CONGENITAL INTESTINAL ATRESIA*

EDWARD E. JEMERIN, M.D. AND CYRILLE R. HALKIN, M.D.
NEW YORK, N. Y.

CONGENITAL INTESTINAL ATRESIA is an uncommon condition although the true incidence is probably considerably higher than that indicated by the number of cases which have reached the literature. Most cases have not been recorded until recently, and even now many infant deaths due to intestinal atresia remain undiagnosed or are not reported. Until some central Registry for the recording of such cases is set up, it is incumbent upon any physician encountering one to see that at least a brief report is entered in the literature. This is particularly the case in view of the extremely high mortality associated with the malformation, a mortality which early recognition and prompt adequate treatment seems to be lessening appreciably. The case encountered by the authors was that of a complete high jejunal atresia in a three and one-half pound premature infant who recovered following jejuno-jejunostomy, the smallest case on record in which a successful result was achieved. An incomplete rotation of the colon was also present but this was not obstructing and did not therefore influence the clinical picture or appreciably alter the surgical problem.

CASE REPORT

F. G., Sydenham Hospital No. 108318, was a premature male infant spontaneously delivered on March 12, 1948, after a 30-week gestation (Dr. Daniel Wanderman). There had been two previous pregnancies; a full term stillbirth four years before, and a full term forceps delivery of a normal boy two years ago. The mother was Rh negative with no demonstrable antibody formation. No history of virus infection during pregnancy could be elicited.

The infant weighed 3 lbs. 14 ozs. at birth and was cyanotic but responded to aspiration and oxygen. Physical examination revealed marked overriding of all skull bones at the parietal, sagittal and occipital sutures, with complete closure of the anterior and posterior fontanelles. The eyes were wideset and a cataract-like lesion of the right was noted. The lungs were fully aerated, the heart normal. The abdomen was soft, the liver edge palpable one finger below the costal margin.

The infant was placed in an incubator with continuous oxygen and 5 mg. vitamin K daily. Pediatric care was supervised by Dr. Lawrence B. Slobody. After twelve hours of starvation, small amounts of glucose water were offered for twelve hours followed by a 1:3 evaporated milk formula. On the second day after delivery, regurgitation after feeding began, the milk curds being mixed with thin yellow fluid. Moderate icterus and a petechial rash on the bregma and the dorsum of the feet were noted, and the temperature rose to 102.8 degrees. A small meconium stool was passed. All oral feeding was stopped and intensive parenteral hydration with saline and Hartman's solution was begun. Intramuscular penicillin was also given. Intermittent vomiting continued and by the evening of the third day, the infant appeared markedly dehydrated and in very grave condition. There was no appreciable distention and no peristaltic waves or pyloric tumor were observed.

* Submitted for publication, December, 1948.

On the fourth day, the general condition was somewhat better. The icterus had deepened but the state of hydration had improved. Regurgitation continued but the infant seemed to retain small amounts of gavage feedings which had been added to the regimen. Several more meconium stools were passed. The blood count revealed a hemoglobin of 14.5 Gm., a red blood count of 5,020,000 per cu. mm., and a white blood count of 24,600 per cu. mm. with 16 immature forms, 57 mature polymorphonuclear leucocytes, 1 eosinophil, 22 lymphocytes, 4 monocytes and 1 nucleated red blood cell per 100 whites. A slight trace of bile was present in the urine. A plain roentgen film of the abdomen

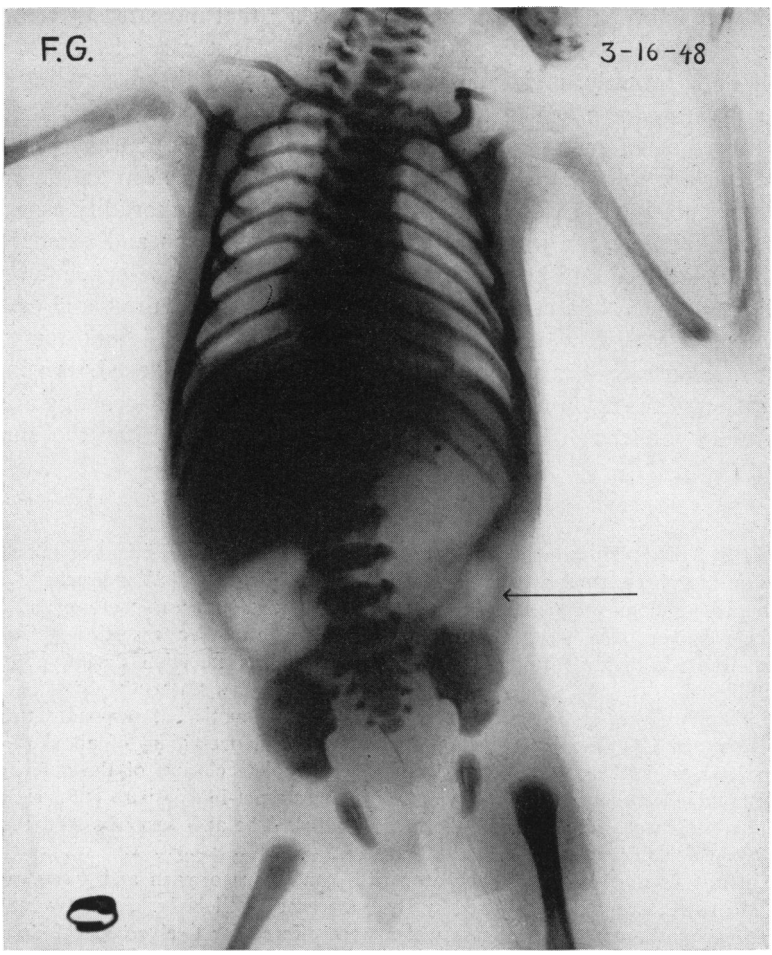

FIG. 1.—March 16, 1948; plain film of abdomen showing stomach, duodenum, and beginning of jejunum (arrow) markedly distended by gas. No gas is seen in remainder of intestine.

(March 16, 1948) revealed a large collection of air in the stomach, duodenum, and proximal few inches of jejunum. No gas was observed in the intestine distal to this (Fig. 1).

The appearance and clinical course were suggestive of high intestinal obstruction but because of the absence of abdominal distention or peristaltic waves and the apparent partial retention of gavage feedings and the passage of meconium stools, a thin barium mixture

was given by tube to corroborate the diagnosis. Meanwhile, hydration and supportive therapy were continued. The roentgenogram after barium administration showed passage of the mixture only as far as the first part of the jejunum (Fig. 2). The diagnosis of obstruction of the upper jejunum was now clear and efforts were made to prepare the infant for operation. By the following day (March 18, 1948) his condition was sufficiently improved to attempt surgery. The weight on this day was 3½ lbs.

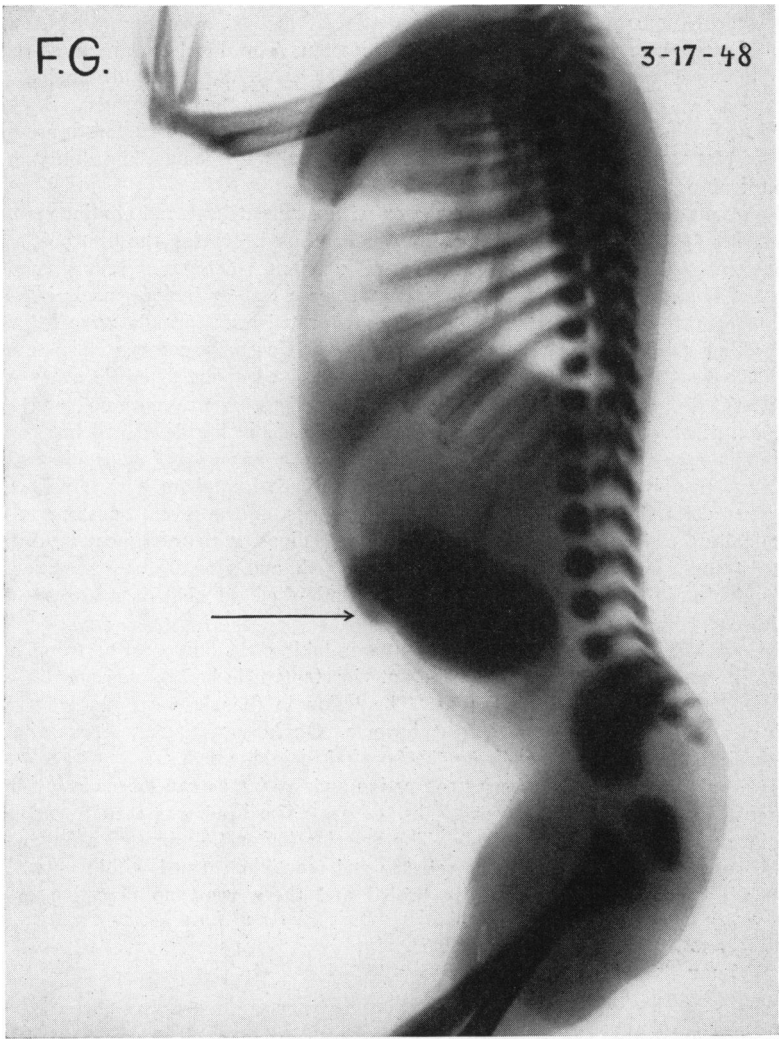

FIG. 2.—March 17, 1948; Film after barium showing abrupt termination of barium column at proximal jejunum (arrow).

With a blood transfusion running and a gastric tube indwelling, laparotomy was performed (E. E. J.) The abdomen was opened through a right rectus muscle splitting incision. All the intestines were delivered. The large intestine was found to be incompletely rotated with the cecum subhepatic in location and the ascending and transverse

colon adherent to the transverse mesocolon and posterior peritoneum. These had to be dissected free before the pathologic condition responsible for the obstruction could be appreciated and exposed. After division of the adhesions and liberation of the colon, marked distention of the stomach, duodenum, and first portion of the jejunum was seen. This terminated about an inch beyond the ligament of Treitz in a blind pouch without continuity with the distal jejunum. Though tremendously dilated the bowel was viable. The distal blind jejunal end was located by tracing proximally from the ileocecal valve. The proximal segment of jejunum beyond the ligament of Treitz was too short to permit anastomosis. This technical difficulty was solved by reduction of the proximal segment through the fossa of Treitz, made possible by the blind termination. The distal bowel was so collapsed that its diameter was less than 0.5 cm. Its caliber was enlarged by distending it with saline and a two layer side-to-side jejuno-jejunostomy was then performed. Exploration revealed no further abnormalities. The wound was closed with interrupted figure-of-eight sutures of No. 32 stainless steel wire through fascia and peritoneum. The skin was closed with interrupted silk sutures.

The infant was replaced in the incubator with external heat and continuous oxygen. The gastric tube was left in situ. Penicillin was continued. During the first postoperative day the temperature rose to 102.4 degrees. The infant voided, and regurgitated small amounts of bile-stained fluid. Fluid requirements were met by hypodermoclysis. Twenty hours postoperatively small amounts of a 1:3 evaporated milk formula were fed through the indwelling tube. At this time the red blood count was 2,850,000 per cu. mm. and the hemoglobin 8.9 Gm. On the second day the baby retained about ½ ounce every 2 hours with occasional regurgitation of bile-stained fluid. Clyses were continued and a blood transfusion given. Twitching motions of the right arm and leg developed but responded to calcium therapy. Thirty-six hours after operation a formed yellow stool was passed which was strongly positive for occult blood, and thereafter about 3 to 4 yellow stools were passed daily. The infant's weight was 4 lbs. 2 ozs. on the seventh postoperative day. He gradually took larger and more concentrated feedings, with infrequent regurgitation. Repeated transfusions were given on the 2nd, 8th, 11th, and 33rd days for recurrent drops in hemoglobin. The icterus meanwhile had diminished so that it was only faintly appreciable.

Five weeks after the operation the infant was taking 2½ ounces of a strong formula. His weight at this time was 5½ lbs. The anterior fontanelle had opened and the sutures also seemed to be separating somewhat. The lesion in the right eye, according to the opinion of an ophthalmologist, was an embryoma. On May 7, 1948, 7 weeks after operation, the infant was discharged from the hospital weighing 6 lbs.

His development since has been progressive and, as far as can be made out, normal. On August 3, 1948, his weight was 10 lbs. 12 ozs. The head was seen to have grown proportionately to the rest of the body. The anterior fontanelle admitted a finger-tip and the skull sutures were palpable. The reflexes and reactions seemed within normal limits for the age. The wound was solidly healed and there were no digestive or bowel disturbances.

DISCUSSION

Atresia of the intestine may occur in the form of an internal diaphragm or, more commonly, the intestine ends as a blind sac discontinuous with the distal bowel or connected only by a fibrous strand. Multiple atresias, while less common, occur. Proximal to the obstruction the bowel is markedly distended; distally, it is very small and contains no gas. The cause is an arrest in development during the second or third month of fetal life when the bowel which has become filled with rapidly proliferating epithelium normally reestablishes its lumen.

The actual incidence of the lesion is difficult to appraise because of incomplete reporting and missed diagnoses. Without attempting to drain the literature, a few references will help to give an impression of the incidence. In 1922 Davis and Poynter collected 401 cases and added one. This figure included 134 atresias of the duodenum and 67 of the colon. Webb and Wangenstein in 1931 estimated the incidence of atresia and stenosis combined as about 1 to 20,000 infants. Including all the intestine below the pylorus and above the rectum, they found about 500 cases reported previously and added 13 cases of atresia and 4 of stenosis. Cohen in 1941 stated that since Webb and Wangenstein's paper at least 150 additional cases had been reported but they included all varieties of intestinal obstruction. In the same year Ladd and Gross reported 52 cases of intestinal atresia, only two of which were in the colon. Since 1941 individual cases or small series have been added by Duckett; by Glover, Smith and Eitzen; by Impink and Clammer; by Stock and Cannon; by Miller, Greengard, Raycraft and McFadden; by Erb and Smith; by Arnheim; by Biggs and Pontius; by Darne; by Judd, and others.

Symptoms usually begin on the first day of life. Vomiting is the presenting symptom. Persistent vomiting leads rapidly to severe dehydration. The vomitus almost always contains bile. The character of the vomitus will of course depend upon the level of the obstruction. While the stools may resemble normal meconium they are usually smaller and dryer. Farber's test which demonstrates the presence or absence of vernix cells in a stool specimen is of great usefulness in determining whether a complete obstruction exists. Abdominal distention may be present if the obstruction is low but may be difficult to evaluate if the atresia is high. Peristaltic waves crossing the abdomen may be seen but these are not constant.

A plain roentgenogram of the abdomen can give considerable information. No air is present in the intestine distal to the site of obstruction and if this be high, a dilated air-filled stomach, duodenum and upper jejunum with no air in the lower part of the field may establish the diagnosis. With ileal obstruction, multiple dilated loops with fluid levels are seen which are characteristic of intestinal obstruction but not necessarily of ileal atresia. Barium may be given to aid in the roentgen diagnosis but the danger of aspirating vomited barium accompanies such a procedure.

Surgical relief of the obstruction offers the only hope for survival. Otherwise, the markedly distended proximal blind loop undergoes necrosis, and perforation with peritonitis supervene. The procedure of choice is a side-tracking anastomosis. Naturally, resection of any nonviable bowel must be performed. Enterostomy should not be done as it always results in death due to rapid dehydration. Technically the operation presents certain difficulties due to the friability and distention of the proximal loop and the extreme narrow caliber of the distal. An excellent description of the technic may be found in Ladd and Gross.

Of equal importance to the operation is of course the pre- and postoperative handling of the infant. Fluid balance must be maintained and this is not an easy

procedure since it entails the administration of parenteral fluid several times a day. Blood and other supportive therapy must be utilized as indicated. A gastric tube should be introduced before the operation and should be continued after operation for as long as necessary. Operation should not be performed until the general condition has been brought to its best possible level even if this involves some delay. The infant frequently is not able to take feedings for several days after surgery and the stomach should be kept empty as long as vomiting persists. Feedings must be started carefully and increased judiciously. In all, the immediate period of postoperative care is one which requires painstaking and meticulous attention.

The mortality without operation is 100 per cent. According to Davis and Poynter, without operation death occurs after an average of 5 days when the obstruction is at the duodenal level as compared to 8 days at the colonic level. Even with surgery the mortality is quite high although it has lessened appreciably with modern techniques and improved preparation and postoperative care. The first recovery is credited to Fockens in 1911 who established a side-to-side anastomosis seven days after birth in a case of atresia at the junction of the middle and lower thirds of the small intestine. Of the 500 cases collected by Webb and Wangenstein only 10 recovered following surgery although the number operated upon is not given. Seven of Ladd and Gross' series of 52 cases treated surgically recovered. In Arnheim's report in 1945 he states that only 11 cases treated surgically to that date survived the procedure. While a number of individual successes have been added since then it is apparent that the mortality is still quite high.

The case reported here is of particular interest because it illustrates that even in a three and one-half pound premature infant a successful result may be achieved. The clinical picture and roentgen findings were typical of high jejunal atresia. The picture was complicated by the presence of icterus and other congenital defects were present in the form of closed fontanelles, an embryoma of the right eye, and an incompletely rotated colon. The last was not obstructing but required liberation and reduction before the primary pathology could be exposed and jejuno-jejunostomy performed.

SUMMARY

1. A case of jejunal atresia in a three and one-half pound premature infant, successfully treated by jejuno-jejunostomy, is presented.

2. Congenital intestinal atresia is briefly reviewed.

BIBLIOGRAPHY

1 Arnheim, E. E.: Congenital Ileal Atresia with Gangrene, Perforation and Peritonitis in a Newborn Infant. Am. J. Dis. Children, **69**: 108, 1945.

2 Biggs, A. D., and G. V. Pontius: Intestinal Obstruction of the Newborn. J. Pediat., **30**: 306, 1947.

3 Cohen, P.: Congenital Intestinal Obstruction. Am. J. Dis. Children, **62**: 135, 1941.

4 Darne, F.: Congenital Atresia of the Small Intestine. Brit. J. Surg., **34**: 315, 1947.

5 Davis, D. L., and C. W. M. Poynter: Congenital Occlusions of the Intestine with Report of a Case of Multiple Atresia of the Jejunum. Surg. Gynec. & Obst., **34**: 35, 1922.

[6] Duckett, J. W.: Intestinal Obstruction in the Newborn. Ann. Surg., **116**: 321, 1942.
[7] Erb, W. H., and D. C. Smith: Atresia of Small Intestine. Ann. Surg., **120**: 66, 1944.
[8] Farber, S.: Congenital Atresia of the Alimentary Tract: Diagnosis by Microscopic Examination of Meconium. J. A. M. A., **100**: 1753, 1933.
[9] Fockens, P.: Ein operation geheilter Fall von Kongenitales Dünndarmatresia. Zentralbl. f. Chir., **38**: 532, 1911.
[10] Glover, D. M., S. Smith and O. Eitzen: Multiple Atresia of the Small Intestine. Ann. Surg., **116**: 1942.
[11] Impink, R. R., and G. R. Clammer: Atresia of the Duodenum. Ann. Surg., **116**: 321, 1942.
[12] Judd, J. R.: Atresia of the Ileum. J. Pediat., **30**: 679, 1947.
[13] Ladd, W. E., and R. E. Gross: Abdominal Surgery of Infancy and Childhood. Philadelphia, W. B. Saunders, 1941.
[14] Miller, E. M., J. Greengard, W. B. Raycraft and I. McFadden: Congenital Atresia of the Duodenum and the Ileum. Am. J. Dis. Children, **66**: 272, 1943.
[15] Stock, F. E., and D. A. Cannon: Atresia of the Small Intestine. Brit. J. Surg., **31**: 96, 1943.
[16] Webb, C. H., and O. H. Wangenstein: Congenital Intestinal Atresia. Am. J. Dis. Children, **41**: 262, 1931.