LETTERS TO THE EDITOR

Intestinal lymphangiectasia and protein losing enteropathy responding to small bowel resection

EDITOR,—Primary intestinal lymphatic obstruction with protein losing enteropathy (PLE) may be associated with other lymphatic drainage abnormalities outside the gastrointestinal tract. The pathogenesis, classification, management, and prognosis of these conditions continues to be discussed.^{1,2} We present a patient with PLE whose disorder has been followed up for nearly 20 years.

The patient, was aged 13 months when he originally presented to the Paediatric University Hospital Centre in Rijeka, Croatia with watery diarrhoea and generalised oedema. The laboratory features, including a low serum total protein and albumin, were consistent with PLE. He was treated with a high protein and low fat diet supplemented with medium chain triglycerides.

After initial improvement of his symptoms, oedema, and hypoproteinaemia he relapsed at the age of 3 years, when severe oedema of his left leg was also observed. Barium radiography of the stomach and small bowel was normal, but consent for small bowel biopsy was declined. He was treated with frequent transfusions of albumin and diuretics in addition to the dietary regimen. His symptoms improved but his left leg remained swollen.

Subsequently his course was characterised by relapses with watery diarrhoea and generalised oedema, even when he apparently complied with his diet. There was persisting hypoproteinaemia, hypoalbuminaemia, hypoglobulinaemia, and lymphopenia. At 6 years small bowel radiography showed thickening of mucosal folds and hyperfoculation of barium. A biopsy specimen of the small bowel did not show dilated lymphatics, but a later specimen at 10 years, done at different levels, confirmed dilated lymphatics.

At 14 years, when he was receiving transfusions of albumin weekly, he was admitted to St Thomas's Hospital, London for reinvestigation. A ⁵¹Cr labelled albumin study suggested PLE (18% recovery), and small bowel radiography showed an abnormal mucosal pattern throughout the entire small bowel. Lymphangiography showed normal right sided iliac and intra-abdominal lymphatics, a normal thoracic duct, but significant lymphatic obliteration in both legs.

In view of the severe and uncontrollable protein loss, surgical resection of the small bowel was carried out. At laparotomy the small bowel was of normal size, but the mesenteric lymph nodes were hard and there were many dilated lymphatics on the surface of the bowel. Injection of a dye (blue violet) into one of these lymphatics showed the dye running across the surface of the bowel with none of it transversing the mesenteric lymphatics or reaching the mesenteric lymph nodes. The small bowel was 671 cm long. Approximately 305 cm, which was palpably thicker than the rest, was resected and an end

to end anastomosis was carried out. When the resected bowel was opened up it had the typical appearance of lymphatic obstruction, a white cobblestone appearance.

After surgery, symptoms of PLE abated, intestinal protein loss was reduced, and his serum proteins and immunoglobulins stabilised to a level just below the normal range. He was last reviewed in January 1996 at the age of 20 years when he was on a normal diet, and growing normally. He remained free of symptoms, and there was no generalised oedema. However, his left leg was grossly swollen (elephantiasis) and also his right leg was swollen.

This patient illustrates that in resistant cases of PLE small bowel resection can control enteric protein loss and the resulting symptoms. However, the prognosis is uncertain, and we need to learn more about the pathogenesis and evolution of the underlying lymphatic abnormality in this group of disorders.³

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Cystic fibrosis in Down's syndrome—diagnostic pitfalls and implications for the clinician

EDITOR,—Cystic fibrosis and Down's syndrome are two frequent genetic diseases (1:2000 and 1:600).¹ However, few reports describe their coexistence.^{2,3}

We therefore report on a female patient (birth weight 2150 g at 37 weeks' gestation, maternal age 40 years, fifth child) born with the typical clinical features of trisomy 21. Chromosomal analysis confirmed the diagnosis. A positive family history of cystic fibrosis with an affected sister who died at the age of 14 years two years earlier, gave her a 25% risk for this disease too. A pathological sweat test (sodium chloride 180 mmol/l) and homozygosity for the mutation delta F508 on chromosome 7 confirmed the diagnosis of cystic fibrosis. Clinically she exhibited marked failure to thrive, malnutrition, signs of maldigestion, and recurrent pneumonia. Although the patient received intense treatment including antibiotics, high energy nutrition and enzyme replacement, her condition deteriorated gradually. She died at the age of 6 months.

Our patient is the first reported so far with DNA analysis confirming both diagnoses, cystic fibrosis and trisomy 21. Diagnosis of cystic fibrosis in the four patients reported previously relied on sweat tests and clinical features with respect to cystic fibrosis. However, abnormalities of osmolality and sodium chloride concentrations of sweat were reported in trisomy 21, thus rendering the validity of sweat tests questionable.⁴ Also, as

clinical symptoms such as failure to thrive, malnutrition, signs of maldigestion, and recurrent pneumonia could be easily attributed to either disease, clinical diagnosis is hampered. It is important to consider the diagnosis of cystic fibrosis in children with Down's syndrome and the above clinical signs. DNA analysis is then essential in confirming or rejecting a diagnosis of cystic fibrosis.

The risk for the occurrence of trisomy 21 and cystic fibrosis in the general population is 1:1 200 000 (1:600 × 1:2000). In a family with one child suffering from cystic fibrosis the risk for siblings is increased (1:600 × 1:4 = 1:2400) and rises further with maternal age. Therefore, the coexistence of cystic fibrosis and trisomy 21 should be more frequent.

The prognosis in this combined disease seems to be very poor. None of the affected children reported survived longer than our patient.

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Nutritional impact of antipseudomonas intravenous antibiotic courses in cystic fibrosis

EDITOR,—In their study of cystic fibrosis patients who were nutritionally assessed at the start and end of a 14 day period of home intravenous antibiotic treatment for chest disease, Vic et al concluded that increased weight was a result of increased fat storage.¹ While this may in part be true, their methodology dictates caution in drawing conclusions.

They used bioelectrical impedance analysis (BIA) at 50 Hz to estimate body water, and four site skinfold thickness to estimate the percentage body fat in their patients. From these data and weights at day 1 and day 14, they estimated changes in fat mass by two indirect methods. The validity of this approach rests on changes in body composition being reflected in changes in BIA and skinfolds respectively. The authors detail neither the changes over their study period in BIA and skinfold thickness, nor the variability of the skinfold measurements. These data are necessary to support their assertion that body fat stores have changed.

In the course of nutritional rehabilitation the initial weight gain is often a result of

increased total body water. The apparent increases in body fat in this study may be an artefact resulting from the reliance of both indirect methods on body weight at some point in the calculations. Azcue *et al* who performed a validation of BIA in patients with cystic fibrosis found a standard error of the estimate of total body water of 3.19 litres.² Such a large standard error makes the interpretation of short term changes in BIA estimated body water difficult. Changes in total body water content may be better estimated from swept frequency BIA, but whether single or swept frequency techniques are used there is a need to test the assumption that short term changes in BIA reflect changes in labelled water ($H_2^{18}O$) distribution. We would not therefore support the routine use of BIA in monitoring nutrition in individual cystic fibrosis patients. In experimental studies the current paper would justify the interchangeable use of BIA and anthropometry to estimate body fat when an underestimate of 0.7 kg and an overestimate of 1.1 kg is not of biological significance.

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Asthma—time for a change of name?

EDITOR,—Professor Silverman and Dr Wilson propose to use the diagnosis 'asthma' in children only when it is accompanied by a description of qualifying features such as the pattern and severity of the disease and possible causative or aggravating factors.¹ As much as their suggestion deserves endorsement, both from a clinical and a scientific point of view, history has taught that getting physicians to accept such an approach may be very difficult, or even impossible.

In 1961, Professor Orie in Groningen considered that it is often very difficult to separate asthma from chronic obstructive pulmonary disease (COPD) in adults. Moreover, the two diagnoses could be encountered in the same patient over time. Finally, the approach to treatment to both disorders was often the same.² He, therefore, proposed to use an umbrella term, chronic non-specific lung disease (or CARA in Dutch) for patients with asthma or COPD, provided it was accompanied by defining criteria (at that time considered to be allergy, airways hyper-responsiveness, and reversibility of airways obstruction). He hoped that this approach would improve communication between doctors and, eventually, patient care.

Unfortunately, he was wrong. The 'Dutch hypothesis' was strongly opposed, especially from the UK and the USA, by physicians who found it incomprehensible that the Dutch would lump together two diseases so obviously different as atopic asthma and chronic bronchitis.³ In practice, it turned out to be

impossible to get doctors to use comprehensive defining criteria to describe individual patients, simply because doctors want simple diagnostic labels that can be communicated quickly.⁴ This has led to the unfortunate practice of giving a diagnosis of 'CARA' to any patient with cough and/or wheeze in our country for a number of years. Over the past five years or so the Dutch have learned their lesson and the term 'CARA' is being abandoned now, being replaced by more traditional diagnoses such as asthma and COPD.

The parallel to childhood asthma as illustrated by Silverman and Wilson is obvious. Their suggestion of using 'asthma' as an umbrella term only when accompanied by a description of the disease in more detail, logical and balanced as it is, is going to be difficult to implement in practice. It requires extra work and discipline from busy practising physicians who prefer economic communication in short labels.

Only if practitioners can be convinced that the approach proposed by Silverman and Wilson is going to be useful and meaningful in patient care will it have a chance of succeeding.

I sincerely hope that the time is more right for asthma with descriptive features in 1997 than it was in 1961. I wish Silverman and Wilson much support from paediatricians. They can count on the Dutch!

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Colic, wheeze, croup, allergy, and asthma

EDITOR,—Paediatrics is full of terms such as these, which can mean many things to a wide range of health professionals and parents. Professor Silverman and Dr Wilson elegantly describe the problems involved in attempting to define paediatric asthma.¹ As long as our understanding of the range of pathological processes resulting in breathlessness and wheeze remains incomplete, these problems will persist.

In clinical practice an explanation of the probable cause of symptoms, the usefulness of treatment, and the likely outlook is helpful even if there is no name to accompany it. Parents should be entitled to this type of approach, rather than a diagnosis that at present has no agreed meaning.

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Adolescent health services

EDITOR,—We agree with Oppong-Diseng and Heycock that adolescents have clear views regarding the nature of services which they would like to see provided.¹ We know that adolescents worry about their health and often feel anxious about consulting their own family doctor. Confidentiality is a major issue and previous work has shown that many adolescents write anonymously to "agony aunts" to have their questions answered.² The advice columnist from *MIZZ* magazine receives up to 400 letters per week from adolescents whom she says "feel they do not have anyone else to turn to for help", and *Sugar* magazine gets up to 300 letters per month. Girls write mainly about their periods, breast size and shape, body image, and with concerns about eating disorders. Both males and females worry about sexual orientation, HIV, acne, and dying and bereavement.

To try and answer some of our adolescent questions locally we liaised with form tutors to provide health related seminars. We asked pupils to anonymously contribute questions for discussion. Questions were wide ranging, for example, "How big is the average male penis?" "Why are my breasts asymmetrical?" "How much exercise should I take?" "Why are my periods not regular?" The questions were addressed in the tutor group sessions and written material provided after each session. These tutorials were rated by pupils as the most popular and informative sessions of the year long personal and social development programme.

Studies such as that of Oppong-Diseng and Heycock highlight various issues regarding health services for adolescents. Adolescents are asking for more information on topics that worry them and they are asking for drop-in centres and specific adolescent health specialists. We must ensure that adolescents know what their local health related services are and must make sure that these services are user friendly and staffed by adolescent friendly doctors. Taking adolescent views into consideration and planning services accordingly are the first steps in persuading adolescents to use these services.

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How to organise the paediatric MRCP (UK) part II clinical examination

EDITOR,—The review by Skinner *et al* of the organisation of the paediatric MRCP (UK) part II examination¹ gives insufficient recognition to the role of nurses, medical secretaries, and play therapists, who are at least as important as the registrar and host examiner. Our secretaries typed letters of invitation, summaries, and thank you letters and helped contact and fielded inquiries from parents. Much of this had to be completed on top of their NHS workload. Nursing staff set up the venue, collected equipment, provided

refreshments, and made up name badges. At our centre, each candidate is shadowed by a nurse helper who ensures that they fill out the correct paperwork, directs them around the venue, finds them refreshment between sections of the examination, and provides support when a case or viva has gone badly. Examiners and candidates commented favourably on this system, and many candidates show their appreciation by contacting

their nurse helper when they pass! Our play therapist was the key to occupying the children during the long periods of waiting required, and in making the examination an enjoyable experience for parents and children.

These staff give generously of their time with none of the professional kudos gained by the registrar. Their commitment is vital for a successful examination.

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