Management of Meconium Ileus:

Resection, Roux-en-Y Anastomosis and Ileostomy Irrigation with Pancreatic Enzymes *

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INCREASING interest in the management of intestinal obstruction due to inspissated, abnormal meconium in the newborn has occurred in the last ten years. This interest has been encouraged by our advancing knowledge of the medical management of pancreatic fibrocystic disease, the underlying disease in all such infants.

Although fibrocystic disease of the pancreas is still far from a conquered problem, many youngsters, relieved of meconium obstruction in infancy, are growing well with the addition of pancreatic extracts to their diets, and measures to control or prevent the commonly associated respiratory problems. Numerous reviews [1, 4, 6] dealing with pancreatic fibrocystic disease (a poor name for the over-all disease since intestinal, bronchial, salivary, and sweat glands are also affected) have been published outlining the current methods of handling these various non-surgical problems.

All infants born with fibrocystic disease of the pancreas have a deficiency of enzymes normally released into the intestinal tract by the pancreas. It is this deficiency which, in utero, leads to the abnormal character of the meconium, causing it to be tenacious, putty-like, and hence obstructing. This meconium, in vitro, or in the infant's bowel, is liquefied by the addition of either human or animal pancreatic juice. Unfortunately, this liquefaction takes place only gradually.

It is difficult to explain why less than 10 per cent of the infants born with fibrocystic disease have intestinal obstruction at the time of birth. This complete intraluminal obstruction, when it occurs, has a typical pathologic appearance at operation (Fig. 1). The bowel distal to the obstruction is generally empty and collapsed, although fully capable of normal function and size when the fecal stream is established. Proximal to the collapsed bowel, the ileum frequently resembles a strand of beads as the bowel wall conforms to the shape of the intraluminal, yellow-white, waxy pellets of abnormal, bile-free meconium. Proximal to these pellets the bowel dilates rather abruptly to a diameter of two to four centimeters, and is tightly packed with either green or black, putty-like, sticky meconium which adheres not only to itself but to the intestinal mucosa as well. This obstructing segment varies from a few cm. up to 30 cm. in length. It keeps its indented shape when squeezed, and the bowel wall is of normal thickness. The bowel immediately above this segment is hypertrophied, and of even greater diameter. This segment is maximally dilated, being at times five centimeters in width. Within its lumen is black liquid meconium overlaid with air in the various loops. This distention with gas and liquid meconium usually extends back into the jejunum, but at times can extend as far as the duodenum.

These classical findings, seen at operation, readily explain the preoperative dis-

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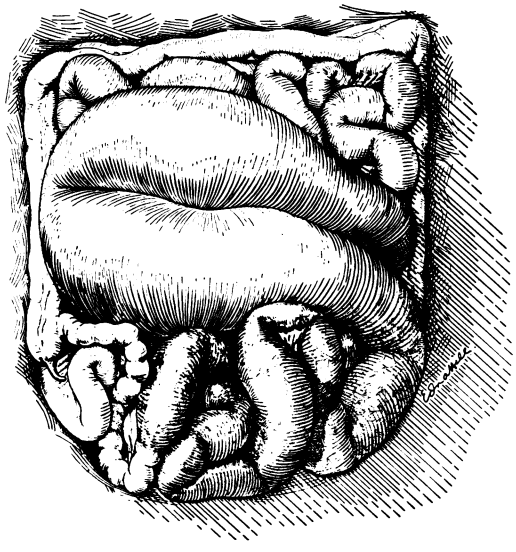

FIG. 1. Typical appearance of the intestines in meconium ileus (see text for description).

tention, the vomiting, and the complete lack of stools or merely the passage of several small waxy pellets. The dilated loops seen through the thin, shiny abdominal wall, may be soft and compressible if they are jejunal loops, or firm and capable of being indented if in the mid- or lower ileum. Roentgenograms of the abdomen characteristically show the obstructed loops of varying size, and often a mottled appearance of air bubbles forced into the putty-like more radio-opaque meconium. A barium enema, although not needed for diagnosis, will show the so-called "microcolon," a misnomer and better described as a small, as yet unused, colon.

Procrastination, watchful waiting, gastric suction, rectal examinations, enemas of saline or enemas containing pancreatic enzymes will not relieve the obstruction if true meconium ileus is present, and operation should be planned and executed without delay.

Many methods have been recommended for relief of the obstruction. Ileotomy with manual emptying of the bowel is usually impossible due to the tenacious character of the meconium. Ileotomy with irrigation of saline has been successfully executed by Hiatt,[3] and Olim [5] has reported success using dilute hydrogen peroxide as the irrigating solution. Both these methods have been successfully employed by us, but we have not been happy with their use. Neither saline nor peroxide dissolves the meconium, and so it is only of help in partially separating the meconium from the mucosa of the bowel. The bowel must then be milked to empty the meconium through the ileotomy. This is quite traumatic, since it must usually be repeated many times. In our experience, this method is poorly tolerated by these infants who already are in physiologic imbalance because of their obstruction.

Resection of the obstructing segment of small bowel with primary anastomosis has been discouraging even though occasionally successful. This may require the removal of too long a segment of bowel, and after the upper dilated part of the bowel is emptied of its liquid meconium and gas, one is left with a tremendous disparity between the size of the upper and lower limbs of bowel for anastomosis. Such anastomoses in the newborn, even when end-to-side, leave much to be desired.

Loop ileostomy with postoperative irrigation of the distal limb with pancreatic extracts to liquefy the meconium, followed later by an intraperitoneal closure of the ileum, has also been used. There can be many complications of this staged method. Wound problems around this type of ileostomy are common since the abdomen must be closed around the exteriorized loop of ileum in the presence of still obstructed intestinal loops. Fluid and electrolyte losses through such an ileostomy are undesirable, although with diligence and hourly reevaluation, the infant's replacement needs can be met successfully. A second intraperitoneal operation to close the ileostomy is another major, relatively dangerous, operation in any infant.

Gross [2] has, in recent years, been exte-

riorizing the maximally distended and hypertrophied loop of bowel, constructing a Mikulicz type of ileostomy, with resection of the exteriorized loop immediately upon closure of the abdominal wall. Later the distal bowel is opened by irrigation with pancreatic extracts. After the intraluminal obstruction is relieved, a specially constructed spur crushing clamp is applied which gradually causes necrosis of the common wall between the two limbs, and, finally, an extraperitoneal closure of the ileostomy is carried out. This method of management has saved a number of these infants and has many advantages. It allows a safer and easier closure of the abdominal wall with exteriorization and resection of the large loop, and contamination of the peritoneal cavity is avoided. It does not, however, eliminate the problems of fluid and electrolytes associated with an ileostomy.

The need for the spur crushing clamp is a serious drawback to this method of management. Its insertion into the two limbs requires care, and can be difficult if the two limbs angulate just within the peritoneal cavity. Frequently it must be applied a second time to insure a deep enough common opening below the level of the parietal peritoneum. Often the tightening of the clamp causes a worrisome, transient fever and tachycardia, and the infant refuses or regurgitates feedings while the clamp is in place. The clamp also creates a nursing problem since it protrudes above the abdominal wall and its rough handling can tear or disrupt the ileostomy.

The closure of the ileostomy, being extraperitoneal, certainly is safer than an intraperitoneal anastomosis, but it is a procedure that requires another general anesthesia with its possible complications in the presence of the abnormal bronchial secretions in such infants. The closure necessitates a bloody dissection, and, hence, another intravenous cut down and transfusion of blood usually is necessary. Wound complications are common and although corrected

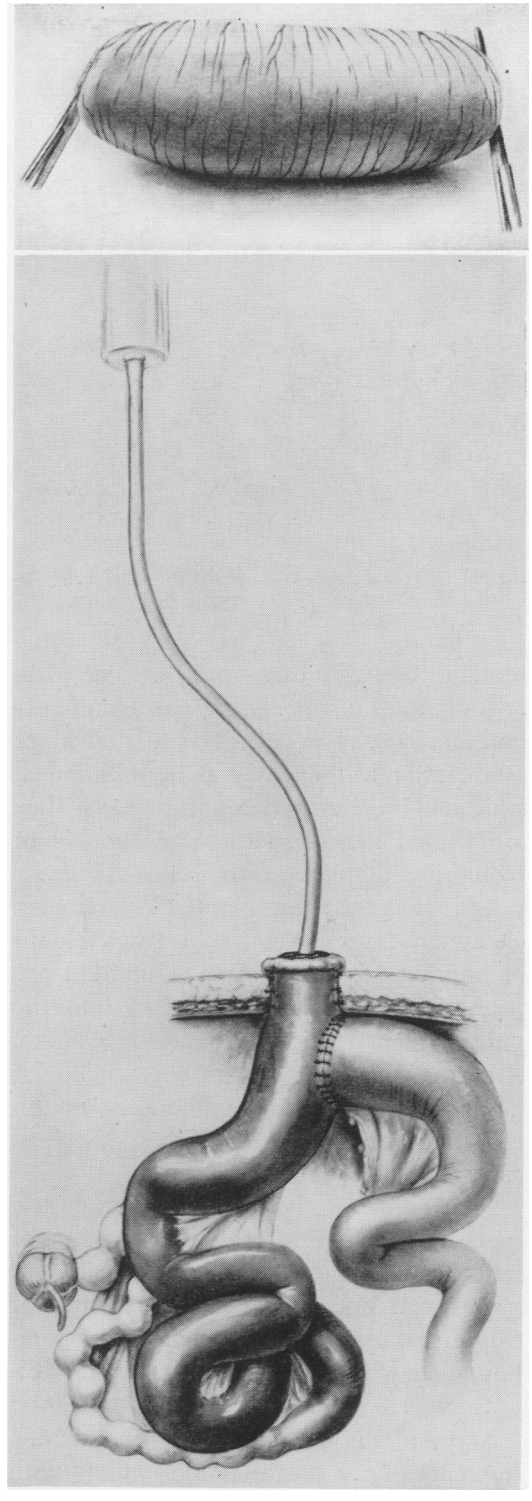

FIG. 2. Method of managing meconium ileus by resection, Roux-en-Y anastomosis, ileostomy, irrigation with pancreatic enzymes and secondary extraperitoneal closure.

in time, can increase the morbidity and prolong the period of hospitalization.

We have recently used a method, to our knowledge previously unreported, that has been successful in relieving the obstruction of meconium ileus (Fig. 2). A Roux-en-Y anastomosis with ileostomy is performed and, although not without its dangers, incorporates the safety of partially removing the maximally distended and hypertrophied segment with the establishment of a safety valve. The single limb ileostomy allows instillation of pancreatic extracts into the distal, still obstructed, ileum. Once the bowel lumen has been cleared of abnormal meconium, the ileostomy being against the peristaltic stream, is often non-functioning and so there is no need for early closure. There is, of course, the hazard associated with an intraperitoneal anastomosis, but the anastomosis can be made between bowel segments large enough to suture with ease, and be certain an adequate lumen is present.

THE METHOD

With a polyethylene catheter in an ankle vein, and type-specific blood available, the abdomen is opened through a mid-abdominal, right transverse incision. The diagnosis is confirmed and the bowel examined to rule out other anomalies.

A segment of the maximally distended hypertrophied ileum is resected, and without attempting to empty the remaining bowel, the end of the upper is anastomosed to the side of the lower leaving a Roux-en-Y limb approximately four centimeters in length. The anastomosis is done between bowel segments which are quite hypertrophied and dilated. The procedure is carried out as an open anastomosis with an inner layer of 5-0 chromic catgut, and an outer layer using interrupted atraumatic 5-0 braided silk.

The Roux-en-Y limb is brought out through a stab wound above or lateral to the transverse incision. The meconium in the proximal ileostomy limb is emptied down to the anastomosis, allowing a patent lumen from mouth to ileostomy. A small #8 French, soft rubber catheter is threaded down into the distal ileum to allow postoperative instillation of pancreatic enzymes which will gradually liquefy the obstructing meconium. This catheter does not pass through the anastomosis, and generally need not remain in place for more than a day or so.

Human duodenal juice, kept frozen in a deep freeze unit, has been used for instillation into the ileostomy. Any extra duodenal juice taken during diagnostic duodenal drainage studies is saved for this specific purpose. Viokase® powder, U.S.P. pancreatin, a desiccated, raw animal pancreas preparation, is used (one-half teaspoon in 30 ml. of saline) if human duodenal juice is not available.

Instillations of pancreatic extract through the catheter in the ileostomy, and also rectal instillation using the same solution result in patency of the lower intestinal tract 24 or 48 hours postoperatively. When the lower bowel, distal to the anastomosis, is patent, the ileostomy generally ceases to function and so losses of fluid and electrolytes from this route cease. Therefore, there is no urgency to close the then non-functioning ileostomy. Closure can be performed at some future date, extraperitoneally at the level of the deep fascia. In fact, probably a mere resecting of the ileum down to the subcutaneous tissue level will allow the shrunken, nonfunctioning limb and skin to close by secondary intention.

This method was first used by us in October, 1955. A female infant (C. H., #55-6251) weighing 2,200 grams was operated upon at 44 hours of age. The preoperative diagnosis of meconium ileus was confirmed at operation. Ten centimeters of upper ileum were resected, and a single limb ileostomy with a Roux-en-Y anastomosis was constructed. Irrigation with human duodenal juice through the ileostomy and

rectum, unblocked the bowel distal to the ileostomy. After the third postoperative day, the ileostomy drainage lessened and stools were passed rectally. By the fifth poostoperative day, a Nutramigen® formula was tolerated by mouth and the ileostomy drainage stopped completely. The infant did well on antibiotics and oral alimentation until the twelfth postoperative day when coughing and mild respiratory distress first appeared. A roentgenogram of the chest showed areas of atelectasis and emphysema characteristically seen when the lungs become involved in a patient with fibrocystic disease. Unfortunately, the pulmonary disease progressed in spite of all efforts to liquefy the bronchial secretions, and the infant died a respiratory death on the sixteenth postoperative day.

At autopsy the pancreas was fibrotic and small. The bronchial tree contained tenacious mucus and bronchopneumonia was widespread. The Roux-en-Y anastomosis was well healed and the single-limb ileostomy was of smaller caliber than the functioning bowel.

This method of managing obstruction due to meconium ileus is still under trial. Although certainly not free of potential hazards, its valuable features seem to us to warrant its use in future cases. In the rare case where perforation has occurred leading to meconium peritonitis, this method, which requires an intraperitoneal anastomosis, might not be advisable.

SUMMARY

The patient with meconium obstruction associated with fibrocystic disease of the pancreas presents a challenge to the surgeon during the early neonatal period, and to the pediatrician during infancy and childhood.

The appearance of the intestines at laparotomy is described, and the various methods of surgical management reviewed. Our current method of resection, Roux-en-Y anastomosis, ileostomy, and postoperative irrigation with pancreatic enzymes is presented.

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