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Congenital anomalies of the gastrointestinal tract causing obstruction

Bryce G. Shopp
University of Nebraska Medical Center

CONGENITAL ANOMALIES OF THE GASTROINTESTINAL TRACT CAUSING OBSTRUCTION

BRYCE G. SHOPP

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OUTLINE

INTRODUCTION

Congenital anomalies of the gastro-intestinal tract are frequently the cause of underlying factor in obstruction. Except in some conditions such as intra-abdominal herniae, duplications, and Meckel's diverticula, a larger percentage of signs and symptoms are present in the newborn than any other period of life.

I. CONGENITAL ANOMALIES OF THE ESOPHAGUS

Embryology - Etiology

The embryonic digestive tube consists of an internal tube of entoderm which becomes the epithelial lining and an investing layer of mesoderm that forms connective tissue, muscle, and serosa.

In the 4mm. embryo the esophagus is a laterally flattened epithelial tube having a well-defined dorso-ventral cleft as a lumen. In older embryos the size of the lumen becomes not only relatively, but actually lessened in size. By the time the 20mm. stage is reached there are numerous "vacuoles" in the esophageal epithelium so that it appears to have several lumina noted by Schultze (1) in 1897. The presence of these vacuoles has been demonstrated by others (Kreuter 1905) who also concluded that the lumen became temporarily occluded at about this stage, but Forssner (1907), Schridde, (1) (1908) and others have failed to find any evidence that the lumen is ever occluded. However, Jordan and Kindred (2) show sections in their text of the occluded lumen, but

Arey (3) states it is never occluded as in the lower vertebrates. At six weeks there are two distinct layers of epithelial cells which at birth has increased to ten layers containing some ciliated cells. Superficial glands begin to develop in the third to fourth month, the deeper ones much later.

At lo mm. the circular muscle coat is present as a concentric layer of myoblasts. The longitudinal muscle is indicated at 30-42 mm. and at 55 mm. is a definite layer.

Keith (4) attributes the formation of congenital anomalies of the esophagus to a failure of the vacuoles to coalesce so that continuity of the lumen is not re-established after this stage. Bremer also supports this theory.

An arrest in development or failure of the mosoderm to separate completely the trachea from the esophagus results in formation of a tracheo-esophageal fistula.

Historical Note:

The first description of atresia of the esophagus has been attributed to Durston, (1670) and in 1821 Martin (6) published the first case report. Gibson observed such a case in 1696 and reported it in 1703 (7). In 1931 Rosenthal (8) reported 255 cases, in 1933 O'Hare (9) reported 281 cases and by 1941 Ashley (10) collected 314 cases. In 1944 Ladd (6) added Seventy-two cases from the Boston Children's Hospital making a total of 400 and N.L. Leven (11) added forty-one cases in 1945.

The first successfully treated patient was operated on by N. Logan Leven of Minnesota in 1939 and Ladd of Boston reported a successfully handled by direct end-to-end anastomosis is that of Cameron Haight of the University of Michigan, March 15, 1941. Since these cases there have been numerous other successful results at anastomosis both by the direct primary anastomosis as well as the multiple stage procedures.

Congenital Anomalies

Classification according to Vogt (12):

Type I. Complete absence of esophagus.

Type II. Atresia of the esophagus with an upper and lower esophageal segment, each ending in a blind pouch.

Type III. Atresia of the esophagus with tracheo-esophageal fistula.

a. With fistula between upper segment and trachea.

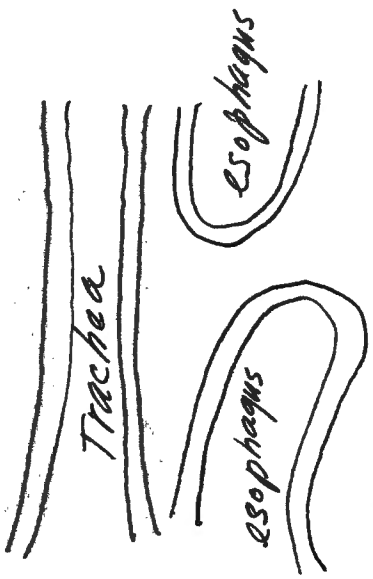
b. With fistula between lower segment and trachea.

c. With fistula between both segments and trachea.

Holt, Haight and Hodges would add one more type to this--the form of a single tracheo-esophageal fistula without esophageal atresia. R. P. O'Bannon (13) reported the first case of congenital partial atresia with a congenital diverticulum.

Congenital anomalies of esophagus are of the above types or with minor variation; and incidence has been estimated at 1:50,000.

Esophageal duplications occur which are cystic structures of variable size with thick walls and mucous membrane lining. They arise in the posterior mediastinum and expand into the thorax retro-pleurally. The mucosa resembles



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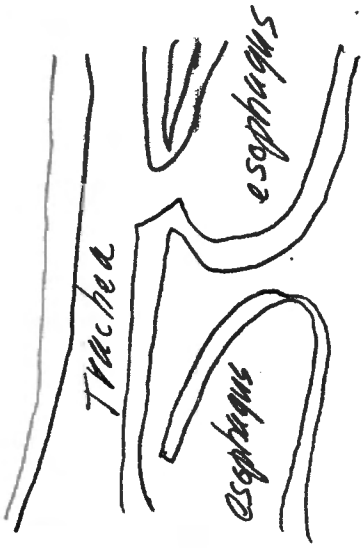
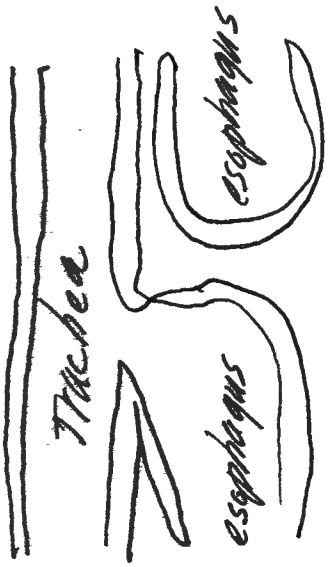
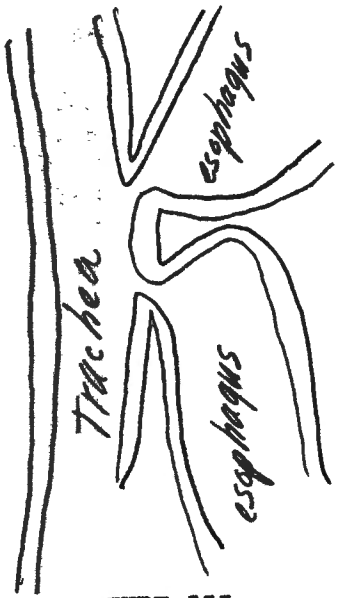


TYPE IIIa.

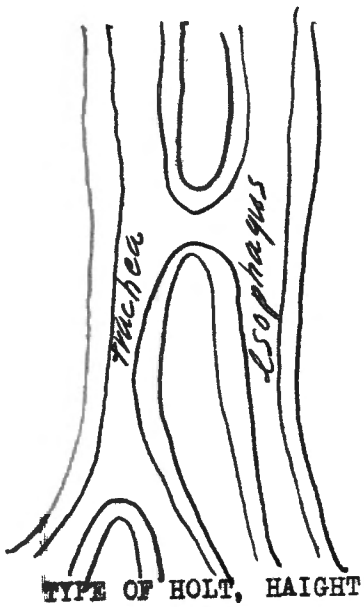


TYPE IIIb.

TYPE I. Complete absence of esophagus.
TYPE II. Atresia with upper and lower blind pouch.
TYPE IIIa. Atresia with fistula between upper segment and trachea.
TYPE IIIb. Atresia with fistula between lower segment and trachea.



TYPE IIIc.



TYPE OF HOLT, HAIGHT AND HODGES

O'BANNON'S CASE

TYPE IIIc. Atresia with fistula between both segments and trachea.
TYPE OF HOLT, HAIGHT AND HODGES - single fistula without atresia.
O'BANNON'S CASE - partial esophageal atresia with diverticulum.

that of the esophagus and may contain gastric mucosa which may secrete, ulcerate, and hemorrhage. Occasionally they are intimately connected with the muscularture of the esophagus but do not communicate with it. They cause symptoms due to pulmonary, tracheal and esophageal pressure.

The condition of the congenital short esophagus with thoracic stomach causes symptoms most often of pain and dysphagia (14). Vomiting may occur but the picture of obstruction is seldom found.

Diagnosis:

This condition demands early diagnosis and treatment. Congenital anomalies of the esophagus in almost all instances include an atresia so that it is impossible for the infant to ingest nourishment hence the conditions are incompatible with life and the signs and symptoms are evident in the early newborn period. Excessive mucous and salivation are usually present with regurgitation of food with choking and dyspnea. Cyanosis develops due to aspirated food or bronchial obstruction due to aspirated mucous. Feedings are vomited almost immediately.

Roentgenographic examination of lungs should be made first: these also are valuable preoperatively. A soft No. 8F or 10F catheter may be introduced to see if there is obstruction. This should be done under the fluroscope to be certain of the catheter's position. Through this catheter a small amount, .5-1.cc., of iodized oil may be introduced to ascertain the type of anomaly. Flat films should also be taken to determine if there is any air in the stomach. With types II and IIIa, there will be no air in

stomach or intestines. In type IIIb, there will be air in the stomach but no iodized oil enters the trachea, while in IIIc, there will be iodized oil in the trachea as well as air in the gastro-intestinal tract. As much of the oil should be removed as possible by suction. With iodized oil in the esophagus the infant should be kept in prone position and excursions of the esophagus with breathing noted under fluoroscopy, since roentgenographic study may show it to be much shorter than it actually is and the thought of primary anastomosis abandoned when it could be accomplished. Air in the stomach and intestines with a proven blind upper esophageal pouch indicates a trachea-esophageal fistula. When a fistula is present the stomach is usually distended and abdomen tympanitic. Barium is inadvisable to use as a contrast medium as it may lead to serious pulmonary complications if it gains entrance into the bronchi. On preliminary chest films the trachea is often compressed in an anterior-posterior plane by the dilated upper esophageal segment.

Whenever congenital abnormalities occur it must be kept in mind that there may be others and they should be sought for. In Ladd's (15) series of eighty-two patients there were ninety-one malformations. Of these, eighteen were of sufficient seriousness to alter the care of the patient or to cause death. Most of the associated anomalies involved the intestines or the heart and aorta.

Treatment:

There are two fundamental problems to combat in these patients,

namely, prevention of aspiration pneumonia and starvation which are the two main causes of death.

After the diagnosis has been made and the type of anomaly determined insofar as possible, an evaluation of the pulmonary status should be made and measures such as aspiration, ozygen administration, and chemo-therapy instituted if necessary. The patient should be further prepared for surgery as soon as possible, by correction of fluid balance, administration of plasma or blood, and vitamins C and K.

Up until 1939 there were no successful operated cases with these anomalies. Most men considered the condition inoperable. F.A. Trump (16) kept a patient living for thirty-seven days by administration of glucose intraperitoneally. Leven has the oldest living patient and Ladd of Boston a patient twenty-four hours younger. These were operated upon in 1939 and had preliminary gastrostomy, followed by extrapleural ligation of the fistula and esophagostomy. Haight (13A) of Michigan had the first successful case of primary end-to-end anastomosis in 1941.

Early attempts at surgery consisted of a number of different procedures. With only atresia gastrostomy should be sufficient for feeding purposes (17). This was first done by Charles Steele (18) in 1899. In cases where a tracheo-esophageal fistula existed gastrostomy was not sufficient as it did not prevent regurgitation into the trachea. Gastrostomy plus ligation at the cardia did not

prevent aspiration via the upper segment. Dorsal esophagostomy advocated by Mixter, technically seemed to solve the problems except that of a mediastinitis and pleuritis so it was soon abandoned. Gage and Ochsner (19) tied off the cardia, marsupialized the upper esophageal segment and did a gastrostomy. Objections to this plan are that the long esophageal pouch led to aspiration and infection later and that the cardiac end sloughed with regurgitation and aspiration.

N. Carter (20) brought the lower end of the esophagus down through the diaphragm and exteriorized it but this left a long lower esophageal pouch.

Direct attack on the fistula was suggested by Keith (21) in 1910. Shaw (22) reported the first operation of this type in 1939 in which article he mentions that Samson used the same technic previously. Lanman (23) had also used this technic in 1936 and 1937.

Haight was the first to use successfully the direct attack of primary end-to-end anastomosis in 1941. He used a left paravetebral incision but others now use a right paravertebral incision feeling that this gives better exposure. Details of technic are beyond the scope of this paper. However, the fistula is ligated extrapleurally and primary end-to-end anastomosis of the esophageal segments is accomplished. Continuous upper pouch suction is maintained and in from twenty-four to forty-eight hours a gastrostomy is performed for feeding. If there is no leakage at the

anastomosis oral feedings are begun about the twelfth day.

When primary anastomosis is not possible due to wide separation of the segments, a multiple stage procedure must be resorted to. The fistula is closed as in the primary anastomosis procedure by cutting and tying. Constant suction is maintained in the upper segment. A gastrostomy is done in the next two or three days. Next the upper end of the esophagus is exteriorized at the base of the neck. At some later date the anterior thoracic esophagus can be constructed. An epithelial lined tube is constructed over which a tube grat is sutured forming the subcutaneous esophagus.

II. CONGENITAL ANOMALIES OF THE STOMACH

Embryology:

In the 4 mm. embryo the stomach is a spindle-shaped enlargement of the foregut. At the 10 mm. stage the gastric epithelium is much thicker than that of the esophagus and by 16-19 mm. it shows vacuoles and pits as in the esophagus. At this same period the organ grows in length and the dorsal wall grows faster than the ventral producing a greater and lesser curvature. The dorsal mesentery also expands more rapidly than the ventral. These factors aid in rotation of the stomach 90° explaining the positions of the vagus nerves. The enlarging liver pushes the cephaled portion of the stomach to the left.

At seven weeks mucosal pits appear which show gastric gland buds at their bases at fourteen weeks.

The circular muscle is distinct in the mesenchyme at 16 mm. and at 24.0 mm. the longitudinal muscle is indicated.

Congenital Pyloric Stenosis - Etiology and Pathology

Congenital pyloric stenosis is the most common condition that requires surgery in the newborn period. The etiological factors of this disease are not known. There appears to be an hereditary influence as it often occurs in two or more in a family. Males are more often affected than females in a ratio of 4:1.

The condition is caused by hypertrophy of the circular pyloric musculature which increases in the number of muscle fibers present into a smooth mass approximately 2 by 2 cm's. This muscle compresses the underlying mucosa resulting in a stenosis of the lumen of the intestine. Whether the hypertrophy occurs before birth is not known. Some attribute the condition to a mechanism of work hypertrophy and others to a pylorospasm plus work hypertrophy.

Historical Note:

Scattered accounts of cases apparently of congenital pyloric stenosis were published in the 17th century (24). The case of Hildnus, a famous German pediatrician, was probably the first in 1646. Blair discussed a case in 1717; C. Weber in 1758. Armstrong in 1777 described these post-mortem findings:

> "There was no morbid appearance to be observed anywhere but in the stomach and the viscus being so full while the intestines were almost empty, it looked as if the disease had been chiefly owing to a spasm in the pylorus which prevented the contents of the stomach from passing into the duodenum."

The first case of pyloric stenosis reported in this country was by Hezekiah Beardley in 1788, "Case of Scirrhus in Pylorus of an Infant", in the first Medical Periodical in the United States.

Diagnosis:

Congenital occlusive lesions of the stomach and the small intestine cause signs and symptoms of high intestinal obstruction. The signs and symptoms of pyloric stenosis usually appear from the third to eighth week of life and most from the fourth to the sixth week (25). The child begins to vomit shortly after its feeding and progresses from regurgitation to projectile vomiting. The vomitus is not bile strained and this is an important point in differentiating this condition from other causes of obstruction. Distention is confined to the epigastrium in the main and x-ray reveals a distended usually dilated stomach. The child obviously remains hungry and hence will nurse even shortly after vomiting. The stools decrease, the child becomes dehydrated and loses weight rapidly.

Examination of the abdomen may in many cases reveal peristaltic waves passing from left to right if the infant has been recently fed. Palpation of the right upper quadrant will in some cases reveal the olive-shaped tumor. The child may be sedated with chloral hydrate by rectum in order to facilitate this examination.

Vomiting from this cause has to be differentiated from that due to intracranial lesions, in which case it occurs more irregularly and there is no relation to food. Intracranial lesions may cause

convulsions and there may be bulging fontanels and bloody cerebrospinal fluid under increased pressure. Physical examination reveals no signs of obstruction and no palpable tumor. The feeding regimen must be evaluated so as to rule out this as a cause of the vomiting.

Pylorospasm as distinguished from hypertrophic stenosis responds to atropine with sedatives, there is no tumor palpable, vomiting is variable, and the child is apt to display a generalized hypertonicity.

Barium mixed with the formula may be given where the diagnosis is obscure. X-ray will reveal an enlarged stomach with a rounded pyloric end, increased peristaltic activity, and no appreciable passage of barium into the small intestine.

Miller and ostrum (26) were enthusiastic about their method of differential diagnosis in giving water-barium mixture through a nipple and changing the infant's position to get the barium into the pyloric antrum. Antispasmodics are given and serial roentgenograms are taken. Cases studied by their technic whom they found would most likely require surgery were those in which a slightly enlarged atonic stomach was found with a narrow pyloric canal elongated more than .5 cm. with more than fifty percent gastric retention at the end of four hours.

Early operations for this condition were gastrojejunostomy with a morality of over fifty percent. Fredet in 1908 introduced his classic operation of splitting the pyloric musculature longitudinally

down to the mucosa and resuturing it transversely. Rammstedt in 1912 did essentially the same but did not suture the muscle. The mucosa pouts into the defect. This technic relieves the obstruction immediately and in almost every case permanently. Choice of anesthetic by most men today is drop ether.

The operation is not an emergency measure but should be postponed only long enough to prepare the patient. Intravenous administration of ten percent glucose in distilled water, 10 cc. per pound, twice daily, followed by subcutaneous normal saline solution, 15 cc. per pound of body weight, is advised by Ladd and Gross (25). Occasionally in a poor risk patient blood transfusion will be necessary. Small oral feedings may be continued, as often a small amount will pass the pylorus. Vitamin C to promote healing is desirable.

Postoperatively feedings are frequent and small, beginning with one ounce of water every two hours for four or five feedings and one-half strength formula for four to five feedings after which the amount and formula strength should be gradually increased to full amount. Parenteral fluids should supplement oral feedings.

Medical treatment for pyloric stenosis is advocated by Todd (27) (London 1947) with selection of cases for surgery. He reported 112 cases treated medically of which twelve died, a mortality of ten percent. Five of the 112 patients were subjected to Rammstedts operation after medical management including eumydrin proved ineffective. He stressed that these patients should be treated in single

cubicles to prevent infections, that breast fed infants did better than bottle fed, and that a thorough routine should be worked out and experienced nursing care available. The criteria used to determine whether medical or surgical care should be used were as follows:

1. Medical treatment should be the routine treatment in infants not severely dehydrated.
2. If after a trial of seven days medical treatment is ineffective, Rammstedt's operation should be done at once unless the infant has infection.
3. Obvious infection is a contra-indication to surgery.
4. Gross dehydration is indication for surgical treatment.
5. Surgical treatment should be employed in babies whose birth weight was below 6-1/2 lbs.

Surgical treatment, at present in experienced hands, has a lower mortality than that of Todd, the hospital stay is much shorter and the morbidity less. There are a number of surgical series published in which the mortality is one and two percent.

Hypertrophic pyloric stenosis is not limited to infants alone, more than 100 cases having been reported in adults (28). The majority of these are associated with peptic ulcer or gastritis.

Theories as to etiology are several. One is that it is a

continuance into adult life of congenital hypertrophic stenosis of the newborn. Others contend that is is due to hypertrophy in adult hood superimposed on a congenital background. Pylorospasm caused by intragastric disease, cholecystitis, or appendicitis also has been thought to be the explanation. Kirklin and Harris (29) describe the most important roentgenologic findings as elongation of the pyloric canal to 2-4 cms. and a crescentic indentation of the bulbar base.

Signs and symptoms are those of partial to complete pyloric obstruction and treatment of choice is gastric resection (30).

Congential anomalies of the stomach other than hypertrophic pyloric stenosis are very rare. Pfaundler, Schlossmann (31) state they are so uncommon as to have no significance. Anders is reported by Touroff (32) as stating only two cases of congenital occlusion of the stomach have been reported, one a membranous obstruction at the pylorus with pyloric hypertrophy, and the other, a growth of accessory pancreas imbedded in the wall of the stomach.

Touroff reported a case of non-hypertrophic pyloric stenosis had been described. Kreuter was able to collect only four cases of pyloric atresia from the literature. Bonner (33) described one case of membranous obstruction of the pylorus with pyloric hypertrophy. Theories as to etiology are several. One is that the septum is formed by a fusion of folds of mucous membrane in this

area. Another is that is is an abnormally high occlusion of the duodenum, or that it represents a failure of the vacuoles to coalesce as in the esophagus and intestine.

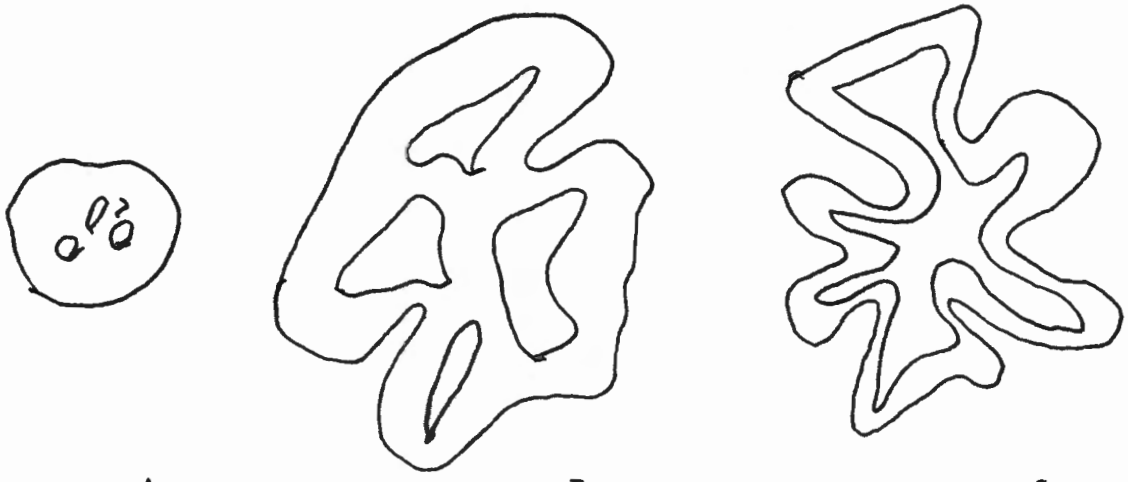
III. ATRESIA OF THE SMALL AND LARGE INTESTINE

Embryology

In the 5 mm. embryo the intestine is a single tube ending in a cloaca, the only recognizable division being the duodenum. The intestine is described in two parts, a cephaled and a caudad limb of the intestinal loop. From 5 mm. to 9 mm. the ventral flexing of the intestinal loop increases and a bulging marks the beginning of the cecum. It now begins to rotate about the superior mesenteric artery as an axis and is thrown into a loop because it grows faster than the abdominal cavity. Some attribute this to internal spiral growth of the gut while others attribute it to the enlarging left umbilical vein dragging the gut with it. The cephaled portion in this process of rotation becomes caudad to the caudal limb by an anti-clockwise rotation. The intestine now grows in length so rapidly that the abdominal cavity cannot contain it and at seven weeks herniates into the imbilical cord. At about ten weeks the abdominal cavity has enlarged sufficiently to contain the intestine and so it returns rather suddenly by either traction by non-herniated intestine, negative pressure due to decrease in liver growth, or by retraction of slower growing mesentery.

The cecum is last to leave the umbilical cord and as it does so has a tendency to straighten which carries it to the crest of ileum on the right where it becomes fixed, and the colon now extends obliquely

from this point to the spenic flexure. As the liver "recedes" in relative size it carries the colon with it and the hepatic flexure is formed. The vermiform process is formed by a rapidly elongating process of the distal tip of the cecum.



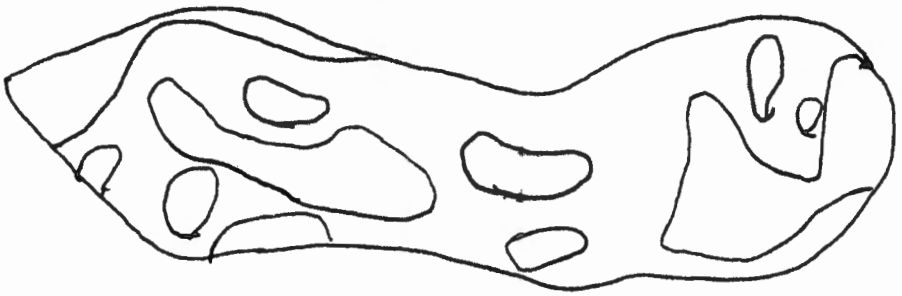
A. B. C.

Cross sections of the duodenal epithelium.

A. Concrescence of intestinal epithelium after five weeks of fetal life.

B. Showing formation of cystic spaces.

C. Intestinal lumen cleared by twelfth week.



Model of the duodenum of a 22.8 mm. embryo, seen in longitudinal section.

At the 6-7 mm. stage the duodenum has a well defined single lumen. The epithelium begins to proliferate rapidly at 10 mm. and vacuoles are formed, and later septa, so that between the 23 mm. and 30 mm. stage there is no continuous lumen. After this time the vacuoles begin to become confluent to re-establish the original central continuous lumen. The lower portion of the small intestine and the colon show these changes also but to a lesser degree. Villi appear at eight weeks as rounded elevations of the epithelium and intestinal glands arise as tubular ingrowths of the epithelium at the bases of the villi appearing at the end of the third month. The duodenal glands (of Brunner) develop shortly after the glands of Lieberkuhn and lymph nodes and Peyer's Patches are present at five months.

The circular muscle can be seen in the duodenum at the 10 mm. stage and gradually appears at lower levels. The longitudinal muscle appears at 75 mm. Mall (1897 and 1898) inferred that peristalsis occurs in 130 mm. embryos since in several he found meconium had been propelled downward toward the cecum.

The large intestine at the 10 mm. stage consists of an epithelial tube, a layer of undifferentiated mesenchyme, and a layer of peritoneal epithelium. Obliteration of the lumen of the large intestine as in the small intestine was described by Kreuter (1) (1905) to begin about the fifth week but was not found by Forssner (1) (1907). Arey says the process occurs as in the small intestine but to a lesser degree.

Villi are present at 55 mm. and glands at 99 mm. Circular muscle is in evidence at 22.9 mm. and found throughout the colon at 42 mm., the longitudinal muscle appearing at 75 mm. Transverse folds in the rectum are seen at 120 mm. and appendices epiploicae at four months.

The appendix is an epithelial tube at 16 mm. surrounded by undifferentiated mesenchyme.

Etiology

The occurrence of congenital intestinal atresia and stenosis has been estimated to be 1:20,000. The first case reported in the literature was by Calder of Edinburgh in 1733. Since then over 500 cases have been reported.

The theories advanced to explain these occurrences have been many. We have mentioned above that before the 5th week of fetal life the intestine has a well defined lumen. From the fifth to twelfth week, however, the lumen becomes obliterated by concrescences and then it is re-established. Any condition that will cause an arrest in development of a segment during that time may cause a stenosis or an atresia or leave a membrane or veil occluding the lumen.

The classification of Forssner and Spriggs (34) (quoted by Bryan)of intrinsic atresia and stenosis of the intestine is as follows:

A. Simple narrowing, stenosis

B. Perforated diaphragm, stenosis

C. Complete diaphragm, atresia

D. Short band, connecting the ends of bowel, atresia

E. Thread-like band along free edge of mesentery, an atresia

F. Gap in intestine and mesentery, an aplasia

Various conditions occurring during the critical period which may be the underlying cause of this anomaly have been listed as inflammation, pressure by the head of the pancreas, vascular changes, syphillis, kinking of the bowel, fetal intussusception, strangulation in a mesenteric opening, volvulus, and extrinsic pressure (35).

The greater majority of atresias and stenoses occur in the ileum, in the duodenum and more rarely in the jejunum.

Diagnosis and treatment

The signs and symptoms of intestinal atresia appear in the first day or two of the newborn period. Infants fed on the normal amount of formula vomit soon afterwards. The vomitus in contrast with that in pyloric stenosis almost always contains bile - an important differential point. Abdominal distention may be absent if the atresia is high or limited to the epigastrium. Dehydration is usually marked.

Examination of the stools if atresia is suspected is of value according to the method of Farber (36). If there are no epithelial cells present in a smear of the center of the meconium, the diagnosis of atresia is substantiated because epithelial cells are swallowed in the amniotic fluid.

Usually barium studies are not necessary to establish the diagnosis, but if they are used, a thin mixture should be given and as much as possible removed from the stomach afterward to minimize pulmonary aspiration when vomiting occurs. Barium enema is of no value and only adds to the distention. Pyloric stenosis seldom causes signs and symptoms before the third or fourth week, whereas those of atresia or stenosis usually appear in the first several days of life.

The first thorough paper written on this subject was by Theremin in 1877, and the first case to be operated upon was that of Tischendorf (1887), who performed an enterostomy. Sutton in 1889 had a similar case.

Until 1911 there had been no cases treated successfully, but in that year Fochens of Rotterdam reported a case of atresia of the ileum in which he did a side-to-side anastomosis and the child recovered. The first case of duodenal atresia treated successufl1y was that of Ernst (37) of Copenhagen in 1916 treated by the same method.

Atresia of the colon is met with less frequently than atresia of the small intestine but it may occur in the same forms. The first case of this type to recover was that of Potts (38) in 1947 in which there was atresia of the transverse colon, in which he performed a side to side anastomosis.

Successful treatment is as dependent upon early recognition of atresia as upon the correct surgical care. Surgery should be attempted promptly after optimum preoperative care. If the atresia is high

in the duodenum and no bile is present in the vomitus, gastro-enterostomy will be required. With a lower atresia however, side-to-side anastomosis is the operation of choice. For atresia of the ileum, many surgeons have performed ileostomy, but this operation has been unsuccessful except in one case, a Chinese infant, in whom ileostomy alone was done by Rudd (39) in 1946 for an atresia of the ileum three inches from the ileocecal valve. The first attempt to close the ileostomy failed but the second succeeded approximately three months following the initial operation.

Brunuzzi and Lyons (40) report a case of Martin's (1945) treated by ileostomy with crushing of the spur and later closure, and a case of their own (1946)with ileostomy followed by side-to-side anastomosis on the fourth day after the original operation when ileal drainage first appeared. Performed before loss of too much succus entericus, this method may offer some encouragement but will need further evaluation.

There is tremendous loss of fluids and it is very difficult to maintain nutrition and electrolyte balance in these patients with ileostomy who are already bad risks. For this reason ileostomy is ordinarily a poor procedure. It is extremely important to explore the intestinal tract at surgery, for atresias are often multiple. In Davis and Poynter's review fifteen percent of the cases had multiple anomalies of atresia or stenosis.

Anastomoses are made by the open method. The proximal bowel may be needled for decompression and the distal segment enlarge slightly

by injecting air or fluid. Postoperatively Ladd and Gross dilate by injecting saline solution from below.

IV. STENOSIS OF THE SMALL AND LARGE INTESTINE

The condition of intestinal stenosis arises in much the same manner as atresia. A partial diaphragm may obstruct the lumen or there may be an incomplete transition from the solid stage to the re-establishment of the lumen. Apparently at least some of the cases of atresia described in the literature have actually been cases of stenosis as the preoperative findings are very similar, and often at operation it is difficult to differentiate the two. Microscopic section of the stenosed area shows areas of heaped-up mucosa and submucosal tissues.

The symptoms of intestinal stenosis are difficult to distinguish from those of atresia. In about half the cases they appear in the first week of life and consist of persistent vomiting, distention, loss of weight, diminished number and size of stools. Cases that appear later have recurrent episodes of the above symptoms and often are handled as "feeding problems". The oldest patient in the series of Ladd and Gross was nines years. Nagel (41) reports a case of a seventy-two year old man who at autopsy had a stenosis of the duodenum and had symptoms of intermittent obstruction since childhood. Findings necessarily will vary with the degree and location of the stenosis, distention will be epigastric if the stenosis is high and generalized if ileal. X-ray will show corresponding bowel dilation. The duodenum may even be palpted if it has not been deflated by vomiting. Administration of barium is usually not necessary but it

done, small amounts of a thin mixture should be used.

Treatment consists of adequate preoperative preparation, followed by a side-to-side intestinal anastomosis. A right paramedian incision isused in infants in older patients the type of incision is dependent upon the location of the stenotic segment. Mechanical dilatation of the stenotic area has been unsatisfactory. An opening made in the intestine just below the stenotic area may be difficult to close because of the small caliber of the bowel. An opening above the stenosis may leak after closure if the dilation has not been entirely successful. Furthermore, recurrence of stenosis would be expected to be frequent after simple dilatation.

Postoperative treatment includes administration of fluids, blood transfusion and gastric suction. Feedings are withheld four or five days.

The prognosis in this anomaly is not favorable. The higher the stenosis or atresia the better the prognosis according to Ladd and Gross. In their series of seventy-four cases of stenosis and atresia operated upon, only seventeen recovered.

	Operated	Recovered
Atresia	52	7
Stenosis	22	10

V. CONGENITAL ANOMALIES OF THE PANCREAS

Two rare congenital conditions in the pancreas may lead to intestinal obstruction. These are annular pancreas; and meconium ileus secondary to pancreatic achylia due to obstruction of the ducts.

The human pancreas arises as two entodermal outgrowths, the dorsal and ventral anlages. The dorsal anlage grows from the dorsal wall of the intestine and the ventral from the inferior anlage formed by the hepatic diverticulum and the intestine. As the stomach rotates the two approach one another and unite. The dorsal part forms the part of the head, the body, and the tail; the ventral the uncinate process and part of the head.

Lecco (42) advanced the explanation that the failure of the ventral anlage to migrate while the duct remains in its usual anatomical relationship results in a strip of pancreatic tissue encircling the duodenum. Tieken (cited by McNaught) (42) suggested that the two anlage, develop- separately, and failing to unite as they normally do, may develop on either side of the duodenum thus encircling it. The semi-annular type he thought was due to over-development of the head which may send out processes which partially surround the bowel.

The diagnosis of annular pancreas is difficult to make preoperatively. Lehman (43) reported the first case diagnosed preoperatively in 1942. Annular pancreas may become manifest at any age. McNaught (44) reported a case in a thirty-seven year old woman and a seventy year old male, and gathered thirty-nine cases from the literature up to 1933, several of which were seventy to seventy-four years of age. Approximately fifty cases have been reported and only eleven had been operated upon by 1944.

The symptoms are those of acute or recurrent duodenal obstruction, namely epigastric pain and distention, nausea, vomiting and malnutrition.

and dehydration. The presence of bile in the vomitus will depend upon the relation of the ring to the ampula of Vater but in most cases there is no interference with the flow of bile (45).

Barium studies may show a constricting lesion having the appearance of a smooth circle of the descending portion of the duodenum.

It is possible for such an anomaly to be asymptomatic and come to light as an acute pancreatitis, at which time the glandular enlargement may cause acute obstruction of the duodenum. In several instances the annular tissue has completely closed off the common bile duct in infants causing death.

Until 1944 only eleven cases had been reported operated upon (46), six of these successfully. Three of four patients upon whom a direct attack on the pancreatic ring was made developed pancreatic fistulae. One of these had to be closed by operation but all survived. The likelihood of a pancreatic fistula following this operation makes it an unsatisfactory one. Posterior gastroenterostomy has been done but unfortunately three of five patients thus treated died of other causes. If the presence of the annular pancreatic tissue occludes the lumen of the duodenum or if the duodenum is congenitally atretic or very markedly stenosed, it is feasible that this operation may not adequately drain the duodenum.

Duodenojejunostomy had been recommended by Howard, Zech (47), and Lehman, and first used by Gross and Chisholm, being advocated by them as the procedure of choice.

Another anomaly which deserves mention occurs rarely and indirectly causes intestinal obstruction is stenosis or atresia of the pancreatic ducts. This condition is pancreatic fibrosis and inspissation of meconium in the intestine due to absence of the pancreatic ferments. The association of these conditions was first reported by Landsteiner in 1905 (48) Kaufmann and Chamberlain (49) believe that maldevelopment, (possible congenital malformation) of the pancreas may be the underlying cause in some cases, since in their patient no imflammation or obstruction was found but rather maldevelopment of the intrapancreatic ducts, ductules and acini.

The normal consistency of meconium composed of bile, mucus, secretions, fat globules, cholesteral, vernix caseosa, epithelial cells and hair is dependent upon the presence of pancreatic secretion and bile in the small intestine. In the absence of these, fats and proteins are not digested and inspissation and hardening occurs. Normal intestinal peristalsis cannot move this tenacious meconium and obstruction results. Dilatation and performation of the bowel may occur above such accumulations with meconium peritonitis. Where meconium peritonitis has occured in utero, x-ray of the abdomen will reveal the presence of calcified plaques in the peritoneal cavity (50). These represent cornified epithelial cells which become calcified in the peritoneal cavity.

In six cases reported by Dodd (51) in which thorough post mortem examinations were made, there were found abnormalities in either the liver or the pancreas. Studies by Hurwitt and Arnheim (52) and others

have failed to demonstrate that the ducts of the pancreas pass through a stage of occlusion by epithelial proliferation as in the biliary tract and duodenum. Their conclusion was that the point of obstruction represents a congenital focal developmental defect.

In most of these patients the diagnosis of intestinal obstruction due to stenosis, atresia or other congenital anomalies has been made preoperatively. The correct diagnosis has been made at the operating table at which time ileostomy or colostomy has been done. This procedure has little to offer and this disease has been universally fatal.

Farber (53) has shown that pancreatin added to the meconium renders it of liquid consistency in a short time. He has also advocated treatment on this basis by irrigation of the ileostomy or colostomy. Four infants were treated by this method at Boston Childrens Hospital. In three of the cases the intestinal tract was cleared of the meconium. Two of these died later of upper respirator infection and one was still living and well when last reported.

IV. DUPLICATIONS OF THE GASTRO-INTESTINAL TRACT

Embryology, Pathology, Etiology

Duplications of the gastro-intestinal tract have also been called enteric cyst, giant diverticula, and inclusion cysts. They are not cysts but duplications of some part of the tract. Most of them have no communication with the bowel. The structure is a two layer muscular coat lined with mucosa and the muscular coat is intimately a

part of the tract to which it is adherent. The mucosa may be like that found in any part of the tract no matter which part the duplication is attached. The fluid is mucoid and clear except where it has become so excessive the pressure has caused necrosis and hemorrhage. Mesenteric cysts must be differentiated, they are of lymphatic origin, are not adherent to the intestine, and are thin walled.

Lewis and Thyng (54) have advanced the most plausible theory as the etiology of duplications. They often found numerous diverticula of the early fetal ileum of animals and humans and postulated that the detachment or pinching off of one of these could readily explain the formation of a duplication -- theory of developmental arrest.

Another theory, Hughes-Jones (55) is that of epithelial sequestration in which an islet of epithelium becomes detached before the formation of the circular muscle begins, then grows to form a cyst, the muscle coats of which are derived as in the intestine.

Meckel, quoted by Evans (56) considered there were two types of diverticula true and false. Those arising from the vitello-intestinal duct he termed the true diverticula, whereas the ones in other parts of the intestinal tract were called false. The true diverticula has walls that repeated those of the intestine but the false were formed by "solutions of continuity" and hence their walls were not

the same as that of the intestine. Due to this classification there was a tendency to call all duplications whose walls and structure were like those of some part of the gastro-intestinal tract true diverticula and to assume that they originated in the vitello-intestinal tract. Much support in the literature for the theory of nearly all duplications arising from the vitello-intestinal tract developed as a result of this classification of Meckel. On the basis of the theories first mentioned above however, there is now more general agreement that Meckel's abnormality (true diverticulum of Meckel) is a distinct congenital anomaly arising from incomplete obliteration of the vitello-intestinal tract and that the other duplications which often coexist originate as described above by Lewis and Thyng.

The occurrence of mucosa of another type in the duplication is accounted for by a process of de-differentiation and re-differentiation under the stimulus of intra-cystic pressure or inflammation (57). The lumen may communicate with that of the intestine but more often does not.

Duplications may occur at any place from the base of the tongue to the rectum but are most frequent in the ileum and more so in the segment of the last few inches of terminal ileum and the first few inches of ascending colon.

In fifty-five cases:

Duodenum		3
Jejunum		4
Ileum except last 4"		16
Last 4" of ileum	)	
Ileocecal valve	)	
Ileocolic angle	)	31
Cecum	)	
First 2" of ascending colon	)	
Transverse colon		1

Other series of fewer cases would seem to have a like distribution. The majority are submucous but may be intermuscular, subperitoneal or mesenteric in position. Cysts may arise in the mesentery of the small or large bowel but are unlike duplications in that they are of lymphatic trunk or misplaced lymphatic tissue which does not communicate with existing systems. Only infrequently do these cause obstruction when they are at the free border of the mesentery, causing pressure on the intestine. More often the findings are pain and gradual enlargement of the abdomen.

Diagnosis

Age incidence in duplications depend in a measure upon the location of the cyst and may cause symptoms at any age, most of them however, in the first year of life. In Hughes-Jones series, cysts of the terminal ileum were usually discovered in the first year of life causing intestinal obstruction, whereas most of those arising in the cecum appear in later years. Cogswell and Thompson reported a duplication of the rectum causing symptoms at two weeks.

The most usual disturbances caused by duplications are those of intestinal obstruction by encroachment on the lumen of the bowel.

The signs and symptoms will necessarily be dependent upon the level of the lesion causing the obstruction. Occasionally pain is the only major finding and may persist intermittently for years, caused by distention of the "cyst" with secretions from its lining. The lining may secrete enough fluid to cause pain from pressure of distention, or the distended cyst may , by pressure on mesenteric vessels, cause gangrene of the mesentery or bowel. In some cases the complaints may be due to abdominal distention and fullness caused by a rapidly enlarging "cyst". Most of the duplications of the small bowel cause partial obstruction with vomiting, colicky pain, and visible peristalsis. Frequently the mass is the factor initiation an intussuception. In a smaller percentage of cases the mass is the cause of a volvulus. The mass is often palpated as movable, rounded and firm. Barium studies in intermittent partial obstruction often will reveal dilated bowel above the lesion and displace loops of bowel, as well as give an idea of the level of the lesion. Severe intestinal hemorrhage may in some cases be the first sign (58).

Complete duplication of the intestine is rare. Gray (1940) found eleven cases of duplication of the colon in the literature and reported the only case of triplication of the colon in the

literature. The structures were complete except that a layer of longitudinal muscle and serosa enclosed all three as a common tunic. Griess (59) et al, (1947) reported a case of complete duplication of the large bowel in a nineteen year old female. Roentgenologic diagnosis was made. There was absence of the right kidney and two uteri were present. There was no communcation between the two. A colostomy of the descending colon was done and later closed to establish a communication. The patient had been operated previously and two appendices removed, and operated twice for bowel obstruction at one of which a cyst was reported drained.

Allard (60) et al recently reported a case of complete duplication of the large bowel in which the terminal ileum was also duplicated and a rectourethral fistula was present. A preliminary colostomy was done and the fistula ligated. Later an ileosigmoidostomy was done to the functioning lumen followed by a subtotal colectomy. The double rectum was converted into one chamber by crushing the spur and later the colostomy was closed.

Occurrence of other anomalies is very frequent with complete duplications. The case of triplication had numerous anomalies (62).

Cogswell and Thompson (61) reported a case of duplicatiom of the rectum which under observation presented the findings of a cyst due to a ball-valve-like communication with the rectum, and became infected, responding to marsupialization.

The treatment for enterogenous cysts is surgical removal. In.

nearly all instances the "duplication" is intimately connected with the bowel and cannot be dissected and enucleated so that segmental resection with end-to-end anastomosis will be necessary. Marsupialization of the cyst can be done but should be reserved for patients who are poor surgical risks. Simple evacuation of the cyst has been done but does not seem feasible since recurrence is almost certain to occur and spread of infection is likely.

Although not an anomaly of the gastro-intestinal tract exactly, omphalocele and umbilical herniae nearly always contain loops of intestine. Obstruction has resulted in one case (63) of clamping the small intestine, when clamping the umbilical cord a second time to stop hemorrhage from it. Peritonitis rather than obstruction occurs in omphaloccle.

Persistence of the vitelline duct as Meckel's Diverticulum or true diverticulum as described by him may be the underlying cause of obstruction at any age. The diverticulum is found within one metre of the ileo cecal valve and consists of an outpouching up to three inches in length. The lining most often consists of gastric and, or, ileal mucosa.

Loops of small intestine may be constricted by a remaining band from the umbilicus to the diverticulum causing acute intestinal bostruction. More often obstruction is due to intussusception for which the inverted diverticulum acts as the initiating factor. The picture is like that of idiopathic intussusception -- intermittent

abdominal pain with vomiting and palpable right sided mass and often this is palpable by rectal examination. Bloody stools appear in 85% of the cases. Treatment of intussusception from any cause should by surgical. It is apparent that attempts to reduce an interssusception by barium enema are frought with danger. There is no way to determine except by operation whether or not a Meckel's diverticulum is the underlying cause, which if present should be excised. Furthermore, one cannot be certain that an intussusception is completely reduced in any particular case by barium enema.

Excision of the diverticulum should be done if the patients condition is good, if not, the obstructing intussusception should be reduced, the diverticulum everted, and excision postponed until the patients condition is improved. Mortality of intussusception from all causes varies directly with the length of time elapsed from onset of symptoms to surgery. A number of series of cases -- Clubbe and Hipsley of Australia, and Ladd and Gross have had mortality rated in the neighborhood of five percent even with illness up to forty-five hours before surgery. In Ladd's series with illness of less than twenty-four hours in 110 patients, there was no deaths.

Snyder, Kraus and Chaffin (64) of Los Angeles have reported a series of 143 consecutive cases of intussusception with a mortality of 4.8%. The average duration of time from onset of symptoms to surgery in their series was twenty-eight hours. They stress the importance of early diagnosis and surgery with improved

pre and postoperative care. When the bowel was badly damaged streptomycin was injected into the lumen above the involved segment.

VII. MALROTATION OF THE INTESTINE

Etiology-Pathology

Incomplete rotation of the intestine is a condition which may cause symptoms at any period of life. In the majority of cases symptoms occur in the newborn period.

The midgut, which consists of two segments, a pre-arterial (that portion between the duodenum and the vitelline duct) and a post-arterial umbilical cord from the sixth to tenth week of embryological development. At the tenth to eleventh week the midgut is drawn back into the peritoneal cavity by a counterclockwise rotation and the terminal ileum, cecum, and ascending and transverse colon lie in the epigastrium a little to the left. Normally the rotation continues so that the cecum passes obliquely to the right and finally reaches the right lower quadrant after which the colon becomes attached in this position and the mesentery of the small intestine becomes attached to the posterior abdominal wall obliquely from the duodenojejunal junction toward the cecum. The naomalies which are caused by an arrest in this process may result in an incompletely rotated cecum, a completely rotated but unattached and mobile cecum, or a mesentery which is unattached to the posterior abdominal wall.

When the cecum fails to rotate, it is found just inferior to the lower half of the stomach with bands of reflected peritoneum

lying across the descending portion of the duodenum obstructing it, or the cecum itself may be directly over the duodenum obstructing it by extrinsic pressure. In malrotation the mesentery of the small intestine is attached to the posterior abdominal wall by only a short attachment and hence volvulus of the midgut often occurs with mechanical intestinal bostruction, with or without strangulation of the superior mesenteric vessels and gangrene of the intestine.

The age at which symptoms occur in this anomaly may vary widely but the majority appear in the first year of life. In the newborn period vomiting is usually the main complaint and the vomitus may or may not contain bile depending upon whether or not the constricting bands are above the ampulla so the vomitus will be bile stained. Abdominal distention occurs, first epigastric, but later generalized due to distention in the small intestine and colon involved in the volvulus. Stools will be scanty due to the volvulus involving the colon. When infarction of the intestine occurs, fever will rise to 103 to 104° and dehydration will be marked. In infants, barium studies are inadvisable if malrotation with volvulus is suspected, since aspiration or inspissation may occur; if used, only small amounts of a thin mixture should be given. A barium enema will show the cecum in the epigastrium. When plain films and clinical study reveal intestinal obstruction, it is better to operate without doing the barium studies. In older patients in whom there have been recurrent attacks of intestinal obstruction or abdominal pain, distention, nausea and vomit-

ing without apparent cause, barium study would not be contra-indicated and would quickly reveal a malrotation. A patient with malrotation may have such complaints for years, even for a life time and the condition only revealed at autopsy. In others, after years without symptoms, a volvulus may bring the patient to surgery abruptly.

In infants during the first week or ten days of life this condition may be indistinguishable from intestinal atresia or stenosis but the treatment is surgical in both.

The treatment in all cases of malrotation is boviously surgical where the condition is causing symptoms. Adequate pre-operative preparation with fluids, blood, and decompression should be carried out. Occasionally this anomaly is discovered on more or less routine barium studies and is asymptomatic in which case it is only necessary to watch the informed patient.

When the cecum or congenital bands lie over the duodenum the bands must be severed and the cecum allowed to retract to the left upper quadrant where it remains. When only the discolored small intestine presents itself on opening the abdomen and the ascending and transverse colon are not seen, there is probably a volvulus and the entire small intestine should be brought out in order to properly reduce it. Ladd has emphasized the importance of not only reducing the volvulus in such cases but also of looking for and severing any peritoneal bands which may be obstructing the duodenum. If the only anomolous finding is a mobile cecum, the ascending colon being normally

attached, the cecum may be secured in its normal position by interrupted sutures.

VIII. INTRA-ABDOMINAL HERNIAE

Historical Note - Etiology

This is one of the rarer causes of intestinal obstruction.

First mention of the peritoneal fossae is attributed by Moynihan (65) to Heusing in 1742.

Gruber and Moynihan (65), in 1859, 1861 and 1862 wrote on the subject and described three cases.

Waldeyer (65) (1868) explained the origin of the fossae as due to separation of the superior hemorrhoidal vein from the posterior abdominal wall as it ascends, thereby forming a peritoneal fold. Treves (65) in Hunterian Lectures attributes the fossae to remaining mesoduodenum.

In 1776 Neubauer (65) reported an "exceptionally rare case of a peritoneal hernial sac holding all the small intestines". Following this date other scattered reports were made of similar findings but the true nature of this anomaly was not realized until the work of Treitz in 1857 who described the paraduodenal fossae and theorized on the etiology of intra-abdominal hernia. He expressed the opinion that the fossae were fromed due to failure of fusion of the peritoneal leaves and that they were deepened by pressure and peristaltic activity of the intestines to form hernial sacs. Eppinger (1870), Langzert (1871), Jonnesco (1890), and Brosihe (1891) all added descriptions and cases which are summed up by Moynihan (65), Moynihan (65) (1906)

explains them as "fusion folds", a failure of the mesentery of the intestine to fuse with the posterior abdominal wall. He further states that, "in its gradual augmentation in size the hernia will be affected by (66) the degree of laxness of the retro-peritoneal tissue, and (60) the extensibility of the peritoneum". (65)

Every possible type of fold and pouch has been described in explaining the etiology of intra-abdominal herniae. Moynihand (65) lists nine such duodenal fossae; four folds about the cecum and appendix forming three fossae; the intersigmoid fossa; and hernia into the foramen of Winslow. Gray (66) has five duodenal, three cecal, and an intersigmoid fossa.

Following their time it was the rule of writers to accept the theories of Trietz and Moynihan. Edmund Andrews (67) in 1923 expressed the belief that duodenal hernia was a misnomer, and that the condition is due to congenital anomaly of imprisonment of the small intestine beneath the mesentery of the developing colon. He lists the following facts to support his point of view:

1. Differential pressure is utterly lacking.
2. There are literally hundreds of similar folds and fossae.
3. In all but a very small minority of cases reported, the degree of herniation has been total or subtotal.
4. Vogt reports a case in which such a hernia was found in a newborn infant. Several others have been reported in the very young. Surely one cannot believe that intra-abdominal pressure in uretero has been

the cause of such a hernia

5. The herniated viscera are never anything but small bowel.
6. In many of the cases there has been an almost universal growing together of the contents of the sac, (adhesions).

To explain the formation of a right duodenal hernia he states that instead of the cecum rotating to the left upper quadrant and lying superior to the small bowel it grows to the right to reach the right lower quadrant; the mesentery of the colon then becomes attached to the right side of the posterior abdominal wall and the mesentery entraps the small intestine. The formation of a left duodenal hernia is explained on this basis by the failure of rotation or even a small amount of reverse rotation which entraps the small intestine on the left side.

Left intra-abdominal hernia is about three times as frequent as right hernia and has its orifice facing toward the right. The inferior mesenteric artery and vein are usually found in the border of this sac while the superior mesenteric artery and vein are found in the anterior broder of the neck of the right hernia.

Callender (68) et al perfer to attribute the anomaly to small bowel invagination of the ascending or decending mesocolon while it is still mobile and to refer to them as "hernia into the ascending or descending mesocolon".

The more recent studies and opinions are for the most part in agreement with those considering this condition as a congenital anomaly which is the result of maldevelopment. Most theories agree

essentially with that of Andrews mentioned above.

Diagnosis and Treatment

The clinical diagnosis of intra-abdominal hernia is a very difficult one to make. Preoperative diagnosis has rarely been made. In 1923 Nagel (69) stated that he had found no case of intra-abdominal hernia which had been diagnosed preoperatively. Longacre (70) gives credit to five preoperative diagnoses, one by Abrasgow, Vautrin, and Slaudenmeyer, and two by Haberer. Exner (71) had a case diagnosed preoperatively and gives credit to Kummer for x-ray diagnosis of this condition. Case and Upson (72), and Taylor (73), also have made x-ray diagnoses.

Clinical signs and symptoms may vary from none to partial or acute complete intestinal obstruction. Others have had recurrent attacks in the nature of chronic obstruction. A tumor may be palpable one one side of the abdomen. Percussion over the tumor will be tympanitic and auscultation should reveal a gurgling peristalsis. Vomiting will not be a prominent symptom unless the obstruction is complete. With strangulated, gangrenous bowel, blood will appear in the stool. Severe abdominal pain is usually present. The vomitus will be bile stained and there may be shock and collapse. Many cases are symptom free and the condition revealed only at autopsy.

The roentgen diagnosis in cases of chronic partial obstruction is suspected by watching the barium meal through the proximal half of the jejunum which does not follow the usual course. Alexander (66)

stresses this point. Exner (71) made the observation that the small intestine does not appreciably change position in the abdominal cavity when under manual palpation or upon postural changes of the patient.

Kummer (1921) (quoted by Exner, 71), stated that the following points were important in diagnosis of left duodenal hernia: (1) total absence of small bowel in the true pelvis in the upright position, (2) the confining of the small intestine to a smooth sharply circumscribed mass, (3) with the patient in the lateral decubitus position with the left side up the small intestine casts an oval shadow on the film the main axis of which lies to the left of the midline.

Wagner, in 1916, (quoted by Exner, 71), mentioned stasis in the stomach, duodenum, and first part of the jejunum, as an important observation. Taylor (73) (1920) described the x-ray appearnce of the small bowel as having a "bunched appearance of the small gut, as if it were contained in a bag". The opinion of most observers appears to favor this and stasis to be the most characteristic findings.

The treatment of this condition will be surgical since the findings which bring such cases to attention are those of obstruction. Sometimes the hernia can be reduced and the neck of the sac closed by suture. Care must be exercised not to injure the vessels which lie along the anterior margin of the sac. Where the neck of the sac is difficult to find it may be opened in an avascular area and the neck approached from within. Strangulation and infarction of the intestine will require resection. Reduction without closing the open-

ing of the sac had resulted in recurrence.

An extremely rare congenital anomaly akin to intra-abdominal hernia is that of passage of a loop of small intestine through a congenital defect in the mesentery. This is probably the rarest of causes of intestinal obstruction. Brown (74), in 1920, gathered twenty cases. Cutler (75) added eight cases in 1925. Since then several more have been reported.

The defect is usually just medial to the artery supplying the terminal ileum, the appendix, and the first part of the cecum. Trauma and inflammation have been given as the cause of these defects but it is more strongly believed that they are of congenital origin (76). The mechanism by which a loop of intestine passes through such a defect has not been explained. Once the process has begun more and more intestine passes through, and strangulation occurs early. Findings will be those of acute small intestinal obstruction. Early diagnosis and operation are essential. In twenty-one operated patients with eight resections the operative mortality was 28.5% (76).

IX. CONGENITAL ANOMALIES OF THE ANUS AND RECTUM

Embryology - Pathology

These are the most frequent anomalies of the gastro-intestinal tract occuring in approximately one in 5,000 newborn. In the 7.5 mm. (5 week) embryo the cloacal cavity is common to both the urogenital and intestinal tracts and beyond it the intestinal tract extends as the tail gut. The separation of the two tracts by the downgrowth of mesoderm between the two takes place and is

normally complete by the 16 mm. (7th week) stage. The perineum is developed from the cloacal membrane and the urogenital sinus acquires an external opening between the sixth and eighth weeks. During this time there is also a dimpling and inpocketing externally to form the proctodeum. Rupturing of the anal membrane about the eighth week establishes the continuity of the lumen from the rectum to anus.

It is apparent that most of the anomalies of the anus and rectum result from an arrest of development at this period from the sixth to eighth week (7mm. to 42mm.). Failure of complete separation of urogenital sinus and cloaca will result in a fistula. Incomplete dissolution of the anal membrane will result in imperforate anus.

Those cases in which the rectum ends in a high blind pouch are possibly due to the obliterative process which causes degeneration of the tail gut extending into and including the lower part of the cloaca which would be the lower part of the rectum. Where the anus is normal and the rectum ends blindly at 4 cms. the process is probably due to obliteration or concrescense occuring at this point of the bulbus terminalis only.

See the following page for types:

Types:

TYPE I.

TYPE II.

TYPE III.

TYPE I. Stenosis, and, or incomplete rupture of the anal membranes up to 4 cms.

TYPE II. Membranous imperforated anus.
TYPE III. Imperforate anus and high rectal pouch.

TYPE IV.

TYPE V.

TYPE VI.

TYPE IV. High rectal pouch, normal anus and anal pouch.
TYPE V. Common congenital fistulae in females.
TYPE VI. Common congenital fistulae in males.

Any type of anal or rectal abnormality may have an associated fistula. In the male the fistula may be rectoperineal, rectovesical, or rectourethral. In the female the fistula is usually rectovaginal but the other types occasionally occur.

Paul of Aegina has been credited with recognizing this anomaly and describing an operation for its relief in the seventh century. He passed a bistoury and later dilated the opening. In 1640 Scultet successfully dilated an anal stenosis with gentian roots (77). Saviard reported such a case in 1693. This was the treatment used until 1835 when Amussat (77) recommended proctoplasty and in 1844 French surgeons advised inguinal colostomy when perineal exploration failed. Dieffenbach, reported by Harken (78), performed a proctoplasty for this anomaly as early as 1826 but was not reported until 1845.

Many classifications of these anomalies have been made but that of Ladd and Gross (79) is the most practical and most widely used.

Diagnosis of abnormalities of the anus and rectum is not difficult. The age at which patients have symptoms is dependent upon the type of anomaly. Those with a complete block will come to attention in the first few days of life whereas those with stenosis (type I) or a fistula large enough to allow bowel movement to occur will be seen later, sometimes after many years. Obstipation is usually marked to severe in these patients and especially so when enamas are not given. The stool is ribbonlike if the stenosis is marked. Abdominal distention often occurs.

Patients with a fistulous tract large enough to partially or completely empty the rectum receive attention because of pain on defecation, obstipation, or abnormal opening discharging feces and may not be seen until as old as ten to eleven years. The average age in the series of Ladd and Gross was fourteen months. Complete obstruction in these occur rarely but abdominal distention is common.

When an imperforate membrane is present (type II) the symptoms are acute and therefore these cases are seen early -- in the first several days of life. The child may refuse to nurse, have vomiting and abdominal distention. Often someone has noticed that no meconium has been passed or that a rectal thermometer or enema tube cannot be passed. The dark meconium can be seen through the bulging membrane. Those with a high blind rectal pouch in the pelvis (type III) will have the same findings with a dimpling where the anus should be.

Type IV patients, with a normal anus and a high blind pouch will often receive attention late, since the anomaly is not suspected and often are not seen until the 3rd to 5th day of life which makes them poorer surgical risks. The importance of examination of all newborn in order to detect these anomalies early should be emphasized.

Wangensteen and Rice (80) described a method of determining the position of the blind pouch by holding the infant head down, allowing the air in the colon to rise to the end of the pouch. Ladd and Gross stress the point that unless sufficient time has elapsed from birth for adequate gas to be present in the colon before this study is done, it may yield no information or lead to an erroneous

conclusion that there is an atresia of the colon or small intestine. They feel that after twenty-four hours from birth is sufficient time to give an accurate picture. Wilson (81) made the point that the infant should be inverted for a minimum of ten minutes in order for sufficient gas to collect in the lower end of the rectal canal to give an accurate picture. He also stressed the importance of lateral films in determining the amount of tissue between the rectal pouch and the perineum.

Fistulous tracts may be injected with opaque oils to determine their course and extent. This may also be done into the distal loop of patients with a colostomy to outline the anomalies present.

Treatment for anal or rectal stenosis is repeated dilatation and in most cases is sufficient. In cases of an imperforate membrane incision and dilatation is all that is necessary.

In anomalies of type three method of treatment will depend upon the heighth of the rectal pouch. The perineal approach is the method of choice. If the puch is two or three cms. above the anus it can mobilized and brought down. When the pouch is higher a transverse colostomy should be done and proctoplasty postponed as there is often too much difficulty mobilizing the high pouch or too much tension on the suture line with sloughing, giving a poor result.

A primary colostomy with a delayed perineal operation is the method of choice in type four patients. Often the two blind pouches can be anastomosed by an oblique junction to prevent stenosis.

In occasional cases where the patient is in too poor condition to tolerate any type of surgery and a fair sized fistula exists this can be dilated and obstruction relieved. This has the disadvantage of making the fistula more difficult to close later.

When a recto-vesical fistula co-exists with any of the four types, and the fistula is small it will often close spontaneously following re-establishment of normal anal outlet. Some recto-vaginal and most recto-perineal fistulae can be treated successfully at the time of initial surgery, but the recto-vesical, recto-urethral, and high recto-vaginal fistulae as well as any other type that presents difficulties of dissection should be left until the child is older.

Brenner (82), in 1915, reported a series of cases with a twenty-vie percent mortality and there has been no drastic reduction in this figure.

There is considerable difference of opinion expressed in the literature as to the existence of anal sphincter muscles in these anomalies. The concensus of opinion seems to be that it is always present unless destroyed by dissection at the time of operation. Since the sphincter arises from the mesoderm and is not dependent upon the existence of the hind-gut, it should be present in most cases.

SUMMARY

Congenital anomalies of the esophagus are attributed to the failure of the embryonic vacuoles to coalesce so that continuity of the lumen is not re-established after this stage. An arrest in development or failure of the mesoderm to separate completely the trachea from the esophagus results in formation of a tracheo-esophageal fistula. Congenital anomalies of the esophagus occur in about 1 in 50,000 births. Atresia is usually present preventing adequate nourishment making the condition incompatible with life. The primary symptoms are excessive mucus, salivation, regurgitation, choking and dyspnea. Feedings are vomited almost immediately. Diagnosis can be made by roentgenographic examination of the gastro intestinal and respiratory systems. After the diagnosis has been made, the patient is further prepared for surgery. As soon as possible by correction of fluid balance, administration of plasma or blood and vitamins C and K. A right paravertebral incision is used, the fistula is ligated extrapleurally and primary end to end anastomosis is done. When primary anastomosis is not possible, a multiple stage procedure must be resorted to.

Congenital pyloric stenosis is the most common condition requiring surgery in the newborn period. The condition is caused by hypertrophy of the circular pyloric musculature which compresses the underlying mucosa producing stenosis of the lumen of the intestine. The signs and symptoms appear from the fourth to eighth week usually. The vomitus does not contain bile which is important in differentiating

it from a lower obstruction. Also, it must be differentiated from intracranial lesions and pylorospasm. The electrolyte disturbance must be corrected preoperatively. Ether is the anesthesia of choice and the Rammstedt technique is most commonly used today. Post-op feedings are frequent and small. There are a few men who treat this condition medically, but it definitely has a higher mortality rate than surgery when performed by experienced men.

Congenital intestinal atresia and stenosis occur in about 1 in 20,000 births. The majority of atresias and stenoses occur in the duodenum, in the jejunum and more rarely in the ileum. The symptoms begin in first day or two of life and the vomitus always contains bile. Also, there are no epithelial cells found in the meconium. Gastroenterostomy is required for atresia of duodenum. Side to side anastomosis is used for lower atresia. Multiple atresias must always be ruled out at time of surgery. The prognosis for stenosis is still very poor.

Annular pancreas and meconium ileus secondary to pancreatic achylia due to obstruction of the ducts are two rare anomalies of the pancreas. Duodenojejunostomy is the procedure of choice.

Duplications of the GI tract may occur at any place from the base of the tongue to the rectum but are most frequent in the terminal ileum and the just few inches of the ascending colon. The symptoms may occur at any age and are usually those of intestinal bostruction by encroahment on the lumen of the bowel. Surgical removal is the treatment of choice. Enucleation should be done, but if impossible end-to-end

anastomosis or marsupialization of the cyst can be done.

Incomplete rotation of the intestine is a condition which may cause symptoms at any age, but the majority appear in the first year of life. Vomiting, abdominal distention, fever and dehydration are the outstanding signs. Volvulus is commonly associated with it and surgery is required.

Intra-abdominal herniae are very rare causes of intestinal obstruction. Left hernia is about three times as frequent as right. It is now generally concluded that is a congenital anomaly which is a result of maldevelopment. Preoperative diagnosis is rarely made. Surgical repair is treatment of choice.

Congenital anomalies of the anus and rectum are the most frequent anomalies in the GI tract occuring one in 5,000 newborn. These anomalies commonly have rectoperineal, rectovesical or rectourethral fistulae associated with them. Surgery is indicated if there is complete obstruction while repeated dilatation will suffice for simple rectal stenosis.

CONCLUSION:

Although congenital anomalies causing obstruction have a low relatively incidence, it is important to recognize them because they are amenable to surgery and have a good prognosis in general.

1. The literature was reviewed concerning common congenital lesions of the gastro intestinal tract causing obstruction.

2. The direct attack of primary end-to-end anastomosis is the treatment of choice for atresia of the esophagus.

3. Pyloric stenosis in infants is best treated surgically by using the Rammstedt Technique.

4. Pyloric Stenosis in adults (secondary to ulcer) is best treated by gastric resection.

5. Gastro enterostomy is required for a high atresia of the duodenum.

6. Side-to-side anastomosis is the operation of choice for lower atresia of duodenum.
Ileostomy followed by a side-to-side anastomosis is operation of choice for atresia of ileum.

7. Stenosis of small and large intestine is treated by side-to-side anastomosis, but the prognosis is not favorable.

8. Duodenojejunostomy is the operation of choice for annular pancreas.

9. Surgical removal for enterogenous cysts should be performed if possible. Marsupialization of the cyst can be done but should be reserved for patients who are poor surgical risks.

10. Surgical treatment is required for malrotation of the intestine. If a volvulus co-exists, it must be reduced and all obstructing peritoneal bands must be severed.

11. In intra-abdominal herniae, the hernia must be reduced and the ~~pouch~~ neck of the sac sutured.

12. Treatment for anal and rectal stenosis is repeated dilatation.

13. Colostomy followed by perineal proctoplasty is required when there is a high rectal pouch.

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