection cannot be justified.

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(Accepted 8 February 1980)

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Porphyrin concentrations in plasma and dialysate in a patient who developed porphyria cutanea tarda during maintenance haemodialysis

Porphyrin	Plasma		Dialysate†
	Before haemodialysis	After haemodialysis	
Total porphyrin (nmol/l)*	1630	990	25
Uroporphyrin (%)	61	61	54
Heptacarboxylic porphyrin (%)	33	33	32
Hexacarboxylic porphyrin (%)	6	6	7
Pentacarboxylic porphyrin (%)	..	..	5
Isocoproporphyrin (%)	..	..	<1
Coproporphyrin (%)	..	..	1

*Normal range <5 nmol/l.

†79 l obtained over 5 hours.

Conversion: SI to traditional units

Total porphyrin (as coproporphyrin): 1.0 nmol/l ≈ 0.065 µg/100 ml.

Correction

Alpha-chain disease diagnosed by rectal biopsy

In the short report by Dr J M Rhodes *et al* (12 April, p 1043) the antiserum is stated to have been supplied by Mercia Diagnostics. This has now amalgamated with Brocades (GB) Ltd and for the last two years has been trading under the name of Mercia-Brocades Ltd.