Colonic strictures in cystic fibrosis

EDITOR.—We report two colonic abnormalities in children with cystic fibrosis.

A. R., a 5-year-old boy, presented with meconium ileus necessitating the removal of 34 cm of ileum including the ileocecal valve. By 3 years of age he had persistently loose stools and deteriorating chest disease with weight on the 10th centile. Barium enema showed a normal anatomy and normal large bowel. His enzyme supplement was changed from Pancrease (Cilag) to Creon 25000 (DuPharm) and five months later to Pancrease HL (Cilag). In response to symptoms the dose was incrementally increased over the next year to 40 capsules a day. This provided 46 000 to 66 000 units/kg/day of lipase and 2300 to 3300 BP units/kg/day of protease. During this period there was no change in his symptoms nor any objective improvement in fat absorption. He had a hypochromic microcytic anaemia and weight remained below the 10th centile. At 4-7 years he had fresh blood in the stools over a two week period without any other change in his symptoms. Repeat barium enema demonstrated an extensive stricture from the remaining ascending colon to the descending colon. A defunctioning ileostomy was fashioned. Histology from full thickness colonic biopsies demonstrated a 4-5 mm thick wall in which there was mild suberosal fibrosis and very prominent deep submucosal fibrosis. The mucosa showed a diffuse mild colitis with lengthening of crypts, mild lamina propria fibrosis, and irregularity of the surface epithelium. He has been changed back to Creon providing 18 500 BP units/kg/day of lipase and 485 BP units/kg/day of protease.

A. P., a 7-year-old who had been on Pancrease HL for 14 months and Nutrizym 22 (Merck) for one month had a barium enema because of persistent abdominal pain and a mass in the right iliac fossa. The ascending colon was featureless and the lumen looked slightly narrowed. Colono-scopic biopsies show mucosa within normal limits but are too superficial to comment on the submucosa. She had been receiving up to 33 500 BP units/kg/day of lipase and 1675 BP units/kg/day of protease.

For the clinic as a whole the change to high lipase and protease has resulted in a reduction in mean capsule number from 30 to 18 per day. There has been no change in fat absorption (mean 86%) but there has been a significant increase in mean lipase (8400 v 18 500 BP units/kg/day) and protease (340 v 600 BP units/kg/day) intake relative to body weight, and to dietary fat content (2400 v 5100 BP units lipase/kg fat intake).

Colonic strictures with massive submucosal fibrosis represent a new finding in cystic fibrosis. Unlike the original cases reported from Liverpool1 these two children had a long history of abdominal problems. The high lipase enzyme intake was above the mean for the clinic. It seems likely that the high lipase enzymes are having a direct toxic effect mediated by an unknown mechanism. This is known to be prolonged in cystic fibrosis and continuing lipolysis and proteolysis in the colon may be contributing to an irritant effect. This may be further exacerbated by alterations in the mucus glycoprotein production.

We suggest that any child receiving high lipase enzymes, or who has done so in the past, who is experiencing abdominal pain or has poorly controlled fat absorption should have a barium enema. We have not found bowel ultrasonography helpful even in the hands of an experienced radiologist.

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2 Horizontal transmission of hepatitis B from children to adoptive parents

EDITOR.—Hepatitis B virus is a highly infectious disease that can cause serious liver disease and death. Nearly half of adult patients acquire infection during childhood,

one quarter after perinatal transmission.2 Horizontal non-sexual transmission between children, frequent in high endemic area, also occurs in families or school contacts of chronic carriers.3 Among 65 chronic hepatitis B patients in our unit (1989-93), 31 were adopted children (21 from Romania, 11 Asia, and one Africa). Mean age on arrival was 32-9 months (range 8-30 months). All had positive HBs antigen and serum DNA (range 0-104 pg/ml, Abbott immunoassay). Anti-hepatitis B virus vaccination was systematically offered to families at the first visit, and familial screening performed. Vaccination was entirely performed in 22 families (40 parents and 42 siblings). Seven families were partially vaccinated: all children, but no fathers and three of seven mothers. Seven (64%) of the 11 non-vaccinated parents from six families developed acute hepatitis B, seven to 29 months after arrival of the child (positive serum HBs ag, Hbe ag, and anti-HBc antibodies). Transaminases reached a mean (SD) 1070 (370) iu/l for aspartate aminotransferase and 2133 (1200) iu/l for alanine aminotransferase. Clinical illness caused work incapacity for three to six weeks. All parents became Hbs antigen negative within six weeks of onset, with normal transaminases. Three additional parents had anti-HBs and anti-Hbc antibodies at screening. One single non-vaccinated parent did not develop acute nor subclinical hepatitis B. Acute hepatitis B occurred also in one 50 year old school teacher, one grandfather, and one 16 year old friend. Blood contamination was recognised in only four cases (wound, scalps, lip cracks, or nasal bleeding).

It seems evident from this and previous results that hepatitis B virus vaccination must be administered to families and classmates of chronic carriers.

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Barry Pless is professor of paediatrics, and of epidemiology and biostatistics, and in this excellent work he successfully combines these perspectives and brings together contributors from the UK, North America, and Australasia. Readers should not be intimated by the opening crash course in epidemiological principles. The examples in the chapters that follow demonstrate how the science is practically applied to paediatrics and elucidate further the basic principles. The book is divided into sections on perinatal, infectious, mental and behavioural and chronic disorders, and on injuries and violence. All the authors follow a common format and review the literature, including a discussion of patterns of occurrence, risk factors, and interventions. The difficulties of obtaining conclusive ‘evidence’ and the limitations of many published studies are explained.

A discussion about the problems associated with low birth weight and prematurity is complemented by the chapter on cerebral palsy. Significant improvements in the treatment of sequelae are not matched by success in the primary prevention of fetal growth retardation and preterm delivery. The prevalence of cerebral palsy is rising, particularly within the preterm subgroup. A major reduction appears unlikely until our understanding of antenatal causes is expanded.

The chapter on communicable diseases includes a timely description of the attempt to eliminate indigenous measles in the United States. An initial 90% reduction was followed by an epidemic of 46 000 cases over a two year period and since 1989 a second dose of measles vaccine has been recommended.

The book would be of value when preparing a presentation on one of the many topics...
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