THE SURGICAL SIGNIFICANCE OF ANOMALIES OF INTESTINAL ROTATION*

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A CLEAR UNDERSTANDING of the normal process of intestinal rotation and of its anomalies is essential to the surgeon. Knowledge of the normal process of rotation and fixation is utilized whenever the colon is mobilized for resection. Unless the surgeon also understands the various possibilities of abnormalities of rotation he may be completely bewildered by the unusual disposition of the viscera when confronted by one of these anomalies and may have to close the abdomen without correcting the condition present.

Anomalies of intestinal rotation are not uncommon. Our interest in them was first aroused in 1934 by an experience with two cases, within a month, of volvulus of the entire mesentery secondary to anomalies of rotation and fixation.[8] In the past year we have seen four infants with volvulus of the entire midgut and one with a huge omphalocele. Because these conditions are still poorly understood by many surgeons it seemed wise to record again the normal steps in the process of intestinal rotation and fixation and to describe abnormalities in each stage as we have encountered them at the Duke Hospital.

Most of our knowledge of intestinal rotation dates from the work of Mall[16] in 1898 and Frazer and Robbins[7] in 1915. Dott[5] was the first to apply this knowledge to practical surgical problems in a masterful article published in 1923. Since then anatomists, pediatricians, gastro-enterologists, roentgenologists, and surgeons have recorded experiences which indicate that anomalies of intestinal rotation are more common than originally believed. When we reviewed the world literature in 1934 105 cases of intestinal obstruction from anomalies of rotation were collected. The literature since then includes at least an equal number of cases, 19 being reported by McIntosh and Donovan.[17]

EMBRYOLOGY

The intestinal tract of the early embryo is a straight structure suspended in the sagittal plane on a common dorsal mesentery. The process by which this primitive position is converted to that seen at birth is called intestinal rotation.

Embryologists divide the primitive intestinal tract into foregut (mouth to duodenojejunal junction), midgut (duodenojejunal junction to midtransverse colon), and hindgut (midtransverse colon to anus). The midgut loop is the one primarily concerned in intestinal obstruction and is the only one under discussion in this presentation. Frazer and Robbins divide the process of

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rotation of this midgut loop into three stages, each of which will be described along with its anomalies.

FIRST STAGE OF MIDGUT ROTATION

Embryology. The primitive intestinal tract is a straight structure suspended on a dorsal mesentery in the sagittal plane (Fig. 1A). As it elongates, the midgut loop bulges through the umbilical orifice into the primitive umbilical cord as a temporary physiologic umbilical herniation. The first stage of intestinal rotation is a contraclockwise rotation of 90° of this intra-umbilical loop from the sagittal (Fig. 1A) to the horizontal plane (Fig. 1B). It occurs at about the eighth week of intra-uterine life.

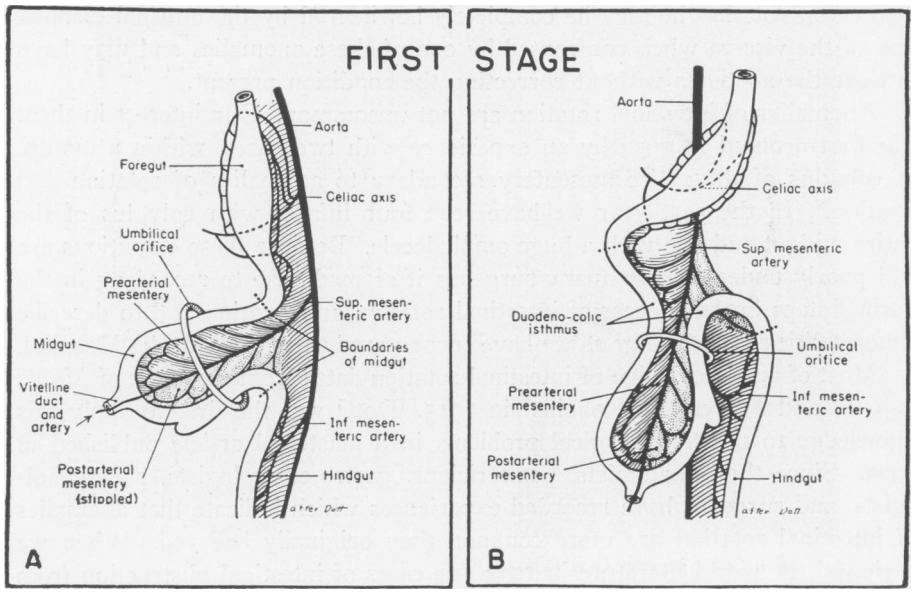

FIG. 1.—First stage of intestinal rotation. (A) Diagrammatic representation of the intestinal tract in the early embryo. The entire intestinal tract hangs suspended in the sagittal plane on a common dorsal mesentery. As the bowel develops the midgut loop bulges into the umbilical stalk as a temporary physiologic umbilical herniation. At about the eighth week of intra-uterine life a 90° contraclockwise rotation of this intra-umbilical, midgut loop from the sagittal plane (A) to the horizontal plane (B) occurs.

Omphalocele. Failure of rotation beyond the first stage and retention of the midgut loop in the umbilical stalk at the time of birth is called omphalocele, amniotic hernia, or exomphalos (Fig. 2). In this condition herniation of intestine and sometimes also of liver and spleen into the umbilical cord is present at birth, the hernial covering being the thin translucent umbilical cord structure.

This anomaly was described by Cullen[3] in his monumental work on the umbilicus as an amniotic hernia. Ladd and Gross[12] refer to it as an ompha-

locele and Dott[5] as exomphalos. It may exist in all degrees of severity. Small herniations may only contain a single loop of bowel. Others, as ours (Fig. 2), may contain the entire intestinal tract, liver, spleen, and pancreas, and be larger than the child's abdominal cavity.

The jellylike cord structure covering such a herniation is delicate and liable to rupture within the first few hours of life. To prevent evisceration it is necessary to close the defect as promptly as possible after delivery. In small herniations peritoneum, fascia, and skin closures can be obtained. In larger herniations it is possible only to close the skin. Hollenberg[10] reports

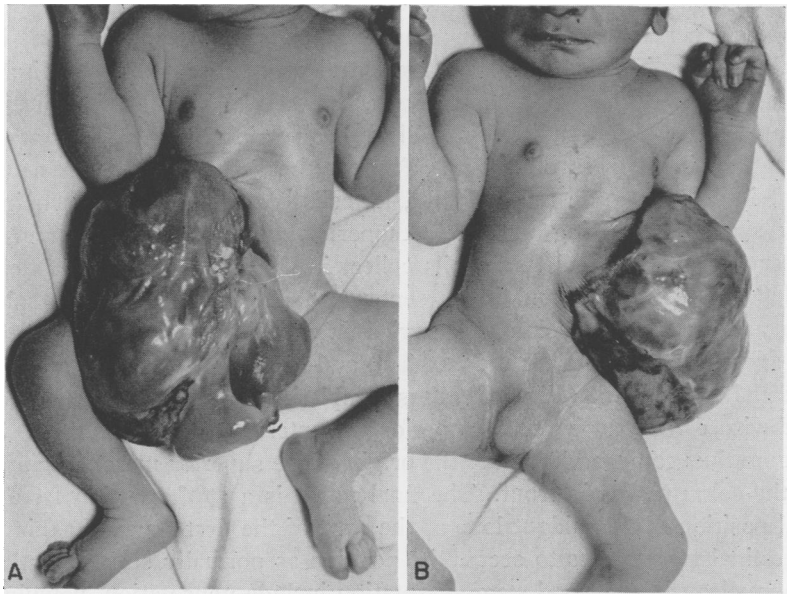

FIG. 2.—Omphalocele (Case 1). Rotation has been arrested in the first stage. The child was born with abdominal viscera retained in the umbilical stalk. Coverings of the hernia are the delicate, jellylike, translucent cord structure. The tie on the umbilical cord is seen at the lower margin of the herniation (A). B shows the transition between skin and cord structure covering the hernia.

four successful cases in which skin was closed over the viscera after the covering membrane had been removed.

Gross[9] more recently has described a technic for the treatment of large omphaloceles in which the amniotic membrane is not removed. Skin mobilized from about the umbilical orifice is utilized to cover the herniated viscera without first removing the covering membrane. After six to 12 months the fascia and muscles are closed over the defect. This technic eliminates evisceration of bowel at operation, and since the viscera are not replaced into the peritoneal cavity there is minimum rise in intra-abdominal pressure, with crowding of the viscera against the diaphragm.

Case 1.—*Omphalocele.* B. B., No. C-46124, a male Negro infant, was seen 12 hours after delivery with a huge omphalocele (Fig. 2). At operation 2 hours later skin over the entire anterior and both lateral portions of the torso was mobilized and used to cover the herniation, no effort being made to remove the translucent cord structure covering the herniation (Gross' technic). The child died 2 hours later. At autopsy the herniation contained liver, pancreas, spleen, and midgut. The lungs were atelectatic. The heart was not completely rotated. The foramen ovale and the ductus arteriosus were patent.

SECOND STAGE OF MIDGUT ROTATION

Embryology. This is the stage when reduction and major rotation of the bowel occurs and is the most important stage of intestinal rotation. The midgut loop returns to the peritoneal cavity from its temporary position in the umbilical herniation and at the same time rotates an additional 180° in a contraclockwise direction about the mesenteric root as a pedicle (Fig. 3). This stage occurs quickly, at about the tenth week of intra-uterine life, and in none of the models studied by Mall or by Frazier and Robbins was the gut found in its process of return. The proximal limb of the pre-arterial segment is thought to be reduced first, its coils entering the abdomen in an orderly sequence, passing under the superior mesenteric vessels and the mesentery (Fig. 3B). As these coils collect in the left side of the abdomen they deflect the hindgut and its mesentery to the left so that the splenic flexure and descending colon are carried into their normal position. The cecum and adjacent colon are reduced last, and as the colon straightens out it is deflected to the right (Fig. 3C), thus completing a 180° rotation in a contraclockwise direction about the superior mesenteric artery as an axis. The duodenum thus comes to lie under the origin of the superior mesenteric artery while the colon passes in front. The final result is a 270° rotation from the sagittal position of the midgut loop at the start of the first stage. In this way the intestinal tract comes to occupy its position as normally seen at birth.

Anomalies of the second stage of rotation include nonrotation, volvulus of the midgut, malrotation, internal hernia, and reversed rotation.

Nonrotation. In nonrotation the midgut loop is returned to the peritoneal cavity from the temporary umbilical herniation without having rotated beyond the horizontal plane it occupied at the end of the first stage (Fig. 1B). The duodenum descends on the right of the superior mesenteric artery (Fig. 4A). The small intestine is entirely in the right side of the abdomen and the colon is on the left. The cecum is in the left lower quadrant. The terminal ileum crosses the midline to enter the cecum from the right. From this point the ascending colon passes upward on the left of the midline to a point behind the greater curvature of the stomach. Between this point and the splenic flexure is a narrow U-shaped loop of transverse colon.

Nonrotation may exist without symptom. Roentgenologists frequently see it as an incidental finding during examination of the barium filled colon and refer to it as "left-sided colon." Its major surgical significance occurs when diseases of the appendix, cecum or ascending colon occur in an unusual position.

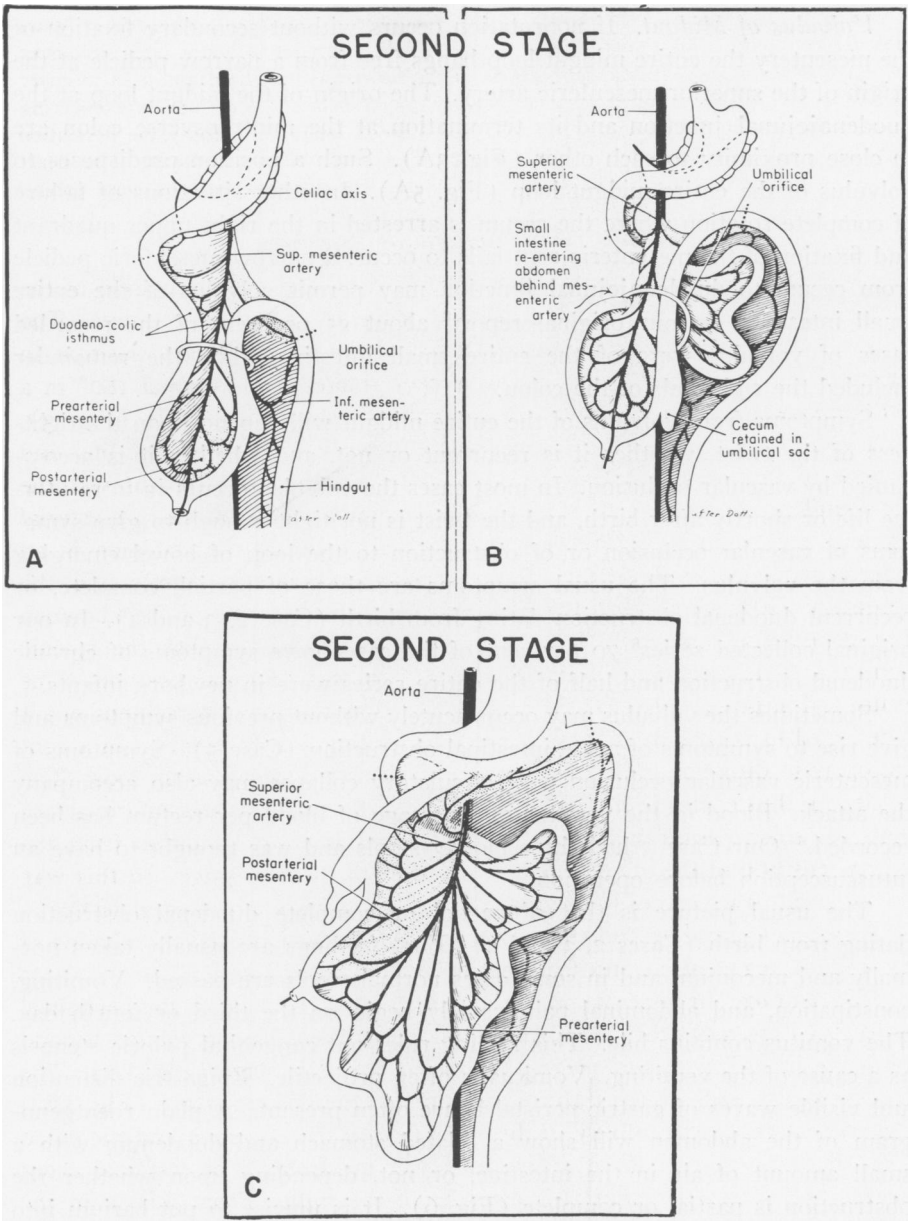

FIG. 3.—Second stage of intestinal rotation. The midgut loop which occupies a position in the umbilical stalk at the beginning of the second stage (A) is reduced back into the peritoneal cavity (B) and in so doing rotates 180° in a contraclockwise direction about the superior mesenteric artery as an axis. The proximal loop of midgut re-enters the peritoneal cavity under the mesenteric root first (B). An orderly reduction of the remainder of the midgut loop then occurs, the cecum and ascending colon being reduced last. As the cecum and ascending colon straighten out they are deflected to the right upper quadrant, completing the 180° contraclockwise rotation of the midgut loop about the mesenteric root. In this way the duodenum is thrown under the superior mesenteric artery and the colon passes in front of it. This stage of rotation occurs quickly at about the tenth week of intra-uterine life.

Volvulus of Midgut. If nonrotation occurs without secondary fixation of the mesentery the entire midgut loop hangs free from a narrow pedicle at the origin of the superior mesenteric artery. The origin of the midgut loop at the duodenojejunal junction and its termination at the midtransverse colon are in close proximity to each other (Fig. 3A). Such a position predisposes to volvulus of the entire midgut loop (Fig. 5A). In other situations of failure of complete rotation where the cecum is arrested in the right upper quadrant and fixation of the mesenteric root fails to occur, a narrow mesenteric pedicle from cecum to duodenojejunal junction may permit volvulus of the entire small intestine. In our original report[8] about 25 per cent of the recorded cases of volvulus were of the entire small intestine while the remainder included the right half of the colon.

Symptoms from volvulus of the entire midgut will depend upon the tightness of the twist, whether it is recurrent or not, and whether it is accompanied by vascular occlusion. In most cases the volvulus occurs in intra-uterine life or shortly after birth, and the twist is not tight enough to give symptoms of vascular occlusion or of obstruction to the loop of bowel emerging from the volvulus. The usual symptoms are those of partial, complete, or recurrent duodenal obstruction dating from birth (Case 2, 3 and 4). In our original collected series[8] 70 per cent of the cases gave symptoms of chronic duodenal obstruction and half of the entire series were in newborn infants.

Sometimes the volvulus may occur acutely without previous symptoms and give rise to symptoms of acute intestinal obstruction (Case 5). Symptoms of mesenteric vascular occlusion with circulatory collapse may also accompany the attack. Blood in the vomitus and passage of blood per rectum has been recorded.[8] Our Case 5 had three bloody stools and was thought to have an intussusception before operation.

The usual picture is that of partial or complete duodenal obstruction dating from birth (Cases 2, 3 and 4). First feedings are usually taken normally and meconium and in some cases normal stools are passed. Vomiting, constipation, and abdominal pain usually begin on the third or fourth day. The vomitus contains bile. This finding rules out congenital pyloric stenosis as a cause of the vomiting. Vomitus is often projectile. Epigastric distention and visible waves of gastric peristalsis are often present. A plain roentgenogram of the abdomen will show a dilated stomach and duodenum with a small amount of air in the intestine, or not, depending upon whether the obstruction is partial or complete (Fig. 6). It is unwise to put barium into the stomach. If the roentgenologist is able to demonstrate that the entire duodenum is on the right side of the vertebral column or that the colon is entirely on the left an anomaly of rotation can be suspected. Usually it is impossible to determine before operation whether the duodenal obstruction is caused by atresia, stenosis, extrinsic bands, or a volvulus of the midgut.

Operative treatment offers the only hope of cure for patients with volvulus of the midgut. In those with symptoms of intestinal obstruction or vascular occlusion immediate operation is imperative (Case 5). In the larger group,

with symptoms of chronic duodenal obstruction, a more leisurely preparation for operation may be pursued. At operation the volvulus of the entire mesentery may be easily overlooked if adequate exploration is not done. The condition may be suspected if the right half of the colon is not found in its normal position or if on palpation a firm cord representing the twisted root of the mesentery can be felt at the site of origin of the superior mesenteric artery. Complete evisceration of all of the intestine is advised as the quickest method of determining the true nature of the condition present. Detorsion of the volvulus is easily accomplished. All of our patients had adhesions attaching bowel and mesentery of the entering and emerging loops of intestine which had been involved in the volvulus (Fig. 5B). These adhesions must be divided as in freeing two adherent leaves of a book. Adhesive bands may also extend from the colon across the duodenum to the posterior abdominal wall. They must be divided, as well as the adhesions uniting entering and emerging loops of bowel, in order that the duodenum be completely freed and separated widely from the colon. If these adhesions are not freed recurrence of the volvulus may occur.

Chronic venous obstruction which has accompanied the volvulus may cause enormous distention of the mesenteric veins (Fig. 5C). Care must be taken not to damage these veins when adherent leaves of mesentery are separated. Lymph nodes in the mesentery are usually much enlarged by the lymphatic obstruction which accompanied the volvulus. In our Case 5 dilated lymphatics could be seen in the mesentery and there was a little chylous free fluid in the peritoneal cavity.

After being freed the bowel is returned to the abdomen in a state of nonrotation, with the duodenum descending on the right of the vertebral column and the colon ascending on the left (Fig. 5C). Entering and emerging loops of bowel involved in the volvulus which were previously adherent are separated as widely as possible when the bowel is replaced. No effort to fix the bowel has been attempted in any of our cases and none of our patients have had recurrence of their volvulus. Recurrences have been reported in cases where adhesions between loops of bowel in the volvulus or adhesive bands crossing the duodenum were not freed.[2, 13, 20] Wangensteen[21] has reported a method of fixation of the bowel in a position of normal rotation. Some effort to fix the nonrotated cecum and ascending colon along the left lumbar gutter would seem to be a simpler method of preventing recurrence of the volvulus, if any fixation is necessary.

Case 2.—*Volvulus of Midgut.* W. D., No. C-48290, was an 8-week-old Negro male with history of vomiting of bile-stained material since the age of 6 days. Bowel movements had been normal. On examination he was dehydrated and undernourished. Roentgen rays showed a stomach hugely distended with gas (Fig. 6A). At operation a 360° clockwise twist of midgut loop was encountered (Fig. 5A). After detorsion and separating of adherent entering and emerging loops of bowel, the bowel was replaced in the abdomen so that the small intestine was in the right half and colon in the left

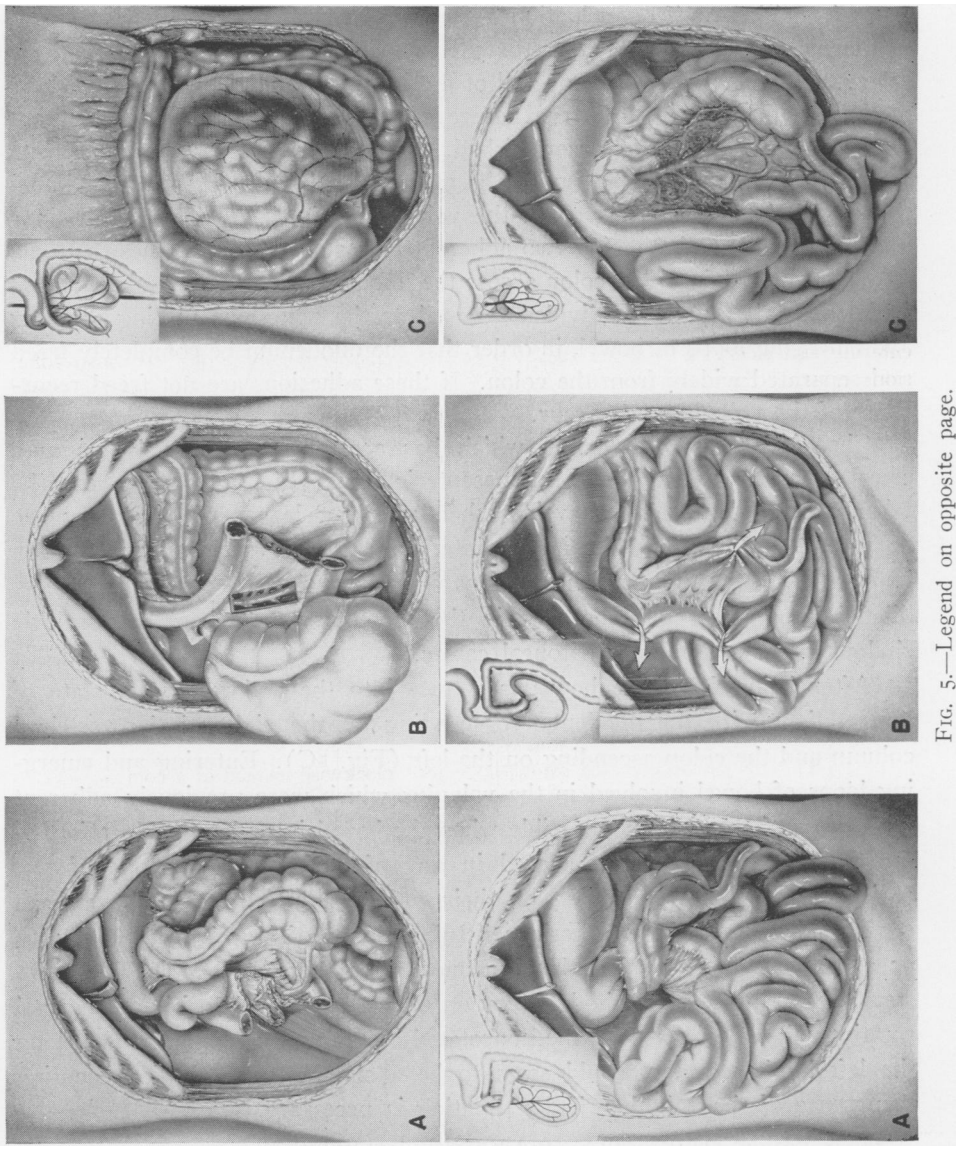

FIG. 4.—Legend on opposite page.

FIG. 5.—Legend on opposite page.

half of the abdomen (Fig. 5C). Following operation he took feedings without vomiting and was discharged, well, on the sixteenth postoperative day.

Case 3.—*Volvulus of Midgut.* B. T., No. C-47820, a 5-day-old Negro male, had a story of vomiting of yellowish material after each feeding since the second day of life. On examination he was dehydrated and weighed 2720 Gm. The outline of a distended stomach was visible in the epigastrium. Roentgen rays (Fig. 6B) showed the stomach full of air and practically no air in the small intestine. At operation exactly the same situation as in Case 1 was found. Following operation he did poorly, became jaundiced and failed to retain feedings well. The operative wound became infected. He died on the twentieth postoperative day.

Case 4.—*Volvulus of Midgut.* J. W., No. C-63611, was a 9-day-old white male. He had vomited since birth and at the age of 4 days had been operated upon elsewhere for pyloric stenosis, but none was found. He continued to vomit biliary material after operation and barium given by mouth 24 hours before we saw him was retained in the stomach and first part of the duodenum (Fig. 6C). His weight on admission was 2553 Gm. At operation a 360° clockwise twist of the midgut loop was found. After detorsion, entering and emerging loops of bowel were found adherent, and there were also adhesions crossing the duodenum to the kidney fossa. These were all freed and the bowel returned to the abdomen in a position of nonrotation. Following operation he did not vomit, but took feedings slowly. He gradually gained in weight and strength and was discharged in good condition on his twenty-seventh postoperative day.

Case 5.—*Volvulus of Midgut.* J. B., No. C-64536, a 6-week-old Negro male infant who had been in excellent health since birth suddenly began having abdominal pain 12 hours before admission. Six hours later he had a bowel movement consisting almost

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FIG. 4.—Abnormalities of the second stage of intestinal rotation.

(A) Nonrotation. The midgut loop has not rotated, the disposition of the viscera being essentially the same as at the end of the first stage of rotation (Fig. 1B) except that the bowel has returned to the abdominal cavity. The colon is entirely on the left and the small bowel on the right of the abdominal cavity.

(B) This shows reversed rotation. A 180° clockwise instead of a contraclockwise rotation during the second stage has occurred. The transverse colon thus comes to lie under the superior mesenteric artery while the duodenum passes in front of it. Subsequent fixation of the mesenteric root has trapped the colon under it.

(C) This shows internal hernia (Case 6). Most of the small intestine is enclosed in a peritoneal sac. The anomaly occurs during the second stage of rotation. When the midgut is reduced from the temporary umbilical herniation the bowel bulges into its own post arterial mesentery (insert Fig. 4C) instead of entering the free peritoneal cavity as it should (Fig. 3B). The wall of the sac is thus the mesentery of the terminal ileum and ascending colon.

FIG. 5.—Volvulus of the midgut (Case 2). Drawings made from sketches and photographs at the operation.

(A) Appearance on opening the abdomen of a 2-month-old infant with signs of chronic duodenal obstruction since birth. A 360° clockwise rotation of the midgut loop about the mesenteric root has occurred. The duodenum is obstructed at the volvulus and is dilated above it. Small bowel loops are not distended.

(B) Appearance on reduction of the volvulus. The mesentery and bowel of the entering loop of jejunum and emerging loop of colon are adherent. Sometimes adhesive bands also cross the duodenum in the region of the upper arrow.

(C) Appearance after separation of adherent bowel and mesentery of entering and emerging loops of the volvulus. Veins are distended and mesenteric lymph nodes enlarged by the chronic obstruction. The bowel is replaced in the abdomen in this position at the termination of the operation. The duodenum descends on the right of the vertebral column and all small bowel is on the right. The colon is entirely in the left side of the abdomen. This is the position of nonrotation.

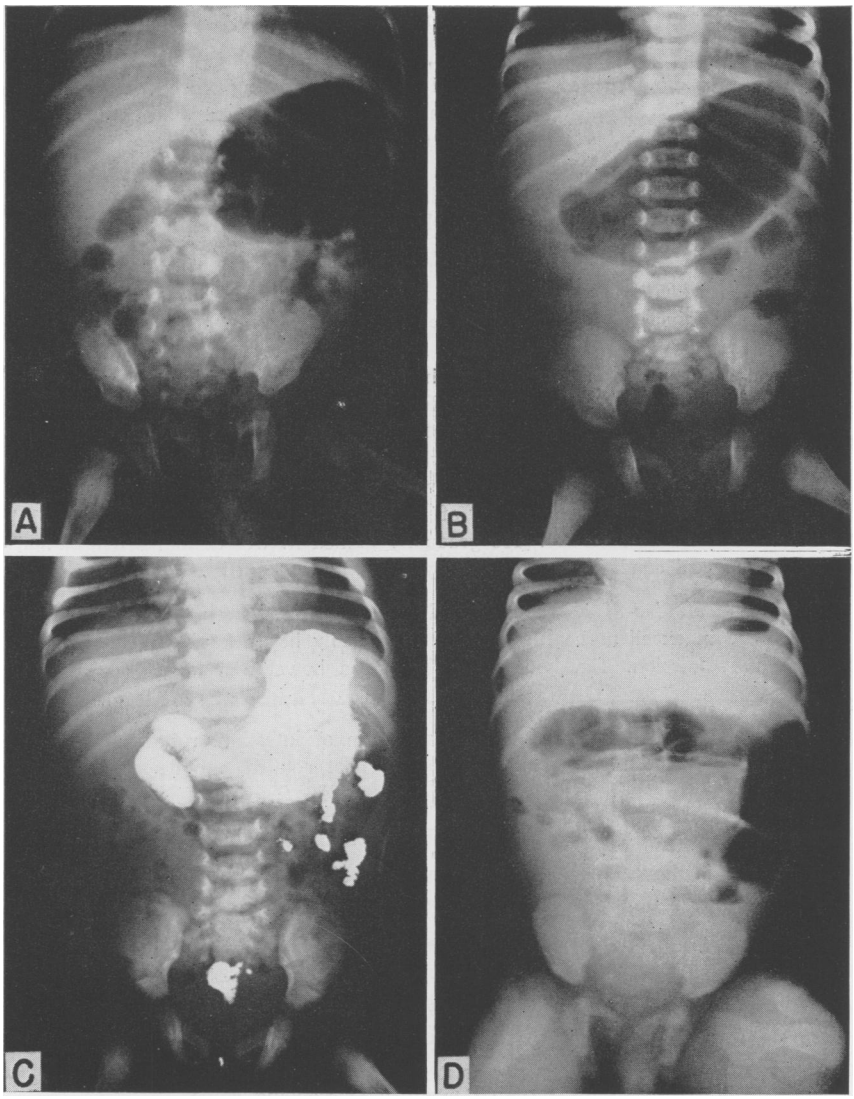

FIG. 6.—Roentgen rays of the abdomen in four infants with volvulus of the midgut is shown. (A)—Case 2. A 2-month-old infant with signs of chronic duodenal obstruction since birth. The stomach is distended with air and there is some gas in the bowel. (B)—Case 3. A 5-day-old infant with a story of biliary vomiting since birth. The stomach is distended with air. There is also a little gas in the colon, probably introduced by enema. (C)—Case 4. A 9-day-old infant with vomiting since birth who had been operated upon elsewhere on the fourth day for pyloric stenosis, but none was found. He was given barium by mouth 24 hours before being sent to us. This roentgen ray shows 24-hour retention of barium in stomach and duodenum with passage of a small amount of barium into the intestinal tract. (D)—Case 5. A 6-week-old infant with signs of acute intestinal obstruction of 12 hours duration. He had had three bloody bowel movements and was thought to have an intussusception before operation. The roentgen ray shows dilated loops of bowel full of gas in the left upper abdomen.

entirely of blood. Subsequently he had 2 more bloody bowel movements. He vomited several times. On admission 12 hours after onset of pain the child was lethargic and acutely ill. The abdomen was distended. No peristaltic sounds were heard. Roentgen rays showed dilated loops of bowel in the left upper abdomen (Fig. 6D). The stomach contained only a tiny gas shadow. A preoperative diagnosis of intussusception was made and immediate operation done. At operation volvulus of the entire midgut 540° in a clockwise direction was demonstrated. Bowel in the volvulus was of good color. There was a little chylous fluid in the peritoneal cavity. Lymphatics in the mesentery were dilated. Detorsion and division of adhesions extending from midtransverse colon across duodenum to right flank were divided and the bowel returned to the abdomen in the position of nonrotation. His postoperative course was satisfactory and he was discharged on the sixteenth postoperative day.

Malrotation. Innumerable irregular defects in intestinal rotation and fixation during the second stage are possible and are grouped together under the term malrotation. They are not difficult to understand, or to unravel at operation, if the normal stages of intestinal rotation are kept in mind.

If fixation of the small bowel mesenteric root over a wide base from Treitz fossa to the right iliac fossa fails to occur, volvulus of the entire mesentery is possible. Or the mesenteric root during the process of its fixation may trap a loop of bowel under it and give the appearance of a mesenteric defect. Other abnormalities of intestinal fixation may kink or compress the lumen of the intestinal tract at any level. These have been described most frequently in the duodenum. Ladd[14] has stressed the importance of inspecting the duodenum in all cases with abnormalities of rotation in order that anomalous bands may be relieved. In other cases the nonrotated cecum may be firmly fixed to, and may partially occlude, the duodenum or small intestine. Or the intraperitoneal bands and adhesions which occur regularly with anomalies of intestinal rotation may unite various combinations of intra-abdominal organs and be the cause of obstructive symptoms.

The inherent possibilities of irregularities of rotation and fixation are unlimited. Symptoms caused by them, if any, may be equally as varied but are usually those of partial or complete obstruction from volvulus or adhesive bands. Usually the true nature of the condition present is recognized only at operation. Waugh[22] emphasizes the "unusualness" of the symptoms, the "emptiness" of the right iliac fossa as a result of absence of the cecum from its normal position, and the roentgen findings after a barium meal as important findings which enable him to make the correct preoperative diagnosis in four of his five cases.

Internal Hernia. Complicated explanations of the origin of paraduodenal, retroperitoneal or internal herniations occur in the literature[18] which describe them as occurring in any one of as many as nine fossae about the duodenojejunal junction and four in the cecal area. Their explanation on the basis of an anomaly of intestinal rotation is much simpler and was first offered by Andrews[1] in 1923. He explained the origin of the paraduodenal hernia as imprisonment of small intestine under the mesentery of the right colon during the process of fixation of the midgut loop after its rotation.

Haymond and Dragstedt[11] in careful autopsy dissections of a case with large internal hernia previously observed at operation, showed that the abnormality was one of malrotation during the second stage of intestinal rotation. The essential feature of the abnormality was a rotation of the bowel into the mesentery of its postarterial segment (later to become the mesentery of the ascending colon (inset Fig. 4C) instead of into the free peritoneal cavity, during the phase of intestinal rotation when the midgut loop was reduced back into the peritoneal cavity from the umbilical orifice. In their case and in ours (Fig. 4C) almost the entire small intestine was in the sac. The amount of bowel involved may vary from a short loop to the entire small intestine.

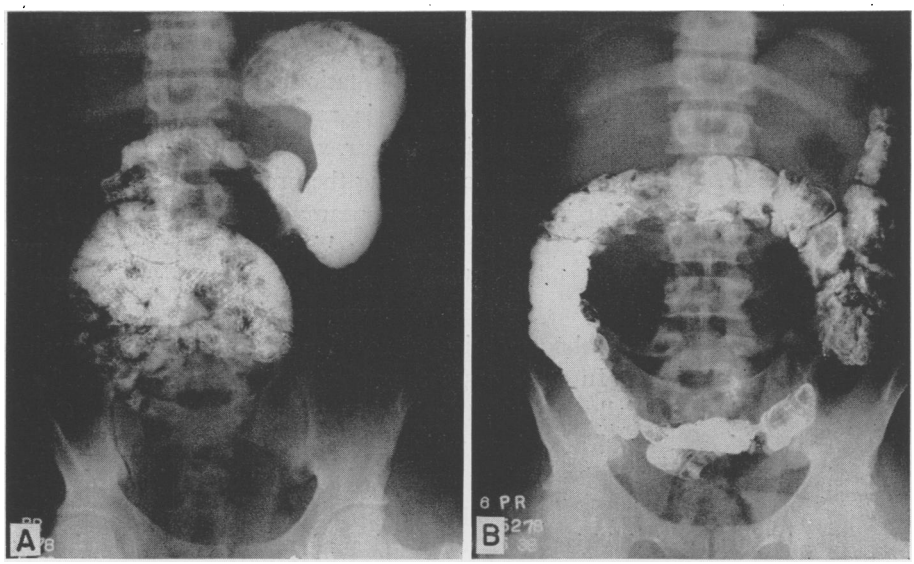

FIG. 7.—Roentgen rays of the barium filled stomach and intestinal tract in a patient (Case 6) with large internal hernia. (A) The small intestine appears to be enclosed in a circular pocket in the center of the abdomen. (B) This pocket is surrounded by colon of the midgut loop (right half of the colon). The splenic flexure and descending colon (hindgut) are normally placed.

Many of the patients with internal hernia have no symptoms and the condition is recognized only incidentally at operation, anatomic dissection or autopsy. If symptoms occur they are usually those of partial or intermittent intestinal obstruction.

In our case precise diagnosis of the condition present was made by our X-ray Department[19] who visualized the barium-filled small intestine enclosed in a circular pocket in the center of the abdomen (Fig. 7A). This pocket was surrounded by colon of the midgut loop (right half) (Fig. 7B). The splenic flexure and descending colon were normally placed.

At operation an adequate exposure, knowledge of embryonic origin, and delivery of as much bowel as is necessary will usually make the situation

clear. Since the wall of the sac is the mesentery of the postarterial segment of the midgut loop (terminal ileum and right colon) care must be taken not to damage its blood vessels. The recommended procedure is to withdraw the bowel from the internal hernial sac and to close its mouth with as many sutures as are necessary.[15]

Case 6.—*Internal Hernia.* J. M., No. A-5278, a 28-year-old Negro male, was admitted with a story of 5 attacks of cramping periumbilical pain during the preceding 2 months. Each attack was precipitated by large meals and was accompanied by vomiting of the food eaten at the preceding meal. Each attack subsided after 6 or 8 hours, during which he usually had to have a hypodermic injection. The physical examination showed nothing of significance. Roentgen rays (Fig. 7) showed the small intestine apparently to be enclosed in a round sac in the midabdomen, which was surrounded by the right half of the colon. At operation almost the entire small intestine was enclosed in a peritoneal sac (Fig. 4C) whose walls formed the mesentery of the right half of the colon. The small bowel could be withdrawn from the sac without difficulty, after which the opening of the sac was occluded by suture. Postoperative course was uneventful.

Reversed Rotation. In this condition a clockwise instead of a contraclockwise rotation of 180° occurs during the second stage. The transverse colon thus comes to lie under the superior mesenteric artery and the duodenum above it (Fig. 4B). If normal fixation of the root of the mesentery toward the right iliac fossa takes place with the bowel in this position the transverse colon becomes trapped in a tunnel beneath this acquired attachment. Fixation of the cecum and ascending colon is usually incomplete and torsion of the mobile right half of the colon with obstruction of the transverse colon at the site of the tunnel through the root of the mesentery may occur.

True reversed rotation is not common. McIntosh and Donovan[17] collected 16 cases and added one of their own. We have never had such a case. The illustration (Fig. 4B) is of Dott's patient, a 68-year-old man with symptoms of acute colonic obstruction of three days duration.

THIRD STAGE OF MIDGUT ROTATION

Embryology. This stage is characterized by descent of the cecum from its subhepatic position to the right lower quadrant and fixation of the mesentery of cecum and ascending colon in the right flank (Fig. 8B). Fixation of the descending colon and of the lower portion of the duodenum also occurs in this stage which is completed about the time of birth.

The important feature of this stage is the fixation of the ascending colon and cecum with its mesentery in the right iliac fossa and with it the fixation of the root of the small bowel mesentery on a wide base from left upper to right lower quadrants. The midgut loop originally dependent from a narrow pedicle at the origin of superior mesenteric artery now acquires a broad oblique attachment to the posterior abdominal wall. It is the absence of this attachment which predisposes to volvulus of the entire mesentery.

Anomalies of the third stage include subhepatic cecum, retrocecal appendix and mobile cecum.

Subhepatic Cecum. If the cecum fails to elongate and descend into the right iliac fossa it remains in a subhepatic position (Fig. 9A). If with the cecum in this position there is also failure of fixation of the small bowel mesentery on a broad attachment to the posterior abdominal wall, volvulus of all of the small intestine may occur. The only other significance of this anomaly is that appendicitis, if it occurs in such a position, may be unrecognized if the possibility of a high lying cecum is not kept in mind.

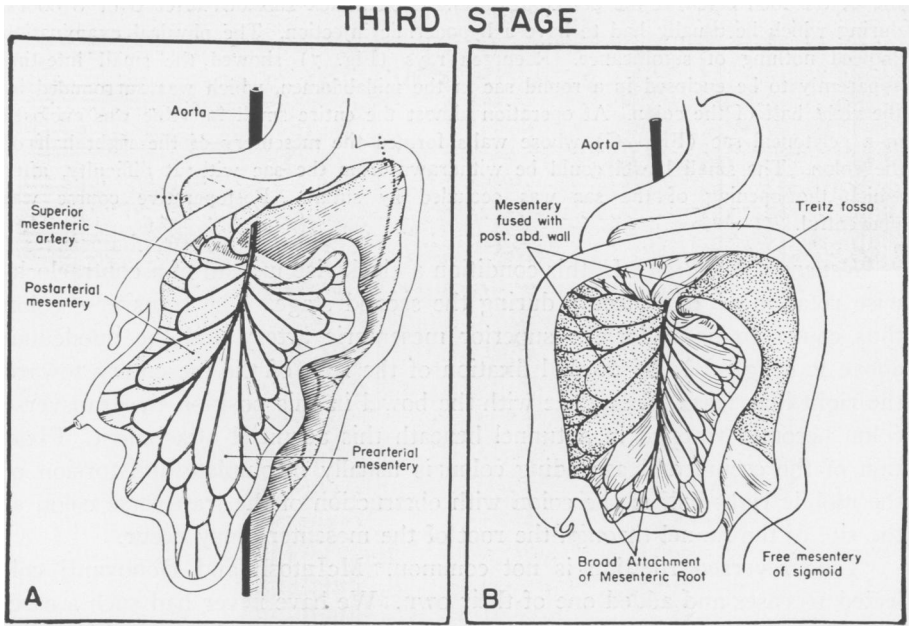

FIG. 8.—Third stage of intestinal rotation. This stage is characterized by descent of the cecum for its subhepatic position (A) to the right lower quadrant and fusion of its mesentery with the peritoneum of the right flank (B). The important feature is the fixation of the cecum and ascending colon in the right flank which gives a broad attachment of the root of the small bowel mesentery from Treitz fossa to right lower quadrant. The midgut loop originally dependent from a narrow pedicle at the origin of the superior mesenteric artery now acquires a broad oblique attachment to the posterior abdominal wall. It is the absence of this attachment which predisposes to volvulus of the entire mesentery. This stage is completed at birth.

Retrocecal Appendix. If the appendix is drawn under the cecum as the cecum descends into the right lower quadrant it may become fixed in a retrocecal position during the normal process of fixation of the cecum and ascending colon to the peritoneum of the right flank. This is a rather common anomaly and is well recognized by surgeons, although they usually fail to consider it an anomaly of rotation.

Mobile Cecum. Failure of fusion of the cecum and ascending colon and its mesentery to the peritoneum of the right iliac fossa in a normal manner allows

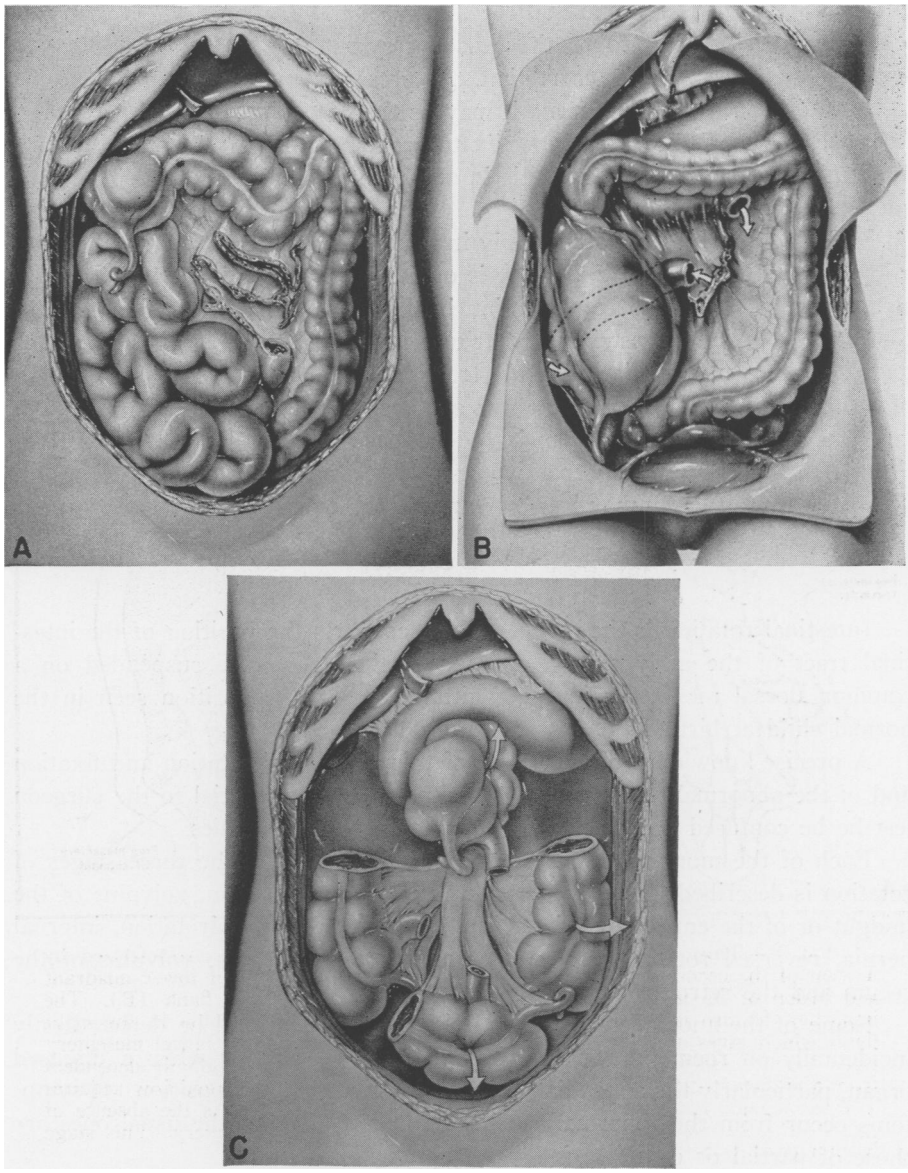

FIG. 9.—Abnormalities of the third stage of intestinal rotation.

(A) Subhepatic cecum. The cecum is arrested in the subhepatic position as at the end of the second stage of rotation. Since fixation of cecum and ascending colon in the right flank has not occurred, the mesentery of the small intestine fails to attain a broad fixation to the posterior abdominal wall from left upper to right lower quadrants and remains suspended from a narrow pedicle at the origin of the superior mesenteric artery. This may predispose to volvulus of the entire small intestine.

(B) Volvulus of the cecum. The cecum and ascending colon are mobile and have twisted 180° in a clockwise direction about the long axis of the ascending colon. The cecum is distended. Small bowel also usually becomes distended. Barium introduced by rectum would show a block at the hepatic flexure.

(C) Mobile cecum. Four hypothetical positions which a mobile ileocecal segment may occupy within the abdomen are shown. The mobile cecum may predispose to volvulus of the cecum. Its most dangerous consequence may be from the anomalous position of the appendix.

undue mobility of the cecum and ascending colon (Fig. 9C). This was formerly in itself thought to be the cause of symptoms, and numerous operations were devised for its correction. At present a mobile cecum is thought to be of significance only because it may be the seat of volvulus of the cecum or because it may allow the appendix to occupy a position almost any place in the abdomen so that appendicitis in an anomalously located appendix may go unrecognized.

Volvulus of the cecum occurs only in a mobile cecum. The twist is usually a longitudinal one about the long axis of the ascending colon[4, 23] (Fig. 9B), although there may also be a rotation in the oblique axis so that the cecum comes to lie in the epigastrium or left upper quadrant. The signs and symptoms are those of acute intestinal obstruction, often with enormous dilatation of the cecum. Roentgenograms show evidence of dilated small bowel plus a large cecal gas shadow. Barium introduced by enema will show an obstruction in the region of the hepatic flexure. At operation detorsion is done. Cecopexy or cecostomy may be necessary. If the bowel is gangrenous it must be resected.

SUMMARY

Intestinal rotation is the process which converts the position of the intestinal tract of the early embryo from a straight structure suspended on a common dorsal mesentery in the sagittal plane to the position seen in the normal child at birth.

A precise knowledge of the stages of this process of rotation and fixation and of the abnormalities which may occur in each is essential to the surgeon lest he be confused when confronted by one of the anomalies.

Each of the more common anomalies which occur in the three stages of rotation is described. They include omphalocele, nonrotation, volvulus of the midgut or of the entire small intestine, various types of malrotation, internal hernia, reversed rotation, subhepatic cecum, mobile cecum, volvulus of the cecum and the retrocecal appendix.

Some of the anomalies may exist without symptom and be detected only incidentally on roentgen ray examination, at operation, or when a diseased organ, particularly the appendix, is found in an anomalous position. If symptoms occur from the abnormality of rotation and fixation the usual ones are those of partial or complete intestinal obstruction.

Volvulus of the entire midgut or of the entire small intestine is the most common cause of intestinal obstruction secondary to anomalies of intestinal rotation. Symptoms depend upon the tightness of the twist and may be those of duodenal obstruction or of acute intestinal obstruction with or without signs of mesenteric vascular occlusion. The usual symptoms are partial or complete duodenal obstruction dating from birth. At operation evisceration, detorsion of the volvulus and release of adhesions uniting entering and emerging loops of bowel and of adhesive bands which may run across the duodenum is necessary. If these adhesions are not divided recurrence of the

volvulus may occur. The bowel is then returned to the abdomen in a position of nonrotation with the small intestine in the right abdomen and the colon on the left.

Omphalocele, amniotic hernia or exomphalos is the only anomaly in the first stage of rotation. The delicate umbilical cord structure covering such a hernia may rupture within the first few hours of life. To prevent evisceration it is necessary to close the defect as soon as possible after delivery.

Malrotation is the term used to designate the innumerable possible irregularities in rotation and fixation which may occur during the second stage. They include a wide variety of anomalous kinks, adhesive bands, and irregular fixations which are not difficult to unravel if the normal process of rotation is understood.

Paraduodenal, retroperitoneal or internal hernia is best explained as an irregularity of rotation wherein the midgut loop rotates into its own mesentery instead of into the free peritoneal cavity during the second stage. Since the wall of the sac is the mesentery of the ascending colon and terminal ileum its vessels must not be disturbed at operation.

Irregularities of descent and fixation of the cecum and ascending colon in the third stage are the most common anomalies of intestinal rotation. They include the subhepatic cecum, mobile cecum, and the retrocecal appendix. The most important feature of these abnormalities is that the appendix may be in an anomalous position.

BIBLIOGRAPHY

[1] Andrews, E.: Duodenal Hernia—A Misnomer. Surg., Gynec. & Obst., **37**: 740, 1923.

[2] Brenner, E. C.: Total Volvulus. Am. J. Surg., **16**: 34, 1932.

[3] Cullen, T. S.: The Umbilicus and Its Diseases. Philadelphia, W. B. Saunders Co., 1916.

[4] Donhauser, J. L., and S. Atwell: Volvulus of the Cecum with a Review of 100 Cases and a Report of Six New Cases. Arch. Surg., **58**: 129, 1949.

[5] Dott, N. M.: Anomalies of Intestinal Rotation: Their Embryological and Surgical Aspects, with Report of Five Cases. Brit. J. Surg., **11**: 251, 1923.

[6] Elman, R.: Ladd's Operation for the Cure of Incomplete Rotation and Volvulus of the Small Intestine Producing Duodenal Obstruction in Infancy. Ann. Surg., **112**: 234, 1940.

[7] Frazer, J. E., and R. H. Robbins: On the Factors Concerned in Causing Rotation of the Intestine in Man. J. Anat. & Physiol., **50**: 75, 1915.

[8] Gardner, C. E., Jr., and D. Hart: Anomalies of Intestinal Rotation as a Cause of Intestinal Obstruction. Arch. Surg., **29**: 942, 1934.

[9] Gross, R. E.: A New Method for Surgical Treatment of Large Omphaloceles. Surgery, **24**: 277, 1948.

[10] Hollenberg, H. G.: Amniotic Hernia. Surgery, **23**: 363, 1948.

[11] Haymond, H. E., and L. R. Dragstedt: Anomalies of Intestinal Rotation. Surg., Gynec. & Obst., **53**: 316, 1931.

[12] Ladd, W. E., and R. E. Gross: Abdominal Surgery in Infancy and Childhood. Philadelphia, W. B. Saunders Co., 1941.

[13] Ladd, W. E.: Congenital Obstructions of the Duodenum in Children. New England J. Med., **206**: 277, 1932.

[14] ————: Congenital Duodenal Obstruction. Surgery, **1**: 878, 1937.

[15] Lahey, F. H., and W. Trevor: Right Paraduodenal Hernia. Ann. Surg., **122**: 436, 1945.
[16] Mall, F. P.: Development of the Human Intestine and Its Position in the Adult. Bull. Johns Hopkins Hosp., **9**: 197, 1898.
[17] McIntosh, R., and E. J. Donovan: Disturbances of Rotation of the Intestinal Tract. Am. J. Dis. Child., **57**: 116, 1939.
[18] Moynihan, B. G. A.: On Retroperitoneal Hernia. New York, William Wood Co., 1906.
[19] Reeves, R. J., F. T. Moran and P. A. Jones: Right Paraduodenal Hernia with Roentgen Diagnosis and Recovery. Am. J. Roentgenol., **59**: 338, 1948.
[20] Stewart, J. S.: Congenital Extrinsic Duodenal Obstruction in the New Born. South. Surg., **14**: 15, 1948.
[21] Wangensteen, O. H.: New Operative Techniques in the Management of Bowel Obstruction. Surg., Gynec. & Obst., **75**: 675, 1942.
[22] Waugh, George E.: Congenital Malformations of the Mesentery: A Clinical Entity. Brit. J. Surg., **15**: 438, 1927.
[23] Weinstein, Mandel: Volvulus of the Cecum and Ascending Colon. Ann. Surg., **107**: 248, 1938.

DISCUSSION—DR. ROBERT L. RHODES, Augusta, Ga.: I did not get the name of the author referred to by the previous speaker, but I rise simply to call attention to two things. One is the monumental contribution to this subject made by one of our own members and a former President. The opening comment of Doctor James E. Thompson in his Presidential Address "Surgery and Embryology" was very similar to the closing comment of our speaker. That is, if one notices and is familiar with these malformations, he does not become lost when he encounters them in the course of an operation.

This was a most excellent presentation, backed up with all types of roentgen rays which, of course, Doctor Thompson did not have the benefit of in 1919. I may say also that I presented a paper on this subject before the first Sectional Meeting of the American College of Surgeons in Birmingham in 1922, under the title of "Arrested Development of the Colon," the purpose being to emphasize what may happen, so that one will not be at a loss when faced with these conditions.

DR. J. W. DUCKETT, Dallas, Texas: There is probably no field of surgery in which the dictum of Halsted, of careful, gentle handling of tissues and complete hemostasis, is more important than in pediatric surgery; of course it is important in all surgery, but here it is essential to achieve such successful results as Doctor Grove and Doctor Gardner have reported in their series of cases. At the Dallas Children's Center we have had some experience with this type of lesion; actually, 15 cases to date since the Center was opened five years ago have grouped themselves into three series of five each. There were five cases of atresia of the duodenum, all of which were low, fortunately, and easily diagnosed early, and all were treated with duodenojejunostomy, as Doctor Grove has mentioned. However, the five atresias of the jejunum and ileum were all treated unsuccessfully, with death, as has been the case in the great majority of such lesions. Our five extrinsic type of obstruction have also been relatively simple, and all have survived.

I call this to your attention because of the fact that it has been universally found that atresias of the ileum and jejunum respond to surgery poorly. This is due to the fact that the atresias are usually multiple and extensive, and the distal segment of the bowel is often poorly developed; it may be hypoplastic or, if not, it is at least very small, collapsed and thin-walled, so that entero-anastomosis is extremely difficult. Even if it can be done technically, with an anastomosis resulting, function is often poor, as Doctor Gardner said.