The Turkish Journal of Pediatrics
1972 / Vol. 14 / No. 3 / pp. 94 – 98

Congenital Megacolon Associated with Enterocolitis and Hypoproteinemia

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Association of congenital megacolon with enterocolitis and hypoproteinemia is a serious, often fatal condition in the newborn period.[1 2] It is rare and the exact etiology is not clear.

J. M., a four week old Puerto Rican male was admitted to St. Christopher's Hospital for Children for diarrhea, vomiting and abdominal distention of three days duration and fever up to 104 °F. On admission to the hospital a pale, quite sick looking infant presented, with marked abdominal distention, dyspnea and grunting respirations. A flat plate of the abdomen revealed a narrow rectal sigmoid area and a widened descending colon adjacent to the narrowed portion. This suggested the diagnosis of congenital megacolon with enterocolitis. Laboratory examination showed, among others, a hemoglobin of 12.4 gms. % and a total protein of 1.9 gms. %.

We subsequently reviewed the clinical and laboratory findings on 30 patients with congenital megacolon. Our patient material suggests that the combination of congenital megacolon with enterocolitis and hypoproteinemia is of more than coincidental importance.

Patient Material

This report is based on review of clinical and laboratory findings of 30 patients with congenital megacolon, of whom 5 had enterocolitis.

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The diagnosis of congenital megacolon was based on histological evidence in all but one of these patients. In this infant the diagnosis was highly suggestive on the basis of history and X-ray findings.

Of the 30 patients, 20 were males, 10 females. The ages ranged from 1 week to 8 years. Patients with enterocolitis were in the younger age group, four patients were less than 2 months, and one was 14 months old.

A number of anomalies were encountered in our patients. These are listed in Table I. Four of these seven patients had enterocolitis.

TABLE I

Patient Name	Anomalies Associated with Congenital Megacolon
M.S.*	Congenital heart disease, microphthalmia.
J.M.*	Congenital heart disease, mongolism.
A.N.	Congenital heart disease.
C.G.	Congenital heart disease, malrotation small intestine.
D.B.*	Absence of 12th rib.
F.R.	Sturge-Weber Syndrome, double collecting system of right kidney
D.R.*	Congenital short bowel, rudimentary auricle.

*Patients with enterocolitis.

Symptoms: In the great majority of patients the onset of symptoms was early, 25 of the 30 infants were symptomatic at birth. One showed his initial symptomatology at the age of 2 weeks, two at 4 weeks and two at 6 weeks. The prominent clinical features were constipation, abdominal distention and vomiting. Table II summarizes the clinical findings for the patients with associated enterocolitis as compared to those with uncomplicated congenital megacolon.

Laboratory Findings: The hemoglobin levels of the patients with enterocolitis were normal. The WBC counts were above 10,000 in all five patients with enterocolitis. In one patient, the WBC was 30,200. The remaining 25 patients had normal WBC counts.

Serum protein levels were done in 15 patients and found low in four patients with enterocolitis. Only one patient with enterocolitis had normal values. Albumin and globulin ratio were unchanged, unfortunately

TABLE II

Patients	Constipation	Alternating Constipation and Diarrhea	Severe Diarrhea	Abdominal Distention	Vomiting	Respiratory Distress	Fever
Congenital Megocolon (25 patients)	20	5	0	15	14	1	0
Congenital Megacolon itwh Enterocolitis (5 patients)	0	2	3	5	4	4	4

no protein electrophoresis were performed. Remaining ten patients who had no enterocolitis had normal serum protein levels.

Stool cultures were done on all patients with enterocolitis. Five patients had Pseudomonas species, four had pathogenic E. coli, in addition to this, and one had also Staphylococcus aureus.

Comments

The presence of diarrhea, abdominal distention with elevation of temperature and leukocyte count clinically, suggests enterocolitis in these five patients with congenital megacolon. This complication has been reported by several authors.[3 4 5 6] The cause of the enterocolitis, however, is not known. It has been suggested[7] that patients with congenital megacolon have partial intestinal obstruction and consequently might have higher intraluminal pressures. This could lead to increased permeability, to bacteria and viruses, and mucosal ulcerations, sloughing off and even perforation can occur. Peritonitis and septicemia are usually fatal complications of enterocolitis. The prognosis of enterocolitis is poor with a reported mortality of 33 percent[4] or higher.[8] Our mortality rate was three out five in spite of vigorous treatment.

The most interesting finding in our patient material was the association of hypoproteinemia in four out of five of our patients with enterocolitis. The normal hemoglobin levels noted on admission would suggest that hemodilution is not the likely cause of the low serum protein levels. Urinalyses failed to reveal albuminuria in any of these patients. Oozing of serum protein from the diseased intestinal mucosa may be the main cause of hypoproteinemia. One patient with enterocolitis, who had a protein level of 1.9 gm. % also had edema and ascites.

Hypoproteinemia associated with congenital megacolon has previously been reported.[1 2] It has been thought to be secondary to malabsorption during the episodes of diarrhea. It is unlikely that the patients with enterocolitis and Hirschsprung's disease can be studied for protein loosing enteropathy on account of their poor condition and the need for intervention. We call attention to the presence of hypoproteinemia in patients with Hirschsprung's disease and enterocolitis as it might possibly represent an additional early sign, amenable to correction.

Another unexpected and incidental finding in our patient material is the high incidence of congenital abnormalities and congenital anomalies in patients with Hirschsprung's disease and enterocolitis. It is of interest that congenital heart disease was found more frequently than the other anomalies.

Summary

Of 30 patients with congenital megacolon, 5 developed enterocolitis. Serum protein levels were found to be low in 4 of these 5 patients and hypoproteinemia is emphasized as an additional sign in the enterocolitis of Hirschsprung's disease.

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