References


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Dr Dodge comments:

I am grateful for Professor Salazar de Sousa’s comments. I had intended my paper to be provocative.

In my own practice, I follow the ESPGAN protocol and subject patients to three biopsies. However, this procedure is followed by only two-thirds of the members of ESPGAN and I imagine that only a few nonspecialist general paediatricians in this country routinely perform a gluten challenge, followed by a biopsy. Moreover, the challenge procedure varies considerably from putting the patient on a free diet to daily administration of a prescribed large amount of gluten. The postchallenge biopsy is essential if a diagnosis of persistent gluten enteropathy is to be made. I want to encourage paediatricians who are daunted by the present protocol to give a gluten challenge to patients who have benefited clinically and, if known, histologically from a gluten-free diet.

Most of my patients in Cardiff seem to adhere very well to their dietary regimen, and it is exceptional to find one in whom the mucosa has not recovered when a second biopsy is performed. However, simply knowing that the mucosa is still abnormal would not ensure future patient compliance, and I believe that our good results can be attributed to the continuing advice and encouragement given to the parents by our dietician. The primary purpose of my paper was not to challenge the usefulness of the ESPGAN criteria (and I agree that we have nothing better at the moment), but to suggest that until we have a satisfactory definition of coeliac disease that is accepted both by physicians and paediatricians, we could improve the clarity of our communications by using the terms ‘gluten intolerance’ and ‘gluten enteropathy’ to describe clinical symptoms and histological appearances. When Professor Salazar de Sousa tells me that a patient has coeliac disease I understand him perfectly because we both use the same diagnostic criteria, but it seems more sensible to use accurate descriptive terminology when the investigative process is incomplete, or when the criteria are not agreed.

References


J A DODGE
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Dissolution of bilateral cystine calculi by penicillamine

Sir,

I read with interest the paper by Ruysch van Dugteren and Wiggelinhuizen.1 I have obtained similar results with alpha-mercaptopropionyl glycine. Cystine-lysine-ornithine-argininuria was diagnosed in two siblings—one with bilateral staghorn calculi—by using high-tension electrohyresis. Calculi were dissolved by means of alpha-mercaptopropionyl glycine in the child with bilateral calculi (Figs 1 and 2), while prevention, without presence of calculi, was achieved with the same drug in the other child. These results show that the drug is useful

![Fig. 1 Patient aged 5 years: (x-rays of abdomen). Bilateral renal calculosis (arrows).](attachment:image)

1. Ruysch van Dugteren K, Wiggelinhuizen C F. Dissolution of bilateral renal cystine calculi by penicillamine.
Correspondence

Fig. 2  Patient aged 7 years: (x-rays of abdomen). After more than 2 years of treatment with alpha-mercaptopyrropropionyl glycine, total dissolution of bilateral calculi.

for the treatment and prevention in homozygotes for the disease. Alpha-mercaptopyrropropionyl glycine may be safer than D-penicillamine.

Reference


Dr Ruysch van Dugteren comments:

D-penicillamine has been, until recently, the only thiol readily available for the treatment of cystine stone-forming patients. Potentially serious side effects have necessitated the withdrawal of this drug in 50% of patients.

Since 1973 alpha-mercaptopyrropropionyl glycine has been used in a number of (presumably) adult patients.1-3 In a total of 26 patients so treated for up to 5 years no serious side effects were observed, and the effectiveness of the drug in reducing cystine excretion in the urine equalled that of D-penicillamine.

Professor Berio’s letter provides further clinical evidence that alpha-mercaptopyrropropionyl glycine may prove to be the drug of choice in children with cystinuria. It must be stressed however that a 3-pronged attack with (1) an increased fluid intake, (2) alkalinisation of urine, and (3) a thiol such as alpha-mercaptopyrropropionyl glycine, has a far better chance of succeeding in dissolving stones than any one of these methods on its own.

References


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Cows’ milk protein-sensitive enteropathy

Sir,

In 1978 we proposed the following criteria for the diagnosis of cows’ milk protein-sensitive enteropathy:1
(1) Clinical disease (diarrhoea with or without vomiting) while receiving cows’ milk protein.
(2) Clinical improvement on a diet free of cows’ milk protein.
(3) Normal or mildly abnormal histology of jejunal mucosa when taken 6-8 weeks after symptoms subside.
(4) Histological relapse, with or without clinical relapse, after re-exposure to cows’ milk protein.

We have now studied a further 60 infants. In analysing the clinical, histological, and enzymological results we have found that cows’ milk protein can cause depletion of the mucosal oligosaccharidases, and malabsorption of xylose in the absence of mucosal damage visible under the light microscope.

These 60 infants were clinically suspected to have intolerance to cows’ milk protein. After their fluid, electrolytes, and acid-base imbalance had been corrected, they were started on a lactose- and cows’ milk protein-free formula which was maintained for 6-8 weeks. After this they were readmitted for milk challenge which was performed as has been described.3 The clinical features of these 60 infants at admission have been described.4

From clinical, histological, and enzymological response to cows’ milk protein, after milk provocation, each infant could be placed in one of 4 groups.

Group 1 consisted of 22 infants with significant histological changes associated with pronounced reductions in disaccharidase levels. All 22 infants developed diarrhoea, some of them with vomiting, fever, or lethargy, 17 of them within 24 hours and five 3 to 28 days after milk provocation. 11 of 12 infants tested had reducing sugar in the stools.

Group 2 consisted of 22 infants with significant histological changes and reduction in enzyme levels (except in