Familial Duodenal Atresia

Congenital atresia of the bowel is infrequent (Harris and Steinberg, 1954); its familial incidence therefore constitutes a finding of special interest. Very few reports have been published on familial congenital intestinal obstruction not associated with mucoviscidosis (Winter and Zeltzer, 1956; Mishalany and Najjar, 1968; Esterly and Talbert, 1969).

To our knowledge no instance of familial duodenal atresia has been described. We wish to report the occurrence of duodenal atresia in 3 sibs.

Case Reports

The parents are young, healthy first cousins. No history of abortions, stillbirths, or premature deaths in the family could be elicited, nor could they recall any periods of ill-health during pregnancy. Their firstborn, a girl, was the product of a normal pregnancy and delivery, and is healthy. The following three pregnancies, during three consecutive years, were complicated by polyhydramnios in the last two months of pregnancy. The infants were a girl weighing 2300 g. at 35 weeks’ gestation, a boy weighing 2800 g. at term, and a girl weighing 2250 g. at 36 weeks’ gestation. The first two were born at the maternity centre in her Arab village, and the last girl was born at the Hillel Yaffe Government Hospital. The clinical, radiological, and operative findings were essentially the same in all three affected babies: continuous vomiting of bile, epigastric distension, and colourless meconium obtained by rectal examination. Abdominal x-rays showed the classical ‘double bubble’ pattern. Laparotomy revealed atresia of the third part of the duodenum, with marked dilatation of the proximal loop. The first two babies were brought to our hospital at the age of 2 days. The third affected sib, born in our hospital, was operated on 12 hours after birth. End-to-oblique duodeno-jejunostomy was performed, with good initial results, but all 3 babies died 5 to 6 days after operation. Necropsy revealed bronchopneumonia and localized peritonitis, but no evidence of mucoviscidosis, rubella, or other local or systemic illness which could be considered responsible for the anomaly.

Chromosome studies from the last case showed a normal karyotype.

Discussion

The literature contains very little information on familial congenital intestinal obstruction. 2 brothers reported by Blanck, Okmian, and Robbe (1965) had atresia of the middle ileum in association with meconium ileus; Winter and Zeltzer (1956) reported 2 sibs with ileal atresia; Mishalany and Najjar (1968) described 3 sibs with jejunal atresia; Esterly and Talbert (1969) reported twins with jejunal atresia and ascribed the lesion to the effects of rubella arteritis. Our report is the first on familial duodenal atresia, and raises again the problem of aetiology of intestinal atresias.

The concept of faulty recanalization after an embryological ‘solid stage’ of the gut as the cause of intestinal atresias has been losing ground. There is mounting evidence that injury to the fetal bowel, especially interference with blood supply to a portion of it, is the main cause of atresias (Grob, 1960; Santulli and Blanc, 1961). This approach has been substantiated by experimental observations (Louw and Barnard, 1955), and is in accordance with the frequent association with mucoviscidosis (Bernstein et al., 1960; Andersen, 1962).
term 'familial' does not seem to apply to such a situation.

While Tandler's faulty recanalization concept may be of lesser importance in explaining atresias beyond the duodenum (where no 'solid stage' has been satisfactorily demonstrated), it may well apply to duodenal atresias. This is supported by the observation that it may be accompanied by other errors of growth, notably Down's syndrome (Bodian et al., 1952). This prompted us to search for a chromosomal aberration, but the karyotype studied was normal.

The occurrence in three consecutive sibs of an uncommon condition such as duodenal atresia strongly suggests genetic determination. The parents being first cousins, it is possible that we are dealing with an autosomal recessive gene.

**Summary**

Three sibs born consecutively with atresia of the third part of the duodenum are described. No additional disease was found, and chromosomal studies from one case were normal. The parents are first cousins, and it is assumed that the anomaly in this kindred is determined by an autosomal recessive gene.

**References**


**Moshe Berant and David Kahana**

The Department of Paediatrics, The Hillel Yaffe Memorial Government Hospital, Hadera, Israel

---

**Immunoglobulins in Protein-Calorie Malnutrition**

It is well established that deficiency of immunoglobulins predisposes to infection. We set out to ascertain whether such a deficiency exists in patients with protein-calorie malnutrition, which might in part account for the common clinical observation of an association between malnutrition and infection. Brown and Katz (1965) reported a significant decrease in the serum IgG levels of 20 children with kwashiorkor compared with 5 normal children. In 7 marasmic infants, Najjar, Stephan, and Asfour (1969) found the mean serum levels of IgG, IgM, and IgA to be higher than in healthy children. In a comparison between 11 children with kwashiorkor and 11 well-fed children suffering from similar infections, Keet and Thom (1969) found no significant difference in the serum levels of IgG and IgM. IgA levels were much increased in the kwashiorkor group.

**Patients and Methods**

Serial estimations of serum IgG, IgA, and IgM were made on admission, during recovery, and during convalescence in 24 patients with protein-calorie malnutrition suffering from a variety of infections. Almost all had gastro-enteritis, and upper respiratory tract infections; pneumonia and viral infections (including 4 cases of chicken-pox and one of measles) were also common. The children were between 7 and 34 months old (mean age 20 months) and the majority were obvious cases of kwashiorkor. Four patients died, all in the first week.

During the same period we were studying another group of patients who had recurrent or persistent infections. For comparative purposes we have tabulated the immunoglobulin levels of 12 of them who showed no evidence of malnutrition, and were of similar ages to the 24 subjects. The nature of the infections in this control group was different. Otitis media, pneumonia, meningitis and upper respiratory tract infections were commonest. Only one child had gastro-enteritis, and one had chicken-pox. The age range was 7–31 months (mean 19 months).

Immunoglobulins were measured by the radial diffusion method, using commercially available antibody-agar plates (Hyland laboratories, Los Angeles). Serum total proteins and albumin were measured by the biuret method, using 28·3% sodium sulphate to precipitate globulins.

**Results**

These are shown in the Table. The means, standard deviations, and Student's t test for small samples were done on the logarithms of the observed values. The 2 SD range about the geometric means recorded is thus exponential, and in most cases exceeds the observed range.
Familial duodenal atresia.

M. Berant and D. Kahana

Arch Dis Child 1970 45: 281-282
doi: 10.1136/adc.45.240.281

Updated information and services can be found at:
http://adc.bmj.com/content/45/240/281.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/