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# Intestinal Obstruction in the Neonatal Period

C. EVERETT KOOP

*School of Medicine and Graduate School of Medicine, University of Pennsylvania; The Children's Hospital, Philadelphia, Pa.*

## GENERAL CONSIDERATIONS

INTESTINAL obstruction in the newborn infant may seem at first consideration to represent a rather small segment of neonatal pathology. Yet it is responsible for most of the surgery in the newborn period and for the most discouraging mortality figures of any of the neonatal surgical entities within reach of therapy. The purpose of this discussion is to provide that perspective which could lead to an aggressive suspicion regarding intestinal obstruction in the neonatal period.

Almost all of the congenital anomalies incompatible with life yet amenable to surgical correction pose problems in intestinal obstruction or produce symptoms of it. Atresia of the esophagus, diaphragmatic hernia, atresia of the small and large bowel, imperforate anus, and extrinsic intestinal obstructions due to a variety of lesions, are all without question among these.

In addition to the obstructive lesions which are uniformly fatal when not corrected by surgery, a surprisingly large group of congenital defects may produce intestinal obstruction in the neonatal period. Such defects, to be sure, can exist without ever producing signs or symptoms or may become associated with intestinal obstruction in adult life, but a sufficient number cause trouble in the newborn period to warrant their enumeration at least.

Except for procedures for an occasional omphalocele or, rarely, for the relief of respiratory obstruction or urinary retention, most neonatal surgery in the pediatrician's experience is for the relief of

intestinal obstruction. Although this condition in neonatal life has long been recognized as a challenge to the surgeon, only in recent years has therapeutic progress been made. Unfortunately, the increasing success obtained by some surgeons has not been widely reported, and a 100 per cent mortality for certain lesions is still considered by some as a reasonable prognosis. Surgeons continue to quote statistics on the pioneer work (26) with neonatal intestinal obstruction, little realizing that these figures were accumulated 20 years ago and apparently unaware that increasing opportunities in the surgery of infancy have led to repeated success in the management of all varieties of intestinal obstruction in the newborn.

Intestinal obstruction in the neonatal period is, to be sure, a many-sided problem. In the first place, among those who care for the newborn, many do not appreciate the frequency of neonatal intestinal obstruction. Second, surgeons are rarely invited to observe the variations in physiology in the evacuation of the meconium in the newborn; they are presented with intestinal obstruction in advanced stages, and frequently treat such patients with little or no previous experience and with the expectation of defeat. Third, single successful case reports appear in the literature, while series of patients treated by surgeons with broad pediatric opportunities are accumulated until a definitive work can be written. The resulting criteria of success and standards of therapy are therefore low.

Because of the small number of lesions producing obstruction in the alimentary tract above the pylorus and because of the similarity of symptoms when they do occur, this discussion will include obstructions from the esophagus to the anus.

The mortality in neonatal obstruction is roughly directly proportional to the patient's age. Some infants, of course, cannot be saved, but the best opportunities lie in the first 2 days of life; thereafter, the mortality rises sharply, with survival in cases of obstruction for a week being rather uncommon. Most surgeons with experience in the management of intestinal obstruction prefer to operate as soon as the diagnosis has been made. But in advanced cases, a delay of several hours, during which fluids can be administered, plasma given, and the infant kept in an atmosphere of high oxygen concentration, may alter the surgical risk sufficiently to make a satisfactory outcome at least a possibility. However, if such preparation lasts more than a few hours, a point of diminishing return is reached at which more

benefit can be expected from the surgical procedure than from continuation of supportive measures.

In the literature, there are frequent references to patients considered too ill for operative procedures. The question arises whether the judicious use of supportive measures might not have altered the picture. The operative risk in all infants with intestinal obstruction is great but in the untreated patients with obstruction, the mortality is 100 per cent. Increasing experience in neonatal surgery indicates that the preoperative condition of a given patient must always be considered in reference to the best result obtainable in that infant, rather than in relation to the ideal result. The temptation to continue measures which seem to improve the infant's preoperative condition is strong. Although the use of several hours for this type of support is to be earnestly recommended, what cannot be accomplished in that time is usually unobtainable and the continued delay of surgery diminishes the chance for success. One learns to be content with any preoperative improvement that can be achieved, and proceeds with operation.

An occasional delay in surgery may be due to the frequency with which multiple congenital anomalies are found in infants with intestinal obstruction; there is a tendency to wait for another lesion incompatible with life to make itself known. Obviously, anticipating the unknown rather than treating the obvious, can only result in the death of many infants who might have been saved. The not too infrequent association of neonatal intestinal obstruction with mongolism is another deterrent to surgical effort. Here again I believe that sufficient error is made in the diagnosis of mental deficiency in the newborn to warrant proceeding with necessary surgery. Mongolian idiots, as a rule, are not good operative risks, as compared with other children; much of the decision is thereby taken out of the surgeon's hands. The surgeon who has seen the successful management of multiple defects leave a child as normal as he might have been without them is naturally less timid in attempting to correct intestinal obstruction in the infant with other anomalies.

Fortunately, the decision concerning the propriety of surgery in these circumstances is only infrequently necessary. Our surgical staff has shared in the heartaches of many parents whose children have died, but only rarely has a child survived whose death after neonatal surgery would, in retrospect, have been a blessing.

### EFFECTS OF INTESTINAL OBSTRUCTION

Because infants lack the reserve found in adults, many of the physiologic disturbances which result from mechanical intestinal obstruction become serious earlier in infants. Not only does the untreated obstruction result in distention of the bowel and delayed return of motor function, thereby preventing the normal nutritional processes from being carried on, but far-reaching and progressive changes in the respiratory and circulatory systems also take place.

With the increased distention and the elevation of the diaphragm, which then becomes limited in its excursion, the thoracic cavity available for breathing space is diminished and dyspnea ensues; the resulting fatigue may lead to exhaustion in a newborn infant. Such a situation also encourages atelectasis and hypoxia. One of the most disturbing problems in infant surgery is that of hypoxia under anesthesia. Surgeons and anesthesiologists alike tend to evaluate their results by the survival of their patients. In my opinion, cerebral damage, which cannot be measured because of the nonexistence of a base line, must inevitably result from prolonged hypoxia under anesthesia. This in turn alters respiratory physiology.

Intestinal obstruction also affects the circulatory system, not only by the loss of plasma as a result of the obstruction, but also by causing hypotension with a resultant decrease in pulse pressure and a less efficient blood flow through the tissues. This further complicates the problem of hypoxia.

The combination of respiratory and circulatory difficulties caused by the obstruction, coupled with the loss of fluid by vomiting and by accumulation in the intestine, is undoubtedly responsible for the high mortality in this group of patients, despite the excellence of surgical technics and the increased knowledge of postoperative fluid and electrolyte problems.

### SYMPTOMATOLOGY

Vomiting, absence of stool, and distention are the classic signs of advanced intestinal obstruction in any age group. In the newborn, however, it is frequently possible to make the diagnosis early. Waiting for the appearance of the symptom triad results in a loss of valuable time, and the delay in diagnosis is reflected in a high mortality. If it is possible to make the diagnosis of intestinal obstruction early, why are so many infants treated expectantly for 4 or 5 days?

The answer to this question is not a simple one, but certainly should include the fact that the signs which lead to a diagnosis of intestinal obstruction are also physiologic signs in many newborn infants. Inaccuracies in charting by nursery attendants, especially in reference to the passage of meconium and regurgitation, contribute to the pediatrician's dilemma and to the delay in diagnosis.

**VOMITING.**—Vomiting may be physiologic, usually due to swallowed amniotic fluid and vernix or to air swallowed in crying. The latter is more common in those understaffed nurseries where feedings are not maintained on schedule. Such regurgitation during the first day of life may be relieved by aspirating the stomach of its contents and giving the baby a fresh start. Should regurgitation be encountered after aspiration of the stomach or for the first time after the first day of life, a roentgenogram of the abdomen should be made without delay.

Since vomiting is the cardinal symptom of intestinal obstruction in the neonatal period, the type of vomiting and character of the vomitus provide important information. Esophageal vomiting is really due to an impediment to swallowing and high esophageal obstruction of the complete variety produces a type of regurgitation which cannot be confused readily with any other. When such vomiting is spontaneous, the vomitus is a sticky material, composed largely of mucus and saliva.

Vomiting produced by obstruction at the pylorus or some point in the duodenum is forceful in character, revealing the action of a hypertrophic gastric musculature. The vomitus is colorless if the obstruction is above the ampulla, but otherwise contains bile. There is much mucus in this vomitus and more often than not, in our experience, particularly in neglected cases, there is changed blood, producing a coffee-grounds appearance.

Vomiting caused by obstructions of the small or large bowel may be quite varied. Even distal obstructions of the colon may have a vomiting pattern not very different from that seen with pyloric or duodenal obstruction. Its onset is usually somewhat later than in the other varieties of vomiting, it is accompanied by distention, and the vomitus may contain meconium. Vomitus without bile may be an early sign of intestinal obstruction, and the suspicion of such obstruction need not wait for the appearance of bile in the vomitus.

It should be remembered that vomiting can also result from lesions other than those of the gastrointestinal tract. Notable among

these are central nervous system lesions and obstructive lesions of the urinary tract; occasionally, vomiting may be associated with cardiorespiratory difficulties.

**VARIATIONS IN PASSAGE OF MECONIUM.**—The physiologic passage of meconium is a variable phenomenon and therefore readily lends itself to confusion with pathologic states associated with intestinal obstruction. In maintaining an aggressive suspicion concerning intestinal obstruction, the meconium must be evaluated both qualitatively and quantitatively.

The infant who passes wet meconium shortly after birth in large quantities is almost never the victim of an intrinsic obstructive lesion.

Rarely, a baby with an extrinsic lesion causing complete or nearly complete obstruction will pass a normal amount of meconium during the first several hours of life and then stop.

The complete absence of meconium is, of course, definitely pathologic, but a diminished amount may also be so. Even in complete intrinsic obstruction of the small or large bowel the diaper may be stained or something that looks like meconium may be passed. It seems needless to suggest that in the presence of distention or vomiting, some inquiry be made into the volume of meconium, which on the patient's chart may be indicated by a check mark. Occasionally, an infant is seen whose referring physician explains the delay in diagnosis to be the attendant's report that meconium stools were passed when actually these were mere stains.

When meconium does not pass early, a rectal examination or perhaps an irrigation is in order. If the presence of an imperforate anus has been ruled out at the time of delivery, a digital examination may reveal a rectal valve, or, more rarely, an atresia of the colon just above the sphincter.

In the newborn, the caliber of the colon is quite small and the rectal ampulla is usually somewhat larger. In an infant with obstruction higher than the descending colon, in whom the meconium has never passed through the anus, the caliber of that portion of the bowel is often even smaller than normal. A finger inserted in such a rectum can seldom get beyond the ampulla and almost never beyond the peritoneal shelf. The sensation is that of reaching a blind end of bowel; this physiologic state may lead the examiner to believe that the site of obstruction is at a low level, which is quite unusual.

If the meconium is inspissated, a digital stimulus may initiate its passage. If no meconium is encountered, irrigation of the lower colon

with saline by means of a fine rubber catheter attached to a syringe may be rewarding. Indeed, a meconium cast of the lower colon may be evacuated, followed by normal meconium stools. If no meconium is produced by either stimulus, roentgen examination is mandatory. Should a small bit of apparent meconium or intestinal contents be obtained, it is then available for chemical and microscopic study.

The small meconium stool which may or may not indicate an obstruction can be studied microscopically, as described by Farber (8), for epithelial cells swallowed *in utero*. The gelatin film test for the presence of trypsin is also helpful on some occasions, if one can afford the time necessary for incubation of the specimen (39). The absence of cells indicates an atresia of the bowel, but their presence does not rule out an incomplete intrinsic lesion or an extrinsic obstruction.

**DISTENTION.**—Distention in the newborn infant may be physiologic until the meconium plug is passed. However, one should not wait too long for the appearance of meconium in a distended infant before thinking of obstruction. When distention is present on the second day of life, or even on the first day following the passage of meconium, a roentgenogram of the abdomen should be made without delay.

Further evaluation of the infant is necessary, particularly with reference to a decision concerning an immediate operation as opposed to a matter of several hours' preparation. But once the suspicion of obstruction has led to a roentgen examination of the abdomen and the presence of obstruction has been established, there is little else to do.

#### ROENTGENOGRAPHY IN DIAGNOSIS OF INTESTINAL OBSTRUCTION

There is little doubt that plain or flat films of the abdomen in the supine and erect positions are the most valuable diagnostic aids when an obstructive lesion of the bowel is suspected. They are quickly obtained, atraumatic, and the examination is easy to perform. The contrast produced by the gas contained in the bowel is as informative in most cases as that produced by a medium administered for contrast, and the risk of aspiration of some radiopaque material which may be vomited is obviated, as well as the hazard of increasing the obstruction with a substance such as barium.

A knowledge of the normal gas pattern is important for a correct evaluation of the pathologic findings. Many observers (7, 38) have

recorded the appearance of gas in the small bowel in the first 5 hours of life and the passage of flatus by rectum as early as the sixth to eighth hour. Most infants show a predominance of small-bowel gas patterns in the first 12 hours of life, with the colon showing some distention thereafter until the bulk of the meconium has been passed. Afterward, the small-bowel gas patterns are predominant and not well delineated.

The best information is obtained from an upright film. The absence of gas in the abdomen indicates an atresia of the esophagus without fistula communicating with the tracheobronchial tree. When the latter is present, the abdominal film does not vary from the normal.

In a diagnosis of obstruction of the duodenum and upper jejunum, the flat film is at its best. There is distention above the point of obstruction and an absence of gas below it. A similar picture may be obtained with midgut volvulus, in association with malrotation of the colon, or a few bubbles beyond the point of maximal obstruction may be seen.

The diagnosis of obstruction is quite obvious on the flat films of lesions of the more distal part of the bowel, but the exact level of the obstruction is obscure. In an adult, it is usually easy to tell the difference between small and large bowel shadows, but in infants the plica circulares of the small bowel and the haustrations of the large bowel are not as well developed as in adults, and the differentiation is always difficult and sometimes impossible. A common error is to assume that the obstruction is lower than it actually is because of the tremendous distention and apparently long segments of dilated bowel.

Storch *et al.* (40) have pointed out two progressive signs that indicate small bowel obstruction in infants: "Squaring off" where the several gas-filled loops have roughly square lines of demarcation, and "continuity" where the squaring off is disappearing and the distended loops are long, broken only by the appearance of septums incompletely interrupting the gas-filled lumen. Fluid levels in normal loops of small bowel in infants do not necessarily indicate intestinal obstruction, but the presence of fluid levels in dilated loops is diagnostic.

In addition to the gas pattern, the shadows of tumor masses, meconium, or calcifications can be sought on roentgenograms. Meconi-

um with the appearance characteristic of adult fecal shadows is strongly suggestive of meconium ileus, whereas calcification outside the bowel or attached to the bowel wall is diagnostic of meconium peritonitis. However, meconium ileus due to pancreatic achylia may exist without this roentgenographic sign.

All these signs can be seen on the flat film alone. Occasionally, a barium enema helps in diagnosing megacolon or in ascertaining the position of the cecum in suspected malrotation of the colon. In general, however, unless such a study will be of extraordinary help to the surgeon, the better plan is to operate with the diagnosis of intestinal obstruction, exact cause unknown. Particularly in hospitals where congenital bowel lesions are uncommon, there is a tendency to accumulate a series of films for teaching purposes. Since infants are easily exhausted, and particularly so if they have abdominal distention with the accompanying strain on cardiorespiratory reserve, the quickest, simplest method of getting information is preferable.

The small patient with partial obstruction or an intermittently obstructing lesion is frequently given an opaque meal. In itself, this may be indicated under special circumstances; but the infant is fluoroscoped repeatedly and kept in the department of radiology for hours at a time while awaiting the completion of a study. Such an examination can be more difficult for the patient than an operative procedure, because in the latter adequate supportive measures are taken while in the former these are completely neglected.

Repeat films have a special place in the problems of imperforate anus, or when a meconium obstruction is suspected and a meconium stool is then passed.

## TREATMENT: GENERAL ASPECTS

### ANESTHESIA

Anesthesia in neonatal surgery is a highly specialized art, but one must use the agents and technics which prove best in a particular institution with its own personnel. Anesthesia is more than the administration of an agent, however, and it is the physiologic approach to the problem of infant anesthesia that has been largely responsible for the success of pediatric surgery.\* Preanesthetic medication is

\* The author gratefully acknowledges the personal skill of Dr. Margery Deming, and her concepts of neonatal anesthesia which are embodied here.

omitted in the infant weighing less than 5 lb. In patients above that weight, 1/600 gr. of scopolamine is used. Morphine is not given to infants not yet fed by mouth because it may easily depress to a critical degree their low internal heat production and their basal metabolism. When infants weigh over 7 lb., and have been fed, 1/480 gr. morphine is given, primarily to control tachypnea. The effect of the morphine is to decrease the respiratory rate and increase the tidal exchange. Efficient respiratory rate in a full term infant under anesthesia is 60 per minute or less.

Preinduction care should include aspiration of the stomach, since regurgitation may occur without evidence of vomiting. Even though a Levin tube or catheter has been used for suction drainage, aspiration should be carried out because the tube may be blocked and the stomach full.

We prefer endotracheal technics and we use intubation for a number of reasons. Adequate assistance to respiration can be given, pulmonary aspiration may be prevented, a patent airway is maintained at all times, and laryngeal spasm due to irritation by the anesthetic and secretions is avoided. The assistance to respiration is particularly helpful because the tidal exchange is decreased and may be inadequate when the abdomen is distended. Closure of the abdomen is also facilitated when respirations are controlled.

Cyclopropane, oxygen, and ether is the combination of agents most frequently used, the last mentioned to prevent the bradycardia which so often accompanies the use of cyclopropane. Curare preparations and muscle relaxants are hazardous, for they cause a fall in blood pressure, delay return of consciousness, and really do not help as much as one would expect. Muscle tone is poor in the newborn and a distended bowel makes wound closure difficult. Succinylcholine is probably the safest of relaxants because of its short action. The dosage, however, is quite variable in the newborn.

Following anesthesia one should make certain that the stomach is empty. The infant should be placed in an oxygen-enriched atmosphere if distended or if the respiratory rate remains rapid.

#### OPERATIVE AND POSTOPERATIVE CARE

Preparation for all possible exigencies should precede any operative procedure. In addition to competently administered anesthesia, an in-dwelling catheter of suitable plastic material or a metal cannula

must be in a vein and fastened securely enough to permit rapid infusion of blood, if needed. In our experience, a wide transverse incision facilitates exposure and wound healing and the cosmetic appearance of scars is superior to that obtained with longitudinal incisions.

Among the standard postoperative measures are: (1) gastric suction drainage; (2) use of chemotherapy, antibiotics, and vitamins; (3) repeated small infusions of blood or plasma; and (4) maintenance of fluid and electrolyte balances. Perhaps most important of all is that infants who have undergone surgery are critically ill and must be seen at repeated short intervals and their management changed as indicated by the postoperative course.

In general, feeding by mouth is attempted as soon as possible. Usually, 5 days must pass before an anastomosis, such as a duodeno-jejunosomy or enteroenterostomy, permits the passage of sufficient fluid and air to prove that it is functioning. This is the very practical reason why search should be made for multiple obstructive lesions at operation; in the absence of knowledge of their existence the prolonged postoperative block to the passage of fluids and gas may lead to an unnecessary exploration at just the time when the patient is least able to withstand it.

Although planning fluid therapy according to well-accepted standards, it is well to lean heavily upon chemical determinations of  $\text{CO}_2$ -combining power, and determinations of serum chloride, serum sodium, and serum potassium.

In-dwelling tubes passing through anastomoses have been used to feed infants postoperatively (18). It is safe to avoid such a practice, if possible, although very fine-caliber plastic tubes (rather than rubber or neoprene catheters of larger caliber) across anastomoses may prove to be quite safe. Our experience with the use of such tubes following end-to-end anastomosis of the esophagus seem to indicate this (23). Only rarely does an infant tolerate feedings placed directly into his jejunum; tube feeding into the jejunum, although mechanically possible, may therefore prove to be physiologically unsound. The use of an incubator, such as the "isolette,"\* seems to have improved the postoperative course of many infants undergoing surgery, although a number of other, seemingly minor, technical advances carried out coincidentally may be at least equally responsible for the lowered morbidity and mortality rates.

\* Air-Shields, Inc., Hatboro, Pa.

## OBSTRUCTIONS BY SITES AND TYPES

## ESOPHAGUS

**ATRESIA.**—Most infants with atresia of the esophagus are born cyanotic, with excessive amounts of mucus in the nasopharynx. Following aspiration and oxygen administration they become pink, and until accumulation of secretions again produces cyanosis are free of pathologic signs for some minutes to hours. An astute observer may suspect esophageal atresia at this time, but in any case the diagnosis should be obvious at the time of the first feeding. The second swallow on its way down meets the forcefully ejected first swallow on its way back in the nasopharynx; the infant chokes, coughs, and sneezes, simultaneously with the appearance of the feeding at the nostrils. Aspiration usually occurs at this time, and aspiration pneumonia, frequently seen first on the right and most commonly in the upper lobe, has its beginning.

Intranasal passage into the esophagus of a no. 8 soft rubber catheter, which will stop at the site of the atresia, usually high in the mediastinum, easily confirms the suspected diagnosis. Further confirmation is not necessary, but can be obtained by injection of several cubic centimeters of iodized oil into the catheter. Barium, so dangerous when aspirated, should not be used.

Atresia of the esophagus is usually accompanied by a tracheoesophageal fistula, and any combination of atresia with or without proximal or distal fistulas is possible. About 80 per cent of patients with this problem have a proximal atresia of the esophagus with a distal tracheoesophageal fistula. The symptoms are due to the atresia and not to the fistula. The latter may complicate the situation by permitting vomiting of stomach contents into the tracheobronchial tree via the fistula. The air which enters the gastrointestinal tract through the fistula, when seen roentgenographically, assures the surgeon that a distal esophagus is present which in other cases may be nonexistent (Plates 1 and 2). In our experience, this lesion is second in frequency only to imperforate anus as a cause of alimentary obstruction in infants.

A number of surgeons have, in the past decade, achieved remarkable results in the operative treatment of this lesion, which before 1943 was invariably fatal. Their procedure consists of a retroperitoneal approach to the defect, with closure of the fistula and end-to-end

anastomosis of the esophagus (15, 42). Even after 10 years, success does not uniformly reward the effort that many have contributed to the correction of this anomaly.

We prefer a transpleural approach to the lesion, a simple anastomosis instead of a two-layer one, and postoperative feedings by mouth or fine polyethylene tubing instead of the usual gastrostomy (21). These departures from the more common retropleural technics with gastrostomy have cut operating and anesthesia time by 60 per cent; have shortened the hospital stay (an economy to the family and hospital), and have virtually eliminated the need for postoperative dilatation of an esophageal stricture at the site of anastomosis, which is frequently encountered when feeding by mouth is delayed. With a team used to managing neonatal problems, a survival rate of about 80 per cent can be expected, but at this writing there are few clinics able to achieve this consistently. Such factors as multiple lesions, delayed diagnoses, and other defects incompatible with life will probably prevent the attainment of much better figures in any long series of cases.

**STENOSIS.**—This is far less frequent than atresia and is not as difficult to manage because the obstruction is incomplete.

**ACHALASIA.**—This condition is amenable to medical management and alterations in position during feeding.

**CONGENITAL SHORT ESOPHAGUS.**—The obstruction produced is actually of the cardiac portion of the stomach as it passes through the diaphragm. In our experience, this is a very rare lesion, representing the most unusual variety of obstruction above the pylorus. Large numbers, however, have been reported from England (48).

**DUPLICATION OF ESOPHAGUS.**—The presenting symptom in these cases, which are seen occasionally, is inability to swallow during the neonatal period. The duplications are usually large (Plate 3) and, as such, displace pulmonary tissue so that respiratory distress or cyanosis may lead to a roentgenographic examination of the chest before the inability to swallow is actually known. The common wall between the esophagus and duplication usually dictates a procedure such as marsupialization, although we have more often than not been able to dispose of the duplication by a one-stage extirpation.

#### STOMACH

Only a few lesions produce obstruction at the level of the stomach in children, and fewer still cause difficulty in the neonatal period.

**DUPLICATION.**—A duplication of the stomach must assume tremendous size or be located at the cardia or pylorus to cause obstruction (14), so that obstruction of the gastrointestinal tract due to duplication of the stomach is more of a possibility than a probability in the neonatal period.

**CONGENITAL HYPERTROPHIC PYLORIC STENOSIS.**—This would seem to be a congenital rarity. Many have been impressed with the marked hypertrophy of the pylorus at a very early age, and the typical pyloric tumor has been seen as early as the seventh month of fetal life. Of interest in this regard is the work of Wallgren (45), who made a roentgenographic study of 1,000 newborns by means of a contrast meal. He found that the roentgenographic features of 5 infants in whom lesions of hypertrophic pyloric stenosis subsequently developed were indistinguishable from those of 995 infants in whom stenosis did not develop.

There are, occasionally, infants who vomit at birth and slowly, but early, develop the typical vomiting pattern of pyloric stenosis and, indeed, have the lesion. Just when these patients pass into the clinical stage of true pyloric stenosis is not known but the tumor has been found at operation as early as the fourth day of life. Most pyloric tumors are palpable, and if positively felt, no roentgenography is necessary. In the neonatal period, however, the diagnosis is unusual enough, so that the possibility is not considered, as a rule, and roentgen examinations are commonly done without any effort to palpate the pylorus. Certainly, the presence of continued vomiting without distention should lead one to consider the diagnosis and confirm it by means most reliable in his hands.

There is no question that a Fredet-Ramstedt type of pyloroplasty is the procedure of choice, through the smallest incision the surgeon can safely use. Some prefer medical management with antispasmodics and atropine-like drugs. In view of the safety of surgery for pyloric stenosis, as well as the certainty of results, nonsurgical management of this condition at the present stage of knowledge, in my opinion fails to make the best use of available treatment.

#### DIAPHRAGM

Defects in the diaphragm may cause obstruction of the gastrointestinal tract at the level of the esophagus, stomach, small intestine, or splenic flexure of the colon. Not infrequently, diaphragmatic hernia on the left is associated with malrotation of the colon, so that either

a volvulus of the small bowel or bands obstructing the duodenum may be the cause of obstructive symptoms rather than the compression of bowel by the edges of the diaphragmatic defect itself. Or, the two phenomena may occur concomitantly.

In our experience, when diaphragmatic hernia is diagnosed in the first few hours or days of life, the signs and symptoms have usually been those of cardiorespiratory embarrassment. Signs of obstruction in the absence of respiratory distress lead to a diagnosis of a diaphragmatic lesion later in the neonatal period. The cardiorespiratory signs are usually a function of the volume of abdominal viscera displaced into the thorax which, in turn, is somewhat a measure of the size of the hernia.

Assuming that the diagnosis of diaphragmatic hernia has not been made in the first few hours of life on the basis of cardiorespiratory signs, obstructive phenomena may make their appearance when gas fills the intestine, the viscera expand, and the loops of large and small bowel are forced into the pleural cavity. Distention, depending on the level of obstruction, usually is of secondary importance to vomiting as a sign which leads to roentgenography of the abdomen, and in turn to a diagnosis.

Fortunately, most abdominal roentgenograms of infants reveal enough of the chest to give a clear view of the diaphragm (Plate 4). The clarification of diagnostic problems concerned with diaphragmatic hernia is not always as easy by roentgen examination as might be surmised. Eventration of the diaphragm, which is rarely, if ever, a cause of obstruction, may be easily accompanied by vomiting, and can be confused with a true diaphragmatic hernia or a diaphragmatic defect permitting the passage of intestine into the thorax.

Occasionally, supposed roentgenographic evidence of gas-filled small bowel in the thorax has proved on operation to have been the shadows of a multiloculated empyema.

Small diaphragmatic defects, as of the foramen of Morgagni, can be the site of incarceration of a loop of bowel which is difficult to prove roentgenographically, particularly if the situation is grave and the use of a radiopaque medium is precluded. Some diaphragmatic defects are asymptomatic throughout life and without neonatal signs. But in those which do produce symptoms in the neonatal period and remain uncorrected, the mortality rises to about 90 per cent by the end of the third month.

Once the suspicion of a diaphragmatic hernia is confirmed by

x-ray, operative correction should be carried out forthwith. Because some diaphragmatic hernias are known to be asymptomatic, there is a tendency to postpone surgery until the infant is older. Procrastination usually results in death, or the necessity of an emergency operation with even greater risk than at the time of first diagnosis.

The aim of all operative procedures is to reposition the abdominal viscera in the peritoneal cavity and to close the diaphragmatic defect. This may be accomplished by the transabdominal or transthoracic routes. Judging by the literature, the former seems to be preferred; but we routinely use the latter, feeling that it presents advantages over the abdominal procedure. The mortality rate, in our experience, is 13 per cent (24). The thoracic approach permits rapid emptying of abdominal organs from the chest in the distressed infant, and the exposure is superior to that achieved with an abdominal procedure. We encounter no greater difficulty in putting the viscera in their proper place through the aperture in the diaphragm than through the abdominal incision. Neither method is easy. In 2 patients with congenital absence of the diaphragm it has been possible, using the transthoracic route to construct a diaphragm out of the abdominal parietes; this probably could not have been done from below. We believe, furthermore, that the abdominal wall adjusts to the overcrowding of viscera better if there has been no abdominal incision.

#### VOLVULUS OF MIDGUT

Fundamental to the understanding of the etiology in midgut volvulus is the knowledge of the embryologic rotation of the colon and its variations. At the risk of oversimplifying a complex series of events it can be said that failure of the cecum to descend into a fixed position in the right lower quadrant can permit the ileocecal valve to lie in close proximity to the duodenojejunal junction with a relatively long, and at times poorly attached, mesentery. Or, failure of attachment of the ascending colon to the abdominal parietes may permit the proximal colon, which is then freely moveable on its blood supply, to behave as though it were an additional length of small intestine.

With two fixed points, the duodenojejunal junction and ileocecal valve or duodenojejunal junction and hepatic flexure lying side by side instead of being widely separated, as in the normal anatomic position, the small bowel is free to twist on itself in either direction and to pull the ascending part of the colon with it when the anatomic situation permits.

Several hundred cases of this lesion have been reported in the literature. About half of them presented symptoms the first day or two of life, and most of the remainder in the first two weeks. Probably, many have a compromised blood supply even *in utero*. Roughly, a quarter of the volvulized loops were of the whole small bowel, while the remainder included the right colon (6, 12, 27).

Persistent bilious vomiting is the cardinal sign of volvulus of the midgut. The infants do not seem to be in pain but seem hard hit by the situation. Distention is not a problem, especially at first. Blood is sometimes seen in the stools but usually after the passage of normal meconium. An abdominal mass is sometimes palpable; in our experience this is due to collapsed bowel and mesentery twisted upon itself.

Other lesions of embryologic variation are not uncommonly associated with malrotation of the colon. Bands crossing the duodenum from the malrotated colon to some other fixed point are perhaps the most usual, but other adventitious (probably not inflammatory) bands, internal hernias, volvulus of the duodenum, and situs inversus, either partial or complete, are also found. Roentgenographic examination of the abdomen confirms the diagnosis of high intestinal obstruction and may lead to an accurate preoperative diagnosis (Plate 5).

The diagnosis of high intestinal obstruction, even without exact knowledge of its etiology, makes surgical exploration mandatory. At operation, the surgical procedure will be indicated by the exact nature of the lesion found, but the operation will not be complete until the malrotation is corrected by the Ladd procedure. In this operation the malrotated colon is detached from its abnormal fixation, leaving it free to fall into the lower abdomen where a greater distance will be maintained between the ileocecal valve and the duodenojejunal junction.

#### INTUSSUSCEPTION

This lesion is very rare in the newborn. Intussusception in the very young is usually of the idiopathic variety rather than due to demonstrable mechanical factors, and usually is ileocolic. It may be ileocecal and be externally visible through a patent omphalomesenteric duct (Plate 6).

Intussusception has been reported in a 7 month premature infant only a few days old. Jeffrey (20) reported on 17 cases in the neonatal period collected from the literature, with only 1 survival. His

own case, which he believed to be the youngest at that time, was 28 hours old. Few neonatal cases have been reported since. In one of our cases, the infant passed the typical stool of blood and mucus immediately after the passage of the meconium plug in the delivery room. Since it normally takes several hours at least after intussusception begins for blood to be passed by rectum, this must have begun *in utero*. Vomiting began almost immediately, and a mass was palpable before laparotomy was performed a few hours later. The infant gave no evidence of discomfort. Recovery was uneventful.

The use of a barium enema to relieve intussusception is permissible only if the surgeon is on hand to take over the management of the patient if barium reduction is unsuccessful and if certain criteria of safety regarding height of the column of barium and abdominal palpation are rigidly enforced. The mortality in our cases of intussusception compares favorably with that of published series in which barium was tried first. The author's preference is for operative treatment, which prevents delay, picks up organic lesions when present, and makes reduction absolutely certain. In the newborn particularly, in whom the diagnosis may not be clear-cut, and in whom delay may be tragic, exploration of the abdomen should probably be carried out without recourse to preliminary barium enema. Abnormalities in blood coagulation should also be searched for in newborn infants who bleed in quantity from the rectum. No bowel resection for neonatal intussusception has been necessary in our clinic, but, if it were, direct anastomosis would be preferred unless the patient's condition permitted no more than the Mikulicz type of external anastomosis.

#### ABNORMAL MECONIUM

Intestinal obstruction in the newborn due to meconium is an example of a disturbed physiology resulting in an organic obstruction. Pathologic obstruction resulting from abnormal meconium must be differentiated from the delayed passage of a physiologic meconium plug. This plug may be spontaneously evacuated after some intestinal distention has been built up behind it, or it may require the stimulation of a digital examination of the rectum or, occasionally, the irrigation of the lower colon with saline. Subsequent studies in patients who pass dry meconium plugs have not, in our experience, contributed any positive knowledge concerning the etiology of this condition (Plate 7, A).

The pathologic group may be divided into four varieties: (1) obstruction caused by chemically abnormal meconium due to pancreatic achylia; (2) altered meconium in conjunction with an apparently associated anatomic defect, such as atresia of the bowel; (3) atypical meconium and a concomitant lesion, such as volvulus of the small bowel; and (4) complications of the meconium obstruction, such as perforation of the bowel with abscess formation or a plastic variety of peritonitis.

The clinical picture varies in accordance with the site of the obstruction; it also depends on associated findings. These infants do not, as a rule, pass meconium by rectum, although occasionally there are reports of diapers being stained green-black. In addition to the vomiting, distention, and absence of stools, the careful observer may note excessively tenacious nasopharyngeal mucus. The roentgenogram is usually diagnostic, presenting "fecal shadows" and other phenomena of intestinal obstruction as demonstrated by gas patterns and fluid levels (Plate 7, B). The characteristic calcification of meconium, in the bowel or extruded into the peritoneal cavity, is diagnostic of atypical meconium. The history of a similar condition in a sibling is helpful, for the familial tendency in pancreatic fibrosis and meconium ileus seems to be well established.

Almost all agree that the absence of pancreatic secretions in the intestines is etiologic in meconium obstruction of the bowel. Attention was first called to this association by Landsteiner in 1905. The pancreatic enzymes may be prevented from reaching the bowel either as a result of pancreatic fibrosis or of atresia of the pancreatic ducts, as Farber described (9, 10). Hinden (17) has reported meconium ileus without pancreatic fibrosis. His patient, however, had a volvulus associated with it at birth and later died of intestinal obstruction, so that there may be some question concerning the inspissated meconium.

The patient with pancreatic insufficiency may have meconium of green-black color, thick, sticky, and mucilaginous, or it may resemble dried putty and leave no stain on one's examining fingers. Attention has been called to the chemical differences between normal meconium and that found with pancreatic achylia, but, unfortunately, seldom is there enough of a specimen in the obstructed infant to aid the preoperative diagnosis. The gelatin film test for trypsin is simple enough to help the surgeon occasionally before his hand is forced by the patient's condition (39).

Meconium obtained at operation may be studied and is of some prognostic significance. Buchanan and Rapoport (5, 33) have noted that normal meconium contains larger amounts of total reducing sugar, as compared with that of patients with pancreatic achylia. Variations were also reported in the nitrogen partition and in pepsin and trypsin activity.

Why only a small percentage of patients with pancreatic fibrosis present the syndrome of meconium ileus is not known. Hurwitt and Arnheim (19) have suggested that the pancreatic lesion is one of early intrauterine life and that the time of its beginning might be roughly estimated on the basis of the distance the meconium has traveled. (Meconium is in the cecum by the fourth month and is in the rectum by the fifth month of fetal life.)

Statements in pediatric literature that meconium ileus is invariably fatal, coupled with the ever-present hope that an obstruction due to meconium will relieve itself, lead not only to delay in the treatment of meconium ileus but encourage resort to ineffective measures in an effort to avoid a laparotomy which might prove unnecessary.

It is safe to say that the laparotomy, unnecessary in retrospect, seldom has an adverse effect on the patient, but that early laparotomy in the case of meconium ileus may result in a successful outcome that would be precluded by delay.

Until quite recently, meconium ileus was uniformly fatal. Since the report of Hiatt and Wilson (16) in 1948, a number of surgeons have successfully treated the obstructive manifestations of this entity in the neonatal period. After the successful management of the obstruction, the infant still has the future of a child with pancreatic fibrosis but we do not believe that this in itself is sufficient reason to withhold surgery from the infant.

At operation, the typical lesion of meconium ileus consists of a segment of bowel tightly packed with meconium of the inspissated variety, the bowel distal to it collapsed, and the proximal intestine dilated with gas and liquid contents. The wall of the bowel containing the meconium is thickened, showing the effects of peristaltic effort to move the meconium distally. Several surgical procedures are possible. The one most commonly used is that of opening the bowel, removing the meconium by mechanical means or irrigation, and then closing the bowel after patency of the distal bowel has been established. Some surgeons use pancreatin suspensions for the irrigation and leave some behind in the bowel before suturing.

Another technic is to resect the area of inspissation and join by anastomosis the proximal dilated and the distal collapsed segments. This technic is limited by the length of the loop containing the firm meconium, and adds to the problems of management of the patient those connected with an anastomosis. If the obstructing meconium is low enough in the bowel, an enterostomy can be performed just proximal to that area, and postoperatively the distal loop irrigated until clear. This technic is especially valuable in the infant who is doing poorly on the operating table and when a short procedure is desired. The limiting factor, of course, is water and electrolyte loss through the enterostomy. We have used all these technics with success, and feel that each has its place. In the majority of cases, if the first method is feasible, it perhaps provides the greatest safety.

If, at laparotomy, the atypical meconium is associated with other lesions, then both, of course, must be treated. The volvulus which is occasionally seen with meconium ileus is apparently caused by the effort of hyperactive peristalsis to dislodge the meconium. The treatment of the meconium obstruction removes the cause of the volvulus which should not recur. Obviously, if the blood supply is compromised, the gangrenous lesion will dictate therapy (Plate 7, *C* and *D*).

Occasionally, an atresia of the bowel is associated with an apparent meconium ileus or meconium peritonitis. The two phenomena can occur concomitantly, but some of the apparent atresias are associated with evidence of peritonitis so that two possibilities may be considered: (1) perforation proximal to an organic obstruction, or (2) apparent organic obstruction as a sequel to obstructive healing after perforation.

Meconium peritonitis (Plate 8) seriously complicates the various problems associated with abnormal meconium. It need not be associated with pancreatic achylia, although it frequently is. Meconium does not appear in the bowel until the end of the third month of gestation. Advanced plastic peritonitis has been seen as early as the sixth month of fetal life (31).

Meconium is considered to be sterile for several days after birth and most meconium peritonitis is assumed to be sterile. It should be remembered, however, that bacteria from the infant's environment can enter the peritoneal cavity via the gastrointestinal tract and cause its perforation in a matter of hours.

Neuhauser (19), in 1944, described the roentgenographic appearance of meconium peritonitis, noting the areas of calcific density

within the peritoneal cavity—within the bowel lumen, within the bowel wall, or lying loosely adherent to the peritoneal surface of the bowel parietes. Neuhauser cited others as believing that the calcifications may begin as early as 24 hours after the onset of peritonitis. Brunkow *et al.* (4) followed the fate of such calcified areas and reported that in 1 case they had completely disappeared by 20 months. Our own experience confirms this.

Before 1950 there were no reports of survival in meconium peritonitis, and even now survivals are not common. If the bowel leak has sealed itself and there is no obstruction, drainage of the abscess cavity is atraumatic and presents the best opportunity for recovery. The surgical management of obstruction in the presence of plastic peritonitis presents as difficult a problem as the surgeon faces, and accounts for the high mortality.

Postoperatively, in addition to the surgical measures of decompression and support, these infants should be given pancreatin daily by mouth. We have also found that essence of caroid, acting as a mucolytic enzyme, is helpful.

#### UMBILICUS

The omphalomesenteric duct may persist as an open limb of the bowel draining meconium to the skin in the region of the umbilicus or as a band extending from the navel to the ileum. In the latter instance, there may be an associated Meckel's diverticulum on the ileum, a cyst within the all-but-obliterated duct, or a mucous sac lying just below the umbilicus and draining onto the skin.

Obstructions caused by the persistent remnant of the omphalomesenteric duct are unusual; when they occur, the mechanism is almost always that of small bowel volvulus around the fixed remnant extending from ileum to navel. Rarely, an intussusception of small bowel through the open fistula is responsible for the obstruction (Plate 6).

Definitive diagnosis is seldom possible; rather, one must be content with a diagnosis of small bowel obstruction, either complete or partial. Occasionally, the presence of drainage of either meconium or mucus at the umbilicus indicates an embryologic vestige and a presumptive diagnosis may be entertained. Exploration of the abdomen is mandatory, and the therapy is dictated by the situation which is found.

Omphaloceles do not produce obstructive symptoms, and the

incarceration of bowel in congenital umbilical hernias is rare indeed. It has been reported that an umbilical cord clamp has crushed small bowel containing an unrecognized omphalocele (1a).

#### ATRESIA OF SMALL BOWEL

Historically, it is interesting to note that single survivals of patients with atresia of the small bowel were recorded in 1911, 1926, and 1927. From then until 1950, there were only 22 others, during which period about 700 atresias of the small bowel were reported. More recently, the mortality rate has improved considerably in many clinics.

#### DUODENUM

Obstructions at the level of the duodenum are caused by a variety of lesions, present signs of high intestinal obstruction early, and require prompt diagnosis and skillful management if the infant is to survive. In general, the prognosis in these cases is better than in lower small intestinal lesions, probably due to earlier diagnosis because of earlier signs as well as to fewer physiologic changes caused by the obstruction.

The signs of duodenal obstruction are early vomiting and distention limited at first to the upper abdomen. Infants with obstructions lower than the duodenum may vomit on the first day of life also, so this sign is not pathognomonic, as sometimes stated. The vomitus may or may not contain bile and may have flecks of meconium in it. In neglected cases, changed blood is frequently seen.

A plain film of the abdomen is usually diagnostic, and the use of a contrast medium is unnecessary. The aspiration of gas from the stomach by catheter, followed by a second film, may delineate the duodenal lesion due to removal of the overlying gastric shadow (Plate 9).

**BANDS.**—These are the simplest obstructions, either adventitious or in association with malrotation of the colon. They cross the duodenum and cause partial or complete obstruction. Their surgical division is readily accomplished; malrotation, if present, should be corrected as noted previously.

**ANNULAR PANCREAS.**—A rare but formidable problem in duodenal obstruction is caused by an annular pancreas. Gross and Chisholm (13) reported a successfully treated case in 1944, and cited only 2 infant cases, with 1 survival, in previous literature. The diagnosis in 2 of our 4 cases was suggested preoperatively by Dr. John Hope on the basis of the "double bubble" of gas seen on the abdominal film

(Plate 10). In 3 of our cases there was an associated intrinsic obstruction of the duodenum; for this reason, in addition to the danger to the ducts, we believe a by-passing operation to be better than one which attempts division of the pancreatic ring alone. Duodenojejunostomy, if it is feasible, is probably the procedure of choice, but in most instances gastroenterostomy may be an easier and a safer procedure.

Except for the diagnostic "double bubble" in annular pancreas, great roentgenographic accuracy is not to be expected in separating one type of small bowel obstruction from another. One must be content with knowing whether the obstruction is in the duodenum or lower, and whether it is complete or partial.

**DIAPHRAGMS.**—When a diaphragm obstructs the lumen of the duodenum, it should not be perforated. The treatment of choice is resection and anastomosis or anastomosis around the lesion. Such lesions may be multiple and an effort should be made to rule out the presence of similar lesions in the lower bowel.

**ATRESIA.**—The management of obstruction depends on the lesion encountered. The level of the obstruction and the relationship of the atresia to the bile and pancreatic ducts will dictate the surgical procedure. Duodenojejunostomy or gastrojejunostomy produce equally satisfactory results initially. The long-term problems of gastrojejunostomy have not been proved in infants, but can be presumed, judging from experience in adults, to entail the additional hazard of marginal peptic ulceration.

#### JEJUNUM AND ILEUM

Generally, lesions obstructing the small intestine more distally are increasingly more difficult to manage. The lower the lesion, the later the onset of vomiting and the greater the distention. These are the patients referred to the surgeon after the longest delay. Postponement of diagnosis is reflected in some series, in which half the patients were considered to be beyond surgical treatment. Although this seems exaggerated, it indicates the poor condition of these infants when seen by the surgeon. Many infants with intrinsic bowel lesions are born prematurely, a factor which increases the risk. Multiple lesions also seem to be more common in association with atresia of the ileum than with that of the duodenum and colon, and the correction of multiple defects adds to the hazard.

The operative procedure in complete obstruction is anastomosis. This may vary in technic, and the choice of end-to-side or side-to-side

varieties must be dictated by previous experience and the anatomic situation. Despite traditional belief to the contrary, the delayed anastomosis of the Mikulicz type may be carried out in the lower quarter of the small bowel provided that the common spur of the two limbs of bowel is promptly crushed and the time during which the infant has an intestinal fistula is not prolonged. Regardless of method, there is no field of surgery where meticulous atraumatic technic and the use of fine suture material is as rewarding as it is in an anastomosis of the small intestine of a newborn infant (18).

**STENOSSES.**—Single or multiple stenoses may occur in any portion of the small intestine. Usually a single stenotic area produces signs of partial or intermittent obstruction; but with multiple lesions distention sometimes builds up rapidly and the resultant state must be considered as complete intestinal obstruction.

**DUPLICATIONS.**—When these produce obstructive symptoms in the neonatal period, the lesion is usually a cystlike structure with one wall in common with the mesenteric wall of the adjacent bowel (Plate 11). Resection of the lesion with an appropriate anastomosis has been uniformly successful in our experience.

**INTERNAL HERNIAS.**—Signs of high or low, complete or partial obstruction are produced by internal hernias, such as the paraduodenal variety, those behind the cecum, or those through abnormal mesenteric or omental apertures. Exact preoperative diagnoses are rare, and treatment is usually made mandatory by the obstructive phenomena. Symptoms may exist from birth and require relief in the neonatal period. More commonly, however, there is either a mild feeding problem or an asymptomatic interval before the situation demands correction in the first few months of life.

**INGUINAL HERNIAS.**—Particularly in premature infants, such hernias may produce signs of intestinal obstruction and indeed may incarcerate or strangulate a portion of the small bowel. The latter condition can usually be prevented by elective herniorrhaphy or by using a yarn truss until surgery can be carried out.

In the infant, an incarcerated hernia does not necessarily make emergency surgery mandatory. Most of these lesions can be reduced normally. It is noteworthy that an infant bowel suffers no damage from longer incarceration than is apparently the case in adults. However, if manual reduction cannot be accomplished early, surgical reduction should not be postponed.

The small bowel is subject to a great variety of obstructive lesions,

among them: polyps; benign and malignant tumors (30) (Plate 12); abscess (36); volvulus around fibrous bands connecting abdominal viscera with the parietes (28); adhesions, perhaps resulting from a prenatal bowel perforation with recovery; granulomas; and inflammatory lesions so old that they undoubtedly originated *in utero* (25). In these unusual situations, the preoperative diagnosis of intestinal obstruction is usually the only possible one. Laparotomy reveals the exact situation and treatment is then based upon the findings. Unfortunately, in most of these rare problems, the obstruction can be relieved only by extirpation of the lesion; this frequently introduces difficulties which may be insurmountable.

#### COLON

Except for imperforate anus and agenesis of the myenteric plexus (megacolon), the obstructive lesions of the colon in the neonatal period are less common than at any other anatomic site. The lesions themselves, however, resemble those occurring elsewhere in the gastrointestinal tract. Obstructions due to atresia, reduplication, and bands are infrequently reported.

Abdominal distention is most marked in distal bowel obstruction but that sign in itself should not be used in diagnosis and indeed is misleading in late cases of small bowel obstruction. We have seen mild bleeding by rectum with extrinsic obstruction of the splenic flexure. One obstructive reduplication of the rectum was visible externally over the buttocks.

Treatment is the same as for obstruction of the small bowel, with the exception of colostomy. A proximal enterostomy in the small bowel is dangerous because of fluid and electrolyte loss, whereas distal to the right transverse colon this aspect of a colostomy can be managed. In general, definitive surgical procedures are to be preferred if they do not entail too great a risk.

#### NEUROGENIC OBSTRUCTIONS

The relationship between certain types of intestinal obstruction and agenesis of ganglion cells in the myenteric plexus of the bowel has been made clear by Zuelzer and Wilson (47) and Swenson and Neuhauer (42-44). The pathologic physiology of neurogenic intestinal obstruction in childhood is therefore better understood now. There are a number of differences between neonatal obstructions due to disturbed

physiology and those due to organic causes. Generally, operation in the former discloses no obvious intrinsic or extrinsic lesion responsible for failure of bowel evacuation nor is there evidence of a work hypertrophy proximal to the neurogenic obstruction. These anatomic findings are reflected in the symptomatology and the roentgenograms. (1) Since a small meconium stool or two is frequently passed by an infant with a neurogenic obstruction and rapidly progressive signs of obstruction are normally absent, diagnosis is late. (2) The gas pattern on the roentgenogram indicates in many instances that obstruction is not complete; this further postpones the definite diagnosis and need for treatment. (3) Finally, the signs may vary in intensity from those of complete obstruction to almost normal meconium evacuation.

However, a fecal impaction in an infant is never normal. It must always be considered indicative of some disturbance, usually far-reaching, in the enzymic content of the intestinal fluid or in the nerve supply of the bowel.

Clinical separation of neurogenic small bowel obstructions from megacolon may be difficult. Usually the gas shadows and distribution of meconium shadows on the roentgenogram will yield a presumptive diagnosis. A barium or iodized oil enema can be used in the newborn to establish the diagnosis of aganglionic megacolon (Plate 13, A and B). However, although megacolon may be positively diagnosed, absence of the typical lesion in the rectum and descending colon does not rule out the involvement of a larger segment of the large bowel or of the colon and small bowel.

The appearance of the intestine at the junction of normal bowel with the aganglionic segment differs according to the anatomic site of the transition. This in turn is probably due to the consistency of the intestinal contents at a given level. For example, in high jejunal obstruction due to agenesis of the myenteric plexus there may be no demonstrable change in the contour of the bowel. In the usual lesion of Hirschsprung's disease, on the other hand, the aganglionic segment of colon is preceded by a markedly hypertrophied and dilated segment of bowel in which longitudinal muscle bundles particularly are seen to be hypertrophied.

The aganglionic segment of bowel is commonly termed spastic or in spasm. This is incorrect. Actually, the affected segment is deprived both of parasympathetic innervation and its local reflexes, resulting in its inability to undergo peristaltic action. This explanation is consistent with the generally accepted view that preganglionic parasymp-

pathetic fibers end around the neurons of the myenteric plexus and that intestinal reflex patterns are begun and controlled by the neurons in this plexus. The attempt by the proximal bowel to propel its contents through the aganglionic segment obviously accounts for the hypertrophy (Plate 13, C). The degree of hypertrophy seems to be related not only to the consistency of the bowel content at that particular level but also to the length of time the infant has survived.

#### MEGACOLON

In their first report on aganglionic megacolon in 26 patients, Swenson *et al.* (43) stated that in all but one case the onset was in the first 2 weeks of life. Hirschsprung had reported 2 such cases and Barrington, in a study of 19 autopsies, considered that 16 cases had been symptomatic from birth. In our series of more than 60 cases, symptoms were present from birth in a large number. The great majority of these have limped along with rectal irrigations and a variety of nostrums by mouth. Some in the age group of a few weeks to a few months have a history of an exploratory laparotomy shortly after birth, at which time the aganglionic megacolon was seen but not recognized. These infants received no surgical relief of obstruction or there was an attempt to milk meconium out of the anus through the aganglionic segment.

A small number of patients with neonatal obstruction due to megacolon have come to us a few hours to a few days old without previous treatment. In such cases, a colostomy proximal to the point of obstruction is performed immediately after establishing the diagnosis, and definitive surgery is deferred until a later date.

The diagnosis of megacolon in the neonatal period is suggested when the flat film of the abdomen shows fecal shadows in the colon of an infant who is passing no stools and is vomiting, or becoming distended, or both. Digital examination may show the rectum to be empty. An enema with an opaque medium reveals on x-ray a distal collapsed segment of dyskinetic bowel above which there is a hypertrophied and dilated segment.

Our clinical and roentgen findings in megacolon parallel those of Swenson and co-workers, and our histologic observations on the absence of ganglion cells in the myenteric plexus of the collapsed segment of bowel are in accord with those of Swenson *et al.*, and Bodian *et al.* (2, 3). The histologic basis of neurogenic obstruction is

apparently still ignored by a number of workers, who continue to use surgical procedures which leave an aganglionic segment of bowel in the patient. If there is a true megacolon, such procedures can only lead to a recurrence of the clinical picture of obstruction. At least half the infants with megacolon have an aganglionic segment extending distally below the pelvic floor; it therefore cannot be adequately removed by an anterior resection of the colon.

Fortunately, the emergency aspects of relieving obstruction in the newborn are paramount; few, if any, surgeons experienced in the treatment of aganglionic megacolon attempt definitive procedures in the first few days of life. In the less severely affected infants, who can be eased along out of the neonatal period without surgery, a choice of treatment may be possible at a later date.

Some believe, notably Potts (32), that because infants tolerate a colostomy so poorly a definitive procedure should be carried out as early as possible. All but 1 of our patients treated with colostomy in the neonatal period or in early infancy have thrived. The single fatality was not due to fluid loss. In our experience, even a transverse colostomy is not difficult to handle, so far as fluid balance is concerned. Of greater importance would seem to be the poor tolerance of all infants with megacolon to any gastrointestinal infection. A child with megacolon who has not had a colostomy or any other operative procedure is laid low by a bout of diarrhea out of proportion to the severity of the disease. Should an infant with megacolon and a colostomy have a bowel infection, the colostomy may perhaps exaggerate his problems, but the blame for fluid balance difficulties cannot fairly be placed on the colostomy itself. The infant with an obstruction due to megacolon demanding surgical relief would do far better with a transverse colostomy than with an immediate attempt at a definitive procedure, such as excising the aganglionic segment of colon.

#### SMALL BOWEL

When the aganglionic segment is not confined to the terminal colon, the symptoms closely resemble those of megacolon, but the problems of management are far greater. Indeed, when the aganglionic segment is proximal to the cecum, no treatment so far offers the hope of survival.

Forshall *et al.* (11) presented clinical material and an interpreta-

tion of it which is in line with the earlier report of Zuelzer and Wilson (47). Rickham's (37) experience with small bowel aganglionic obstruction closely resembles ours: these cases are difficult to separate from megacolon preoperatively; they do not respond to spinal anesthesia; there is no external clue to the site of aganglionosis; and lesions above the terminal ileum are uniformly fatal.

In treating an infant with neurogenic bowel obstruction, laparotomy is advisable, in order to ascertain the level of agenesis. If a typical case of Hirschsprung's disease is found, a proximal colostomy is indicated, with a definitive treatment postponed to a later date. We prefer a transverse colostomy; this leaves a normally innervated segment of bowel, distal to the colostomy, which through disuse will contract to a size suitable for later anastomosis with the rectum. But if the dyskinetic area is proximal to the transverse colon, the choice is between doing nothing, and facing the infant's certain death, and performing an ileostomy, which most infants tolerate poorly. Swenson has performed a successful terminal ileostomy; and at a time when fluid balance was much less well understood than it is now, Dr. W. E. Lee performed terminal ileostomies with surprising success. Nevertheless, infants tolerate poorly any type of small bowel fistula, and the ingenuity of the attending staff is greatly taxed in keeping up with fluid and electrolyte losses. If the infant can be carried over the first 2 weeks, there is apparently a compensatory adjustment in fluid loss which eases the problems of management.

Paralytic ileus of the small intestine must be considered in the differential diagnosis of neurogenic obstruction of the bowel. It may occur with an infarct of the kidney or with retroperitoneal hemorrhage resulting from a traumatic delivery or with spontaneous hemorrhage soon after birth. It is difficult to assess paralytic ileus in the neonatal period. In the presence of apparent obstruction, it is probably safer to perform an exploratory laparotomy, which in retrospect will have been unnecessary, than to procrastinate when laparotomy could have lead to definitive treatment.

Zuelzer and Wilson (47) have called attention to the familial tendency in congenital, neurogenic obstructions of the bowel. Among our cases, in each of 3 families, there have been 3 children with neurogenic obstruction. In 2 of the families, the colon only was affected; in the third, 2 children had small bowel myenteric plexus agenesis and 1 had typical Hirschsprung's disease.

## IMPERFORATE ANUS

The diagnosis of imperforate anus is usually made by observation alone, and usually only a few minutes after birth. Paradoxically, this prompt diagnosis permits the delay of a day, sometimes of 2, before surgery.

There are several varieties of imperforate anus. (1) A transparent membrane behind which meconium can be seen; its perforation is simple, completely effective, and requires no further discussion. (2) A normal anus and rectal canal ending in a blind pouch a few centimeters proximal to the sphincter; this is really an atresia of the colon, and its management is similar to that of other colonic obstructions. (3) Imperforate anus, in which the distance between anal dimple and blind end of colon is 1.5 cm. or less. (4) Imperforate anus in which the distance is greater than 1.5 cm. About the differential diagnosis of the last two and their management are centered the problems of imperforate anus.

To differentiate the high from the low, gas-containing bowel is essential for roentgenographic contrast, and hence the need for some delay after diagnosis before surgical treatment is carried out. If the infant cries and swallows, sufficient air will be taken into the gastrointestinal tract in the first 8 to 12 hours to permit accurate x-ray diagnosis. Roentgenograms at intervals of 8 hours usually show the level of the lesion within 24 hours or occasionally in 36. To prevent excessive distention, suction drainage via a catheter placed in the stomach should be instituted after 12 hours.

Traditionally, roentgenograms for the diagnosis of imperforate anus are taken with the infant held by his heels and with an opaque object over the anal dimple, as described by Wangenstein and Rice (46). However, flexing the thighs on the belly, as advocated by Rhodes (35), may serve better to push gas into the distal bowel (Plate 14). A tight abdominal binder is also helpful. As soon as it is established that the distance between colon and anal dimple is less than 1.5 cm., or when the caliber of the colon increases on successive films without further descent toward the anus, the decision to operate can be made. The procedure is dictated by the distance between anal dimple and colon.

For lesions in which the separation is 1.5 cm. or less, the perineal approach is not only satisfactory, but entails little risk and permits

excellent results. When the separation is more than 1.5 cm., the perineal approach should not be used; a colostomy or definitive surgery by a combined abdominoperineal approach are then the procedures of choice.

A colostomy is lifesaving, simple, and postpones definitive surgery indefinitely. The combined abdominoperineal procedure should not be done unless reliance can be placed on all phases of surgical assistance, including anesthesia and postoperative care. In our practice, the combined operation is used for any infant with a gap of more than 1.5 cm. and a weight of over 4½ lb., in the absence of medical contraindications (22, 34).

Too frequently, the perineal approach is used, with undesirable sequelae, when the combined operation or colostomy would have been preferable. It is possible to bridge a gap of more than 1.5 cm., at the expense of nerve and blood supply to adjacent structures, and pull the mucosa of the colon to the skin edge for suturing. Unfortunately, under tension, these sutures do not hold, the bowel retracts, and a raw canal remains which forms a stricture. Such infants survive their lesion and the surgery, but lifelong problems in defecation and, particularly, in fecal incontinence are initiated.

The foregoing, by and large, refers to males only. In the female, a rectovaginal fistula is commonly associated with an imperforate anus. This prevents intestinal obstruction and may be managed as a fistula by a variety of surgical maneuvers. Males frequently have fistulas also, but these are between the blind pouch of the colon and some portion of the lower urinary tract. Such associated anomalies can be corrected at the time of surgery for imperforate anus by either approach.

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