Operative Management of Congenital Intestinal Atresia*

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THE PROBLEM OF intestinal atresia does not need to be reviewed from the standpoint of incidence, methods of diagnosis, related anomalies and mortality figures. Textbooks and articles in the literature cover these quite well. A review of the literature does reveal a need for an analysis of the operative methods and postoperative problems encountered in the treatment of infants with these anomalies.

This discussion reviews the experience of others, and presents my experience with ten cases treated during the last four years. The study has been confined to lesions of the jejunum, ileum, and colon. Duodenal lesions are omitted. The anatomical relationships of the duodenum are such that the technical considerations of lesions in this region are distinctly different from those of lesions below the ligament of Treitz.

The literature was reviewed with the purpose of finding, if possible, the reasons for both success and failure. There were only a few articles which analyzed a series of cases in sufficient detail to be of value.[5-8, 11] Up to about 1948 there did not seem to be any common denominator to success. Many methods were tried, and one received the impression that a favorable outcome was rare and fortuitous. During the last six years, however, the outlook for infants with these anomalies has become much brighter, and it appears that we are no longer in an era of chance success. Several reports[6, 12] indicate that great strides are being made in the reduction of the mortality, and that successful outcomes are becoming the rule rather than the exception.

The clearest analysis of the causes of failure was made by Louw[8] in 1952. He studied 44 patients with atresia below the duodenum between the years 1926 and 1951. Twenty-five of these died "early deaths" within 48 hours of operation, and 14 died "late deaths," three days to five weeks following operation. The "early deaths" were attributed directly to circulatory collapse and respiratory embarrassment, and indirectly to late diagnosis. The "late deaths" in those treated by primary anastomosis were caused mainly by postoperative ileus and disruption of the suture line; and in those treated by enterostomy, with or without an anastomosis, the late deaths were due to disturbances of nutrition and fluid balance. Glover and Barry[5] listed numerous factors which contribute to the high mortality rate, among them being late diagnosis and operation, inadequate supportive therapy, and inadequate or unphysiological operative management. Gross[6] states that three-quarters of their deaths are due to postoperative obstruction caused by peritoneal adhesions. It is interesting that both Louw and Gross, who report the largest series, state that associated anomalies are a minor factor in the mortality.

The main problem which must be faced today is not that of the "early death." Increased awareness of the condition has led to earlier diagnosis. This combined with our increased knowledge of supportive therapy and improvements in anesthesia, should

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make the "early death" uncommon. The major problem, then, is obviously one of establishing intestinal continuity and restoring intestinal function within a reasonable period of time during which supportive therapy is effective.

The ability to support an infant through a difficult preoperative period, operation, and postoperative period has not only reduced the "early deaths" to a minimum but has allowed a modification of the previous concepts that concerned the operative management of intestinal atresia. For many years it was believed, with justification, that any type of exteriorization of the small intestine of an infant was incompatible with life. Recently there has been a trend back to intestinal exteriorization, with strikingly good results. In 1952 Gross, using a Mikulicz type of exteriorization which is closed in a week or two, reported six recoveries in seven cases of ileal atresia. In 1950 Duckett[4] discussed five consecutive cases of jejunal and ileal atresia treated by Nichols in Los Angeles. He was using a separated double barrelled enterostomy, the fluid from the proximal end being collected and injected into the distal loop. An enteroenterostomy was then done in about ten days.

In addition to these successes with exteriorization more reports are appearing of success with primary anastomosis, but no recent series in which this method was used is available. The ten cases presented here, with one exception, were all treated by primary anastomosis. Experience with the first few infants led to the adoption of two principles which are the basis for the technic now being employed in the management of this anomaly. These two principles which are felt to be so important in obtaining consistently good results are: decompression of the large loop proximal to the anastomosis, and end-to-end anastomosis.

PROXIMAL DECOMPRESSION

The management of these patients has led me to believe that the single, most important factor in the success or failure of any type of procedure for intestinal atresia is a period of adequate decompression of the greatly dilated proximal loop. The musculature of this loop is so stretched and attenuated that a period of decompression is essential if it is to regain its ability to contract effectively. There are numerous reports in the literature of infants who have died because of lack of function of the proximal intestine in spite of anatomically patent anastomoses. As mentioned previously, Louw lists "postoperative ileus" as a major cause of failure. Case 3 in this series is an excellent example of the functional failure of the large proximal loop simply because the muscle fibers were not given the opportunity of regaining tonicity.

Providing for this period of decompression is much more important than the manner in which it is accomplished. Exteriorization procedures are undoubtedly successful because they fulfill this requirement. There does not seem to be any other logical explanation, since these procedures are in essence an anastomosis ten days or so after the initial exteriorization. The only plausible accomplishment of a period of exteriorization is the recovery of effective contractibility of the proximal loop. In contrast to exteriorization, decompression in this group of patients has been effected by means of a polyvinyl enterostomy tube proximal to a primary anastomosis. The tube has been effective in providing continual decompression, and the results have been gratifying.

The use of such an enterostomy tube is not new. Four successfully treated cases have been reported in which such a tube was employed.[1, 3, 9] The first of these was as early as 1933,[1] and the last two were reported by Miller[9] in 1943. Glover and Barry[5] in 1949 pointed out the importance of a Witzel type enterostomy to control distention. Again in 1952, Miller[10] stressed the value of a tiny tube proximal to the anastomosis to take the tension off the suture line. The real rationale for its use, however, has never been clearly stated or emphasized.

END-TO-END ANASTOMOSIS

Realizing that strong effective peristalsis of the proximal loop was essential to overcome the resistance of a small anastomosis and also to dilate this anastomosis, I felt that an end-to-end anastomosis would be more desirable than any other type. Propelling the intestinal stream directly into the anastomosis should be more effective than dissipating some of that propelling force into the blind pouch of a side-to-side anastomosis. The often reported complication of dilatation of the proximal blind pouch in a lateral anastomosis attests to the fact that intraluminal pressure produced by peristaltic activity is wasted in this pouch.

In 1953 Clawson,[2] in a review of this subject, found overwhelming evidence in favor of the end-to-end anastomosis as the most physiological type of intestinal union. The usual complications of side-to-side anastomosis which he cited, namely, dilatation of the blind pouch, ulceration, hemorrhage, and anemia, were all seen in Case 2, and may necessitate further surgical measures.

In addition to the fact that an end-to-end anastomosis is more physiological and avoids the aforementioned complications, it is also more receptive to the total propelling force, a fact which is so important in this particular problem. The theoretical advantages of this type of anastomosis are supported by a distinct clinical impression that end-to-end anastomoses in this series have functioned much more readily and efficiently than the lateral anastomoses.

METHOD

A soft rubber catheter is placed in the stomach, and a plastic cannula in an ankle vein. The abdomen is opened through a right rectus incision unless a left-sided colon lesion is suspected. This can be ascertained by a barium enema if the plain film indicates that this is a possibility. The proximal blind loop of intestine which usually measures 4 to 6 cm. in diameter is easily identified and delivered. A 7 or 8 mm. incision is made directly in the end of this dilated loop, and as much of the proximal bowel as possible is decompressed with suction. This immediate decompression permits restitution of the intestinal wall to a relatively normal thickness, and improvement in its circulation while the distal intestinal tract is being examined. The distal loop of intestine usually measures 5 to 7 mm. in diameter. Examination may reveal an isolated area of atresia or multiple areas with sausage-shaped pieces of intestine between. Distal to any gross defects in the bowel, and without opening the bowel, saline is injected and followed visually throughout the entire remaining bowel down to the rectum. The saline should move along easily, and its passage avoids overlooking another area of atresia or stenosis. The blind end of the distal loop is then cut off transversely, leaving an opening which can be stretched gently up to 7 or 8 mm. in diameter. All isolated segments of atretic bowel are resected, as there is some question as to the ultimate fate of these blind pouches.

A two-layer end-to-end anastomosis is then made between the opening in the large proximal segment and the small distal segment. The defect in the mesentery is closed, with care not to kink the anastomosis. About 10 cm. proximal to the anastomosis a No. 13 needle is inserted into the bowel, and size .065 inch polyvinyl tubing is threaded through it until 5 cm. lie in the lumen of the large dilated loop. The needle is withdrawn and the tubing secured by means of a single purse string suture of silk (Fig. 1).

The intestines are replaced in the abdominal cavity, with the large loop lying anteriorly. The polyvinyl tube is brought out either through the wound or through a separate stab wound, depending upon which avoids kinking. The wound is closed and a dressing applied, through which the tube protrudes. A separate dressing incorporating the tube is then placed on top of the first. This allows easy access to the tube if necessary,

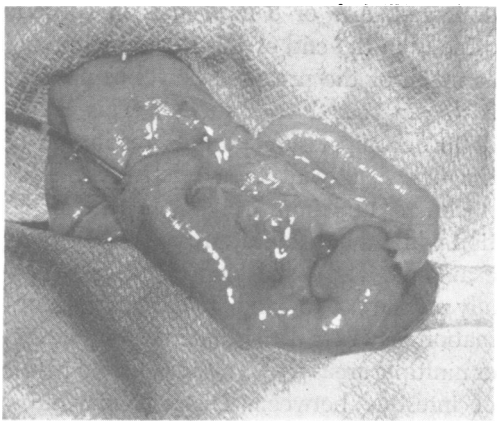

FIG. 1. Primary anastomosis of large proximal loop and small distal loop. Polyvinyl enterostomy tube is seen entering the dilated bowel 8 cm. proximal to the anastomosis.

and protects it from inadvertent removal. At least 12 cm. of tubing should be left outside the abdominal cavity. In one instance it was cut too short, and slipped back into the bowel.

POSTOPERATIVE COURSE

It is to be expected that the diminutive stoma will be temporarily closed because of edema. Constant gastric suction is maintained to prevent as much gas as possible from entering the intestinal tract. The enterostomy tube is permitted to drain spontaneously into the dressing, thus preventing pressure building up in the proximal loop and thereby allowing the musculature of this loop to recover from its prolonged stretching. Gastric suction is discontinued at 48 hours, and feedings are started slowly at 72 hours. If any evidence of obstruction occurs it is most likely due to persistent stomal edema or inefficient contractions of the large proximal loop, since aspiration of this loop through the enterostomy tube immediately stops the symptoms (Cases 8, 9 and 10). Most of these infants have shown evidence of a patent anastomotic stoma in 49 to 72 hours, and feedings have then been increased progressively. The enterostomy tube has functioned as a safety valve protecting the musculature of the proximal loop from stretching and decompensating as evidenced by the drainage, which has ranged from a total of 5 ml. in two cases to as much as 100 ml. in another. Most of the tubes have been removed by the tenth or twelfth day, at which time the infants are eating satisfactorily and displaying no signs of obstruction.

All of these infants have, to a greater or lesser degree, exhibited a period of frequent small loose stools, sometimes as many as 15 a day. This period has varied from a few days to several weeks; the longer periods being the more common. This was seen regardless of the location of the atresia. Changes in the frequency and type of feedings did not alter this phenomenon. All of these infants have eventually developed normal intestinal function.

Penicillin and streptomycin have been used as the antibiotics of choice. There have been no wound infections or serious pulmonary complications. In a few patients thush was quite troublesome, and because of this antibiotics are now used for only 72 hours following the operation.

As noted previously, Gross[6] has stated that the main cause of death in his cases is postoperative obstruction due to peritoneal adhesions. This has been a serious problem not only in this group of patients but in all infants who have been subjected to an extensive intra-abdominal procedure. Two of these infants required re-operation because of adhesions (Cases 2 and 10), and one of them died as a result. One has had several temporary bouts of partial obstruction since leaving the hospital (Case 8), and another has developed a mild celiac-like syndrome (Case 9). It is well known that a clinical syndrome simulating celiac disease can be produced by a chronic partial intestinal obstruction, and it is highly probable that this last infant is an example of this. Until a method is found of preventing peritoneal adhesions this complication will un-

doubtedly cause more difficulty and higher mortality rates than any other single factor.

DUODENAL LESIONS

Atresias and stenosis of the duodenum have not been included in this study because the technical problems are different. A bypassing procedure is preferable to either a primary anastomosis or an exteriorization procedure. Decompression and rest of the stretched proximal duodenum is easily accomplished with an indwelling gastric tube, and in part probably explains why the survival rates of these high lesions have always been better than with the lower lesions.

RESULTS

There have been three deaths in this series of ten consecutive patients. In Case 3, the failure was undoubtedly due to inability of the proximal loop to contract effectively because of persistent distention. Case 5 presented a situation which was hopeless because of the extensive necrosis and infection. In Case 10 the method used in treating the atresia was successful, but the infant succumbed to obstruction owing to adhesions, a complication which offers a real challenge if abdominal surgery in infants is to continue to improve.

Of the seven successful cases, six infants are growing and developing normally. The follow-up has been from six months to four years. The other infant is exhibiting intermittent symptoms of partial intestinal obstruction, probably on the basis of peritoneal adhesions, and may require further surgical therapy.

Case 1. This 1-day-old female infant had an atresia of the upper sigmoid colon treated by end-to-end anastomosis and tube cecostomy. The cecostomy tube was inadvertently removed on the third postoperative day. Her colon functioned poorly as evidenced by intermittent vomiting, distended abdomen, and passage of very small stools. On the 18th postoperative day she became completely obstructed at the site of the anastomosis, and because her condition was so poor, a tube cecostomy was done in preference to re-exploring the anastomotic site. This decompression rapidly improved her condition, she began to have normal stools, and in 7 days she was taking a full formula and gaining weight. The cecostomy tube was removed 11 days after its insertion, and the opening closed spontaneously. The period of decompression apparently allowed the proximal colon to return to an effective functional status. Her course thereafter was uneventful. She is now 4 years old, and in good health.

Case 2. This was a 2-day-old male infant with multiple atresias of the ileum. The atretic segments of ileum were resected and a side-to-side ileo-ileostomy was done, with no proximal decompression. On the fifth postoperative day a wound dihiscence was closed. He did fairly well except for abdominal distention and occasional vomiting for 20 days, at which time he became completely obstructed. Re-exploration revealed that the side-to-side anastomosis was patent but the disparity in size of the two ends had allowed the smaller end to bend at right angles over the large end where it had become adherent and obstructed. This was freed and replaced in the abdomen, hoping it would not recur. He did well for 4 months, at which time he developed a celiac-like syndrome, and failed to gain weight. It was felt that his symptoms were probably on the basis of chronic partial obstruction, and exploration was done. The blind pouch of the proximal ileum had become tremendously dilated, and was adherent to many loops of bowel. This entire area, including the old anastomosis, was resected and another side-to-side ileo-ileostomy was done. Following this procedure he improved dramatically. At the age of 1½ years he had a severe gastro-intestinal hemorrhage, and during the following year had several others. These were all treated conservatively since the source of the bleeding was uncertain; however, roentgenograms revealed that the blind pouch of the lateral anastomosis was greatly dilated, and this probably was the source of the hemorrhage. For the last year and a half he has been asymptomatic and developing normally.

Case 3. This 1-day-old female infant had multiple areas of atresia of the terminal ileum. These were resected and a side-to-side ileo-colostomy was done. Eleven days later it was obvious that she was still almost completely obstructed. Re-operation revealed that the proximal loop was still as dilated, and the colon still as small as at the original procedure. The area was resected and another side-to-side anastomosis was done. Examination of the specimen revealed that the anastomosis was quite patent. She showed no evidence of moving anything into her colon, and 8 days later she was again

explored. The anastomosis was again patent, and it was felt that she probably had a congenital neurological defect of her colon such as that seen in the rectum in Hirschsprung's disease. The proximal half of her colon was resected, and end-to-end ileocolostomy was then done. Shortly after the procedure she vomited in spite of a tube in her stomach, and aspirated. She developed a severe pneumonitis and died 2 days later. Autopsy revealed that the last anastomosis was patent, as were the first two, and that the ganglia in the colon were normal. Subsequent experience has led us to believe that the large proximal loop never had the functional ability to propel its contents, since the musculature was never allowed to regain its tone by some type of decompression.

Case 4. This was a 1-day-old male infant with a complete diaphragm across the lower jejunum. The proximal bowel measured 5 cm. in diameter, and the distal bowel 5 mm. in diameter. A longitudinal incision was made, extending 1 cm. on both sides of the diaphragm. The diaphragm was removed and the bowel closed transversely, giving an adequate stoma. He began to have a few stools but showed signs of marked obstruction. On his tenth postoperative day he was re-explored. The stoma was quite patent but 1 cm. distal to it was a constricted area of bowel with a tiny lumen and rigid wall. Apparently when the distal bowel had been checked for patency the needle had been placed in the bowl distal to this stenotic area. The area was by-passed by means of a jejuno-jejunostomy. Since there was such tremendous dilatation of the proximal jejunum, and because of our experience with Case 3, we decided to use an enterostomy tube. A .065 inch polyvinyl catheter was placed in the jejunum 8 cm. proximal to the anastomosis. Following this procedure the infant had no more obstructive symptoms and the enterostomy tube was removed on the tenth day. He had persistent diarrhea for 6 weeks, which then stopped. He is now 2 years old, and has gained weight at a normal pace.

Case 5. This 2½-day-old female infant was explored and found to have an atresia of the jejunum. There was a volvulus involving a large proportion of the bowel, both proximal and distal to the atretic area, with marked necrosis, perforation, and meconium peritonitis. It was almost impossible to recognize what had happened. Her condition was so critical that the necrotic bowel was rapidly removed and the two ends exteriorized. She did not respond to vigorous supportive therapy, and died 24 hours later.

Case 6. This 2-day-old male infant had an atresia high in the ileum. The large proximal loop had perforated, and the peritoneal cavity was a mass of old fibrous adhesions and contained some brown fluid. Ten centimeters of the large proximal bowel were resected because of questionable viability. The end of the small distal bowel was then anastomosed to the side of the large proximal bowel. A polyvinyl tube enterostomy was placed 8 cm. proximal to the anastomosis. The infant showed no further signs of obstruction, and the tube was removed from the intestine on the tenth day. He had the usual difficulties with diarrhea, which finally subsided. He is now 1½ years old, and has developed normally.

Case 7. This 1-day-old female had multiple atretic areas of the ileum. They were resected and an end-to-end anastomosis was done between the large dilated proximal bowel and the small distal bowel. A tube enterostomy was used. She had no obstructive difficulties and the tube was removed on the tenth postoperative day. She had a very uneventful course, and at the age of 9 months is in good health.

Case 8. This 1-day-old male had an atresia of the terminal ileum. The remaining 1-cm. of terminal ileum distal to the atresia and the cecum were resected, and an end-to-end ileocolostomy was done. A tube enterostomy was placed proximal to the anastomosis. By the fourth postoperative day he was taking feedings well and having stools. During the next 5 days, however, he showed progressive evidence of poor function of the large proximal loop, namely, intermittent vomiting and increasing abdominal distention. Feedings were stopped and the enterostomy tube was aspirated to decompress the dilated proximal loop. This immediately stopped the obstructive symptoms. The large loop was permitted to rest for 24 hours, following which formula was started again, slowly, and no further obstructive symptoms developed. The tube was removed on the 15th day. He gained weight well but had two episodes of partial intestinal obstruction associated with systemic infections. When the infections were controlled his obstructive signs disappeared spontaneously.

Case 9. This 8-hour-old male infant had an atresia of the lower jejunum or upper ileum. There had apparently been a perforation *in utero* as the peritoneal cavity was filled with dense adhesions, but the location of this perforation could not be demonstrated. An end-to-end enteroenterostomy was done, and a polyvinyl tube enterostomy was placed proximal to the anastomosis. The Levin tube was removed on the second postoperative day, and the infant started to vomit immediately. It was found that the enterostomy tube was plugged with blood clot. This was opened and aspirated, and in addition

Levin tube suction was re-instituted for 2 more days. Gastric suction was then stopped, and no further vomiting occurred. Oral feedings were started, and slowly increased. The enterostomy tube drained moderate amounts of green liquid for a few days, which then stopped and the tube was removed on the twelfth postoperative day. His course was similar to the others in that he had a long period of diarrhea which finally was controlled, and he was discharged gaining weight. He developed a severe bronchiolitis several weeks after discharge, and following this he began to exhibit a mild celiac-like syndrome probably on the basis of partial intestinal obstruction. If this persists and interferes with growth he may need re-operation. The most likely cause of this partial obstruction is peritoneal adhesions.

Case 10. This 6-hour-old female infant had an atresia of the upper ileum. An end-to-end ileo-ileostomy was done with a polyvinyl tube enterostomy proximal to the anastomosis. She did quite well for 6 days, at which time she vomited. The dressings were opened to aspirate the enterostomy tube, and it had disappeared. Roentgenograms showed it had slipped into the lumen of the large loop. She continued to vomit small amounts of each feeding, and on the tenth postoperative day, she was re-explored because of this evidence of poor function of the large proximal loop and the loss of the tube for decompression. The tube was recovered from the proximal loop and a new enterostomy tube was inserted. Feedings were again started 24 hours later, and she had no further obstructive difficulty; however, her abdomen remained moderately distended and her weight gain was not satisfactory. On the fourth day following the second procedure intestinal fluid began to leak from the wound around the enterostomy tube. The tube was removed and the drainage ceased spontaneously. At the age of 2 months she developed a severe bronchiolitis, and while recovering from this, she became completely obstructed. Exploration revealed marked peritoneal adhesions, with many points of possible obstruction. Extensive lysis had to be done and she died shortly after this procedure. No autopsy was obtained. It was felt that this procedure was too shocking for an underweight infant just recovering from a severe pulmonary infection.

DISCUSSION AND CONCLUSIONS

A method of management of intestinal atresias below the ligament of Treitz has been presented. It is felt that the most important factor in the successful treatment is a period of adequate decompression of the distended loop proximal to the anastomosis. This can be accomplished either by the method presented here of primary end-to-end anastomosis with a proximal tube enterostomy, or by an exteriorization procedure. The former has certain advantages. It is technically easier to perform, especially for a surgeon who only occasionally is confronted with this problem. It does not require a second operation. Most important, however, is that it avoids the long period of supportive therapy which is necessary for an infant with an exteriorized bowel. Such support requires an abundance of skilled nursing and house staff personnel which is not available universally to practicing surgeons.

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