Three Varieties of Congenital Diverticulum of the Intestine

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A congenital diverticulum of the intestine is a continuous threat to the health of its possessor. If it does not become inflamed or ulcerate, it may produce obstruction. If these hazards fail to materialize, neoplastic changes may appear in later life. Only a few such diverticula remain silent throughout a long life.

The embryogenesis of three varieties of congenital diverticula is described here. There are some other diverticula of doubtful congenital origin and many of demonstrable acquired origin. These are usually smaller and less dramatic, both in actual size and pathology.

Of the three types discussed, only Meckel's diverticulum can be considered common. The other two are rare enough that the possibility of their presence is usually overlooked until revealed at operation or necropsy.

**Meckel's Diverticulum**

The most common intestinal anomaly is the persistent of Meckel's diverticulum, a vestigial structure arising from the terminal ileum. It is present in about 2 per cent of the population and is usually said to be more frequently found in males than in females. This is because more males than females suffer from Meckel's diverticulum disease. The sex bias disappears when the diverticulum is reported incidental to surgery or autopsy. This is illustrated in a short series of our own.

The diseases that call attention to the diverticulum are obstruction, ulceration, inflammation, neoplasm, or umbilical symptoms. Most of the sex difference is caused by the greater frequency of obstruction caused by Meckel's diverticulum in male children.

Meckel's diverticulum represents the persistence of an embryonic structure that normally disappears in the 5th week of gestation, or soon afterward, leaving no trace. It is formed early in development as the connection between the primitive gut and the yolk sac and is first known as the vitelline duct. Just before it separates from the gut, its epithelium shows the start of crypt formation, and mucous granules appear in some cells. This suggests that in normal development the potential for growth and differentiation of this tissue is present but is suppressed. If the suppression mechanism fails to operate, the tissue not only persists but grows and develops at much the same rate as the normal ileum.

Whether a blind-ending diverticulum or a patient omphaloileal fistula is left depends upon the length of the unsuppressed portion of the duct. If the whole of it is withdrawn into the abdomen with the return of the intestines during the 10th week, there will be a diverticulum. It its tip remains in the cord, ligation of the cord at birth will result in a fistula. Very rarely only a distal segment of the duct escapes suppression, and a cyst, umbilical sinus, or umbilical polyp results. Although these latter anomalies are related to Meckel's diverticulum, they are outside the scope of this discussion.
From its developmental origin, Meckel's diverticulum must arise from the antimesenteric border of the ileum, although some diverticula may curl around the ileum and adhere to the mesentery. Diverticula that arise from the mesenteric border are not Meckel's.

Meckel's diverticulum may be the same diameter as the ileum or it may be smaller. Most diverticula will be from 1 to 5 cm long but a few may be much longer. About one-half will be found between 46 and 91 cm from the ileocecal valve. An average distance may be considered to be 40 cm in an infant and 50 cm in an adult. Not less than 5 feet of ileum should be examined in order not to miss a Meckel's diverticulum. There are, of course, limits; we do not believe diverticula arising from the jejunum or the colon to be of Meckelian origin.

In about 25 per cent of Meckel's diverticula the tip is attached by a fibrous cord to the abdominal wall at the umbilicus. The cord represents either the distal vestigial portion of the diverticulum or the fibrosed portion of the superior mesenteric artery beyond the normal ileum. Such attachment increases the danger of obstruction from volvulus around the axis of the diverticulum.

Although Meckel's diverticulum arises from that portion of the intestine destined to become ileum, patches of gastric, pancreatic, duodenal or colonic mucosa in various combinations are common. Gastric mucosa is of special interest because, if it has fundic glands (which secrete hydrochloric acid), symptomatic ulceration of the diverticulum or the adjacent ileum will occur. Gross found gastric mucosa in 54 per cent of 130 diverticula examined histologically. Some writers have placed it as high as 80 per cent. Most of this ectopic mucosa contained fundic glands.

Obviously many Meckel's diverticula remain silent throughout life, but their anatomy predisposes them to some specific lesions.

**Intestinal Obstruction.** A diverticulum attached to the anterior abdominal wall may cause volvulus of the ileum. One not so attached may become the leading point of an intussusception.

**Ulceration.** Hydrochloric acid secreting glands of ectopic gastric mucosa will, if present, produce ulceration.

**Inflammation.** Meckel's diverticulum, like the appendix, may become inflamed.

**Neoplastic Disease.** Nothing in the anatomy or embryology of the diverticulum suggests a cause for the high incidence of neoplasia. More carcinoid tumors and leiomyosarcomas are found in Meckel's diverticulum than would be expected from its size and frequency of occurrence. Curiously, both tumors are found predominantly in males.

**Internal Diverticula**

Internal diverticula produce no visible outpouching of the intestine. The diverticula are contained in the mucosa of the wall (intramural) or hang nearly free in the intestinal lumen (intraluminal). The external appearance of the intestine is that of simple intrinsic obstruction. Both types of internal diverticula are the result of developmental errors in the 5th to 8th
weeks of embryonic life. In both cases the diverticulum, at first small, increases slowly in size, producing first chronic obstruction and eventually acute obstruction in postnatal life.

Starting about the 5th week, the embryonic epithelial tube which represents the future gut elongates faster than does the body of the embryo. In the 6th week the increasing length causes the intestine to herniate into the umbilical cord. Such growth requires rapid multiplication of epithelial cells to cover the increasing surface. Cell division becomes so exuberant that more epithelial cells are produced than are required; the original single layer of cells becomes many layered to the extent that the lumen is, in some places, occluded by epithelial cells. Such occlusion does not take place in all parts of the intestine or, perhaps, in all embryos. It is an incidental result of epithelial proliferation rather than a necessary stage in gut development.

In addition to filling the primary lumen of the intestine, the epithelial cells also bulge into the antimesenteric mesenchyme forming bud-like diverticula. These buds are more frequently observed in the duodenum and the ileum than in the jejunum. Over 40 have been counted in a single embryo.

Normally, as growth of the gut slows, fewer cells divide and vacuoles appear in the thick epithelium; these vacuoles coalesce to restore the intestinal lumen, and the diverticular buds, projecting into the mesoderm, are absorbed. The epithelium is again a single layer by the 10th week.

If the lumen is not restored normally, a pocket may remain in the epithelium, or a solid plug of epithelium may occlude the entire lumen. There is some evidence that mesenchyme is stimulated to grow into such areas of solid epithelial cells, thus establishing a blood supply and a connective tissue core. The pocket will thus become a diverticulum or even a completely closed cystic "duplication" within the intestinal wall. It will be separated from the normal lumen by a septum of connective tissue lamina propria covered on both sides by intestinal mucosa. The mucosa of the diverticulum will correspond to that of the normal intestine at the same level.

Should a solid epithelial plug persist, it will also develop a connective tissue core. Such a septum is often complete, and presents as membranous septal atresia shortly after birth. Sometimes the septum is incomplete, with a central aperture large enough for intestinal contents to pass, but presenting enough resistance that a pocket forms in the valve-like septum. Such a pocket will gradually develop into an elongated sac. This sac is an intraluminal diverticulum. Such diverticula tend to enlarge until chronic or acute obstruction develops.

In some cases, recanalization of the intestinal lumen occurs before the transitory diverticula in the peripheral mesenchyme mentioned above have been absorbed in the growing intestinal wall. The diverticula thus become permanent. They usually remain intramural. If they continue to grow with the intestine they will be true, congenital intestinal diverticula.
Dorsal Intestinal Diverticula

In contrast to Meckelian diverticula that always arise from the antimesenteric border of the intestine, there is another group of diverticula that arises from the mesenteric border. This group includes short diverticula that lie in the mesentery as well as those, more aptly termed duplications, that are several feet in length. The most dramatic are those that arise from the duodenum or jejunum, pass through the diaphragm, traverse the thorax, and attach to a cervical vertebra. These dorsal enteric diverticula or duplications are not restricted to the small intestine but may arise from any portion of the alimentary tube. We will confine ourselves to those arising from the small intestine.

Dorsal diverticula may arise by developmental accident during the 3rd week of embryonic life. Before the notochord appears the embryo consists of two cell layers; ectoderm, from the midline of which will form the neural tube, and endoderm, which will form the future alimentary tract. With the forward growth of the notochord during the 18th to 21st day, the endoderm separates from the ectoderm but becomes attached to the growing notochord. At about the 24th day the mesodermal notochord separates from the endoderm. At this time the dorsal elements of the embryo (ectoderm and notochord) are growing cranially faster than is the endoderm. If, during this shearing movement, the notochord has not completely separated from the endoderm, a thin band of endodermal cells will be pulled cranially and dorsad, forming a bridge between the two structures. Should the endodermal band remain attached at both ends, then it will reorganize itself into a tubular structure similar to embryonic gut. The epithelium will induce the surrounding mesenchyme to form the typical connective tissue and muscular coats, and the result will be a giant diverticulum passing from the intestine and attaching to the vertebra. At the point of attachment, one or more vertebral bodies may show anterior spina bifida.

Much more frequently, the connecting band becomes attenuated and breaks, the portion attached to the gut forming a diverticulum passing cranially in the mesentery. If it is long it will usually be called a duplication. The mucosa of the diverticulum or duplication may be that of any gastrointestinal segment. Gastric mucosa is often present at the cranial end, and eventually causes ulceration.

Ulceration, inflammation, and obstruction of the normal channel by distention of the diverticulum are the usual indications of the anomalous structure.

Excision of these diverticula on the mesenteric side of the gut is more difficult than excision of antimesenteric diverticula. The blood supply to the normal channel passes over the diverticulum so that removal of the diverticulum or duplicated segment requires removal of an equal length of normal intestine. In a few cases a common wall between the diverticulum and the normal gut can be extirpated.

Other Diverticula

The developmental mechanisms described above do not account for all intestinal diverticula. Some of these may be of congenital origin, or may occur at points of congenital weakness of the intestinal wall. The exact etiology of many such diverticula is still obscure.
and often controversial. Only the three groups described above can be certainly attributed and well understood developmental defects.