Commentary

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Nephrologists will be intrigued by this case history, and as the pathological findings conflict with current beliefs about Henoch-Schönlein nephritis some will ask whether this child really had Henoch-Schönlein syndrome. The clinical features of the illness, which followed an upper respiratory tract infection and which involved the alimentary system, the joints, the kidneys and the skin, seem typical but the pathognomic feature of Henoch-Schönlein syndrome is the rash, for it is the characteristics of the rash that allow the clinical diagnosis. It is unfortunate that no photograph of the rash is available, but understandable since the authors had no particular reason to take a photograph at the time when they believed this to be a straightforward case of Henoch-Schönlein syndrome. We have to accept the accuracy of their description and the validity of these experienced clinicians saying that it was characteristic of Henoch-Schönlein purpura.

Thirty years ago β haemolytic streptococcal infections were a common precipitant of Henoch-Schönlein purpura; they are a less common cause now as the incidence of streptococcal infection in the industrialised countries has decreased. This child had such an infection and an appropriate antistreptolysin O titre response. But, the ensuing illness was not a classic post-streptococcal acute glomerulonephritis: serum C3 complement values remained normal and the general features of the ensuing illness, particularly the rash, were characteristic of Henoch-Schönlein syndrome. The twist in this tale is

that although the nephritis that accompanied the Henoch-Schönlein syndrome was clinically appropriate, the renal biopsy did not show the one feature that has come to be regarded as pathognomic of Henoch-Schönlein nephritis. For though on light microscopy there may be a wide variety of different histological appearances, most of them variations of a diffuse mesangial glomerulonephritis, the pathognomic feature of Henoch-Schönlein nephritis is the immunofluorescent finding of IgA deposits in the mesangium. The constancy of these deposits together with the observation that some children with Henoch-Schönlein syndrome have raised serum IgA, and an increased proportion of IgA bearing lymphocytes, as well as the finding of IgA immune complexes has led to a widespread acceptance that IgA immune complexes are a cause of the nephritis. The similarity of the immunofluorescent findings and the immunological observations to those of Berger disease (mesangial IgA nephropathy) which often presents with haematuria, has led to the suggestion that Henoch-Schönlein nephritis and Berger disease are the same and that people with Berger disease could be considered as having Henoch-Schönlein syndrome without the rash.

Now this report seems to suggest that Henoch-Schönlein syndrome and a nephritis may occur in someone with specific deficit of IgA and that the features, including the nephritis, may occur in the absence of IgA deposits. Therefore, some will wonder whether the IgA abnormalities of Henoch-Schönlein syndrome are merely epiphenomena and irrelevant to the aetiology; others will question whether this girl did have Henoch-Schönlein syndrome—and many of us will wish that we had had the opportunity to examine the child and see the rash ourselves when it first occurred.

Symptomatic vitamin E deficiency in cystic fibrosis

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SUMMARY A girl with cystic fibrosis who developed a neurological syndrome probably secondary to vitamin E deficiency at the age of 10 years is described. The severity of the deficiency and the early development of neurological features probably result from reduced intraluminal bile salt concentrations in addition to the pancreatic insufficiency.

Reduced serum concentrations of vitamin E have been documented in cystic fibrosis,¹ and there is now some evidence to indicate that vitamin E is necessary for normal neurological function.² Neuropathological changes³ and a spinocerebellar syndrome have been described in adults with cystic fibrosis and vitamin E deficiency.^{4 5}

We report a girl with cystic fibrosis who had undetectable serum concentrations of vitamin E and

developed a spinocerebellar disorder at the age of 10 years, having had gastrointestinal problems from an early age but few associated pulmonary symptoms.

Case report

This girl was born at 38 weeks' gestation weighing 2.97 kg. Intestinal obstruction developed on the first day and at operation a very dilated jejunum was found, descending down to a volvulus in the mesentery. The ileum was atretic in two places and the appearances were compatible with meconium ileus. Resection of 47 cm of bowel was necessary and a defunctioning 'koop' type ileostomy was fashioned. It was closed at 3 months. Sweat tests after the operation confirmed the diagnosis of cystic fibrosis.

At 3 years of age intestinal obstruction occurred again and a second laparotomy was necessary. The ileum was obstructed by adhesions at the site of the previous ileostomy, and bowel proximal to the obstruction was dilated and hypertrophied. Adhesions were divided, the redundant ileostomy stump was resected, and the appendix removed.

During the following years chest symptoms were minimal but occasional episodes of abdominal pain and vomiting occurred. A barium study (at 8 years) showed a grossly distended segment of jejunum. The patient's weight fell from the 50th to the 25th centile.

At 9 years of age a pre-existing right exotropia became more evident and horizontal nystagmus was noted. An assessment at a neurological department was otherwise normal. An operation for exotropia was performed. In the following year neurological deterioration occurred. On examination at 10 years of age the findings were horizontal nystagmus, noticeable peripheral and truncal disequilibrium, a peripheral neuropathy with wasting and weakness of distal muscles especially in the lower legs, areflexia, and impaired posterior column sensation. Nerve conduction studies showed mild fall out of both sensory and motor fibres in upper and lower limbs. The electromyogram showed motor unit action potentials of noticeably increased complexity and instability, with jitter. The duration was increased and the interference pattern dense because of excessive polyphasia. The abnormalities were neurogenic and indicated vigorous terminal nerve fibre sprouting.

Sural nerve biopsy did not show degenerative change. Occasional foci of acid phosphatase activity were present and several cells contained autofluorescent pigment. The muscle biopsy showed mild denervation with regeneration and strongly autofluorescent pigment in many fibres consistent

with vitamin E deficiency. Serum vitamin E was undetectable on a number of separate blood samples. A load of 1 g fat soluble vitamin E preparation was given and over the following 24 hours serum concentrations were just detectable. Details of subsequent vitamin E estimations and of supplementation of a water miscible preparation are also given (see Table).

Within six weeks of these investigations a massive rectal bleed occurred and on hospital admission the haemoglobin concentration was 5.5 g/dl. No defect of clotting was found, although two weeks previously prothrombin time had been prolonged and konakion (15 mg daily) was prescribed. Transfusion of 9 units of blood was necessary. Physical examination of the abdomen was normal and endoscopy did not show varices. At laparotomy the residual bowel consisted of 46 cm of jejunum and 60 cm of ileum plus the colon. Two ulcers with adherent clot were present at the site of the old anastomosis and a further ulcer in the dilated bowel. Because of the degree of dilatation it was necessary to resect 20 cm of jejunum and a short length of dilated ileum. The liver was shrunken with a multinodular surface and the histology showed biliary fibrosis.

Table Serum vitamin E concentrations and supplementation

Date	Serum vitamin E μmol/l)	Vitamin E supplementation
21 5 82	Undetectable	Nil
18 6 82	0hr Undetectable 2hr Undetectable 4hr Undetectable 6hr Undetectable 8hr Undetectable 24hr 2-3	Oral load of 1 g fat soluble preparation*
19 7 82	3-2	Oral water miscible preparation† (3g/day) from 7 82
8 9 82	12-2	Oral water miscible preparation (3g/day) from 7 82
8 10 82	7-6	Intramuscular preparation† (100mg/wk) from 10 9 82
22 11 82	9-0	Intramuscular preparation† (100mg/wk) from 10 9 82
7 1 83	9-4	Intramuscular preparation† (100mg/wk) from 10 9 82
7 3 83	22-7	Oral water miscible preparation† (3g/day) from 7 1 83
15 4 83	20-7	Oral water miscible preparation† (3g/day) from 7 1 83
25 4 83	17-8	Oral water miscible preparation† (3g/day) from 7 1 83
18 7 83	11-4	Oral water miscible preparation† (3g/day) from 7 1 83
5 12 83	14-4	Oral water miscible preparation† (3g/day) from 7 1 83
Normal range	11.5-35.0	

*Ephynal.

†DL α-tocopheryl acetate.

Conversion—SI to traditional units: vitamin E 1 μmol/l=41.6 μg/100 ml.

At this time the patient's serum lactate, pyruvate, folate, red blood cell folate serum B₁₂, zinc, copper, cholesterol, and the lipoprotein values were all normal. Activities of alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, and γ -glutamyltransferase were raised.

After supplementation with vitamin E for 6 months the nystagmus had resolved. After 18 months of treatment the ataxia had lessened, hand coordination was improved, and Romberg's sign was negative. She was still areflexic.

Discussion

A neurological syndrome secondary to vitamin E deficiency which has responded to vitamin E supplements has been reported in adults with cystic fibrosis.^{4,5} Our patient resembled these previous reports but presented much earlier. This was presumably because of impaired bile salt solubility of the vitamin in addition to pancreatic insufficiency. Luminal bile salt concentrations were likely to have been reduced in our patient. Firstly she had multiple resections of the ileum, the site of the active reabsorption of bile salts; secondly, the situation would be exacerbated by stasis in the dilated jejunum producing a 'blind loop' effect; and finally biliary fibrosis would result in impaired biliary excretion.

Although this neurological complication seems to be relatively uncommon in cystic fibrosis, it is important to be aware of it. A careful neurological examination of these patients for evidence of cerebellar, posterior column, and peripheral nerve dysfunction is emphasised. Monitoring of serum vitamin E concentrations in all patients would be ideal and is recommended if possible, yet limited laboratory facilities for measurement make this

impractical for many paediatricians. In patients with cystic fibrosis, 50 to 400 mg daily of a water miscible product has generally resulted in normal plasma and tissue levels of α -tocopherol.^{1,3,6} In the absence of laboratory facilities regular vitamin E supplementation of a water miscible preparation of vitamin E is recommended in a dosage of 100 to 200 mg daily. If there is severe malabsorption resulting in neurological dysfunction this oral preparation may not give adequate plasma concentrations and intramuscular vitamin E (50 to 100 mg/week) may be required.⁴

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Loss of breast milk nutrients during tube feeding

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SUMMARY Nutrient losses from banked breast milk during tube feeding were determined from laboratory models. Large fat losses occurred, continuous feeding giving rise to significantly greater losses than bolus feeding. More fat loss occurred with low flow rates and there were also significant protein losses.

The effects of delivery and handling on bank breast milk nutrients have commanded much less attention than the nutrient properties themselves. The composition of milk in the feeding syringe may not be the same as that which enters the infant's gastrointestinal tract; significant energy losses have been shown to be lost during the passage down feeding tubes.^{1,2} This study compares fat losses in con-