

Comment

The clinical features in this case were those of the recessively inherited syndrome of chronic cholestasis and lymphoedema described by Aagenaes.² The illness was unique among over 100 cases of neonatal hepatitis studied during the last 15 years³ and incompatible with the simple diagnosis of Wilson's disease. The neurological features in the terminal stage of the disease, however, were those of lenticular degeneration, and the microscopic and chemical changes at necropsy seemed compatible with the proposal that they were caused by copper. We suggest that the patient's initial liver disease interfered with copper excretion, causing copper retention within the liver followed by overflow to the brain, and that the copper caused lenticular damage and contributed to the rapid progression of the liver disease. This sequence of events resembles that in Wilson's disease, except that the primary defect in Wilson's disease presumably affects biliary excretion of copper in a more specific way.

Several important points follow. Firstly, this complication should be sought in other patients with Aagenaes's syndrome with a view to using penicillamine more energetically. Secondly, the observations support the widely held view that the lenticular degeneration of Wilson's disease is a non-specific consequence of overflow of copper from the liver. Finally, this case has caused us to look for copper retention in other children with chronic liver disease. We have observed very high liver copper concentrations in several other children with childhood cirrhosis and have seen a gratifying response to penicillamine therapy in two of them. No particular relationship between the degree of copper retention and the form of the initial liver disease has emerged. Copper retention is particularly prominent in primary biliary cirrhosis. Controlled studies of penicillamine treatment in this disease have been reported and show encouraging results.⁴ Other complications of secondary copper retention have been reported.⁵

We advocate measuring the liver copper content in patients with chronic liver disease at all ages to accumulate information which may allow meaningful trials of treatment with penicillamine in selected cases to test the hypothesis that copper retention can be a factor contributing to progression to cirrhosis.

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¹ Hunt, A H, *et al*, *British Medical Journal*, 1963, **2**, 1498.

² Aagenaes, O, *Acta Paediatrica Scandinavica*, 1974, **63**, 465.

³ Danks, D M, *et al*, *Archives of Disease in Childhood*, 1977, **52**, 360.

⁴ Jain, S, *et al*, *Lancet*, 1977, **1**, 831.

⁵ Fleming, C R, *et al*, *Gastroenterology*, 1976, **71**, 905.

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Department of Paediatrics, Royal Children's Hospital, Parkville, Victoria, Australia

ARNOLD L SMITH, MB, FRACP, assistant gastroenterologist
DAVID M DANKS, MD, FRACP, professor and medical geneticist

Breakfast and dietary aspects of Crohn's disease

There has been considerable speculation about the cause of Crohn's disease. Diet may play a part. A recent study¹ showed that patients with the disease consumed large quantities of refined carbohydrate. James² examined breakfast habits in 34 patients and found that most of them ate cornflakes regularly at the onset of their illness. We have re-examined some aspects of diet, particularly breakfast habits, in 100 patients and 100 matched controls.

Patients, methods, and results

We interviewed 48 men and 52 women with Crohn's disease. Their ages ranged from 15 to 81 years (mean \pm SD 41.7 \pm 15.8). The mean interval since the initial diagnosis was 8.9 \pm 7.2 years. We chose 100 controls, matched for age and sex, from either patients or their relatives attending a fracture clinic. Patients and controls were interviewed by the same person using a

Number of patients and controls regularly consuming certain foods at breakfast at least twice weekly and of those with other habits unrelated to breakfast. Analysis is by χ^2 on one degree of freedom except for subjects who swallowed toothpaste, when a two-tailed Fisher test was used

Dietary and other habits	Crohn's patients (n = 100)	Controls (n = 100)	Significance (P)
Foods at breakfast			
Bread	91	86	
Toast	59	64	
Egg	31	37	
Fruit or fruit juice	14	30	<0.02
Porridge	20	18	
Weetabix, Shreddies, or Shredded Wheat	21	19	
Cornflakes	29	22	
Special K	4	7	
Rice Krispies	6	6	
Sugar Puffs	3	1	
Bran or All Bran	13	12	
Muesli	3	10	
Any cereal	55	55	
Cereals at other times of day	12	12	
Seasonal variation	18	13	
Other habits			
Coca Cola consumption (twice weekly)	38	34	
Alcohol consumption	42	55	
Smokers	46	44	
Toothpaste users	73	74	
Toothpaste swallowers	6	1	
Addition of sugar to:			
Tea or coffee	78	52	<0.001
Mean number of teaspoons to each cup	1.79 \pm 1.3	1.14 \pm 1.3	<0.001
Cereals	57	42	<0.05
Mean number of teaspoons to each bowl	1.38 \pm 1.5	0.84 \pm 1.21	<0.006

questionnaire in an open study. The frequency at which certain breakfast foods were currently eaten was recorded as regular (at least twice weekly) or irregular (less than twice weekly). The results (table) showed no statistical difference except for fruit or fruit juice, which were taken more often by controls ($P < 0.02$). Those who regularly had cereals at another time of day or changed their cereals between winter and summer were also noted. All subjects were asked whether any of a given list of foods precipitated abdominal symptoms. A significant difference was noted for a large range of products which included cheese, chocolate, eggs, nuts, vegetables (especially cabbage), meat (mainly pork), and cereals.

In view of previous reports about cornflakes and Crohn's disease patients were asked whether they knew of an association between food (unspecified) and the disease. Twenty-nine knew of the possible association with cornflakes and 12 of these had stopped eating them, having previously taken them regularly. The corresponding figure for their 29 matched controls was only three ($P < 0.02$). Twenty-one of the 71 patients who were unaware of the association had also discontinued cornflakes. The figure for their 71 controls was 10 ($P < 0.05$). Seemingly patients with Crohn's disease had significantly reduced their consumption of cornflakes compared with controls, irrespective of whether they were aware of the possible association. The amount of sugar added to beverages and cereals was recorded. There were more sugar takers among patients than controls. This was true for addition of sugar to tea and coffee ($\chi^2 = 13.74$; $P < 0.001$) and cereals ($\chi^2 = 3.92$; $P < 0.05$). The amount of sugar consumed was also significantly greater for addition to drinks ($t = 3.52$; $P < 0.001$) and to cereals ($t = 2.80$; $P < 0.006$). Other questions related to toothpaste, smoking, and consumption of alcohol and Coca Cola.

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We looked at current breakfast habits because people's recollection of previous detailed habits may be inaccurate. The difference in sugar consumption between patients and controls is difficult to interpret. Patients may increase their consumption in an attempt to compensate for loss of energy or weight. They may exclude other foods which cause symptoms, or include those which are better tolerated. A direct causal relationship between Crohn's disease and food could be difficult to establish.

¹ Martini, G A, and Brandes, J W, *Klinische Wochenschrift*, 1976, **54**, 367.
² James, A H, *British Medical Journal*, 1977, **1**, 943.

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University Hospital of Wales, Cardiff CF4 4XW

JOHN F MAYBERRY, MB, BCH, clinical research officer
JOHN RHODES, MD, FRCP, consultant physician
R G NEWCOMBE, MA, FSS, statistician