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Duplications of the Alimentary Tract, Mesenteric Cysts, Transitional Forms

By

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The term "duplication" was coined by Ladd [6] who tried to clear up the terminological chaos in the nomenclature of the pertinent anomalies. A multitude of terms were current (enterogenous cyst, enterocyst, giant diverticulum, ileum duplex, etc.) although — essentially they all referred to the same malformation. They are all oval, elongate or tubular cystic structures situated beside the digestive canal, usually on its mesenteric or dorsal side; they may be closed (80 per cent) or communicate with certain parts of the alimentary tract (20 per cent). Blood is generally supplied to them by the vascular apparatus of the adjacent gastrointestinal segment. Their wall contains smooth muscles and a mucous membrane which is not always similar to the lining of the oesophageal, gastric or intestinal segments situated at the same height. Duplications sometimes run along the entire length of the alimentary canal, but occur most frequently around the ileum (70 per cent), sometimes in the posterior mediastinum and even near the mouth [1], the root of the tongue [4], and in the rectal area. Their contents may be serous, mixed with blood, and possibly infected. Thoracic duplications are of two kinds. One type is entirely within the chest (usually in the posterior mediastinum); in the other form a fistula extends across the diaphragm to the upper part of the abdominal cavity and opens there into the duodenum or the jejunum. In an unusual form of abdominal duplications the entire large intestine is duplicated, and there are two anuses and a double urogenital apparatus (bladder, penis or vagina).

The embryological background of the anomaly is still somewhat obscure. According to the earliest theory, advanced by Lewis and Thyng in 1907 [8], duplications originate from epithelial islets or diverticula. Bremer [3] attributed duplications to non-confluent vacuoles which arise at an early stage of intestinal development. Unfortunately, none of these hypotheses explains why cysts always originate from the mesenteric or dorsal aspect and why their mucosa often differs from that of the adjacent segment of the digestive tract. McLetchie et al. [9] were the

first to note that thoracic duplications were usually associated with vertebral malformations, anterior and posterior spina bifida, the Klippel-Feil syndrome, and that abdominal enterocysts were also often accompanied by vertebral anomalies or at least strands running from the cyst abdominal duplications, other cysts and fistulae situated in the vertebral canal and on the dorsal aspect of the spinal column which contain intestinal mucosa, further diastematomyelia and diplomyelia, mesenteric cysts, vesico-intestinal fissure, malrotation as also certain forms of dia-

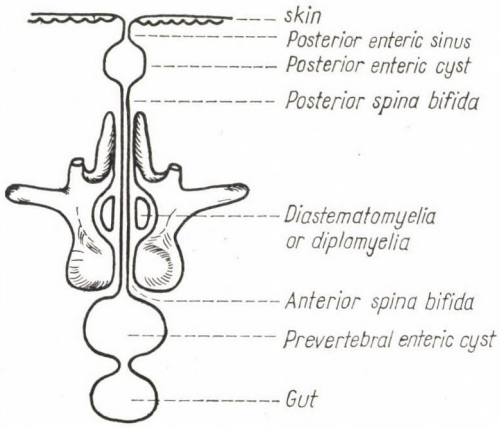

FIG. 1. Diagram of malformations constituting the split notochord syndrome (after BENTLEY and SMITH, 2)

to the spinal malformation. According to their theory, what occurs is the splitting of the dorsal chord and, as a consequence of ento-ectodermal adhesion, a diverticulum-like traction or abstriction of the foregut gives rise to duplication. This is the reason why the anomaly is often accompanied by vertebral changes, while the strand which can be found in some cases is a remnant of the neuroenteric canal. This theory seems to come nearest to reality and is in no contradiction to either the situation or the histological structure of the formations in question. BENTLEY and SMITH [2] suggest that thoracic and phragmatic hernia (with or without anterior or posterior spina bifida) have to be classified with the split notochord syndrome and can be traced back to a developmental disorder of the dorsal chord (Fig. 1).

It is on account of their topographical diversity that the symptoms of oesophageal-gastrointestinal duplications show a great variety, a fact which often makes reliable preoperative diagnosis impossible. Abdominal duplications give rise to intestinal obstruction owing to their enlargement caused by infection. Compression and obstruction of the intestines, or the strangulation of mesenteric

vessels may also bring about an acute abdomen. Haemorrhage, ulceration or perforation may point to the existence of abdominal or thoracic dupli cations containing gastric mucosa. Thoracic duplications may involve pulmonary compression and its consequences (pneumonia, atelectasis). Radiologically demonstrable growths in the thoracic cavity, especially in the posterior mediastinum, and associated with some spinal malformation, also point to the possibility of a duplication.

Treatment consists in the removal of the duplication which — owing to the common mesentery — includes resection of the affected intestinal segment, while the cyst alone has to be extirpated in the cervical, thoracic and rectal areas. When a thoracic duplication passes through the diaphragm and extends to the jejunum, it will suffice to extirpate its thoracic portion and ligate the diaphragmatic part. The time of operation depends on the appearance of symptoms which may occur at any time between birth and adult age. The rate of operative mortality, influenced by numerous factors, has dropped from 30 to 8 per cent in the last 25 years [4].

Most authors make a sharp distinction between duplications and so-called mesenteric cysts although the latter's situation in the abdomen is like that of intestinal duplications; they, too, are situated in the mesentery of the small intestine or the mesocolon, dorsally to the intestinal canal. Mesenteric cysts are held by many authors to be chyle cysts because some of them contain a chylous substance. Their thin walls usually consist of fibrous connective tissue with endothelial lining and their contents are serous, sometimes chylous. These structures may attain a considerable size and may manifest themselves with pain and intestinal obstruction. Larger cysts are usually palpable. Treatment consists in removal which is mostly possible without injuring the mesenteric vessels or the intestine.

Case Reports

Case 1. M.K., a female child 8 years of age, was admitted with an acute abdomen. Surgical exploration revealed an apple-sized, thick-walled, perforated cyst (Fig. 2) which — situated in the lower part of the ileum, 60 cm from the ileocolic valve — was closely adhering to the mesenteric side of the intestine. The cyst was removed together with a 7 cm segment of the small intestine and this was re-united by side-to-side anastomosis. Microscopic examination showed tangled smooth muscle bundles

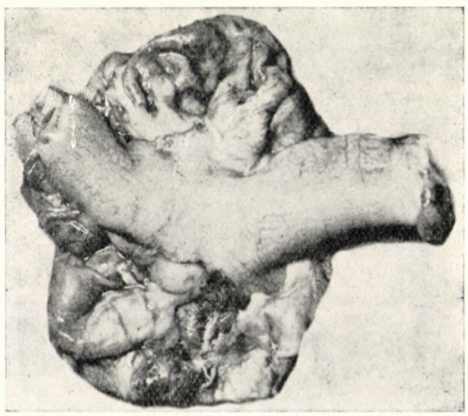

Fig. 2. Case 1. Ileum duplex extirpated together with a segment of the small intestine

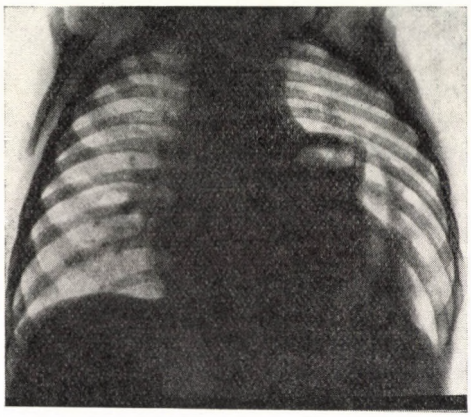

FIG. 3. CASE 2. Radiogram of thoracic enterocyst after puncture

and inflammatory infiltration in the cyst wall lined with several rows of cuboid epithelium. The intestinal surface was covered by inflammatory granulation tissue. Recovery was smooth.

Case 2. D.P., an 8 months old male infant born with 2900 g and weighing 4 kg at admission. The baby had no appetite, vomited frequently, had sudden bouts of temperature. X-rays revealed a small fist-sized intensive homogeneous shadow in the left thorax. Bronchography showed complete filling on the left side, but the lower lobar bronchi were displaced ventrally. Diagnostic puncture proved the structure to be a cyst from which a turbid fluid was collected yielding intestinal bacteria, and thus indicative of a communication between the cyst and the digestive tract. Following puncture, fluid level formation was observed in the cyst (Fig. 3). X-rays of the alimentary tract showed dextrolateral displacement of the lower third of the oesophagus and a normal stomach and duodenum; with the filling of the small intestine contrast material appeared in the lower part of the chest, but the communication was not detected. After injecting contrast material into the cyst, X-rays revealed a tortuous duct running in the midline from the middle of the cyst's right side to below the diaphragm. One hour later, the bulk of the contrast material was seen to have found access to the upper loops of the small intestine, but the site of communication still remained hidden (Fig. 4). By the 12th hour, a minimum of contrast material had only remained in the cyst, the rest having passed into the large intestine. As a preoperative measure repeated punctures of the cyst were done and antibiotics were administered. The general condition showed no improvement, the bouts of temperature persisted so that at the age of 11 months it was decided for surgery. Left posterolateral approach revealed an apple-sized cyst closely attached to the aorta and the oesophagus; its serosa and the course of the vessels were similar to the usual gastric pattern. Since the life of the patient seemed to be in serious danger, two thirds of the cyst were resected and the rest was marsupialized. The baby died three hours later with circulatory failure. At autopsy, a 4 mm wide tortuous thick-walled duct was found; it originated from the medial aspect of the residual cyst; after running first upward and then distad, it pierced the diaphragm between the oesophagus and the vertebral column to end in the lower

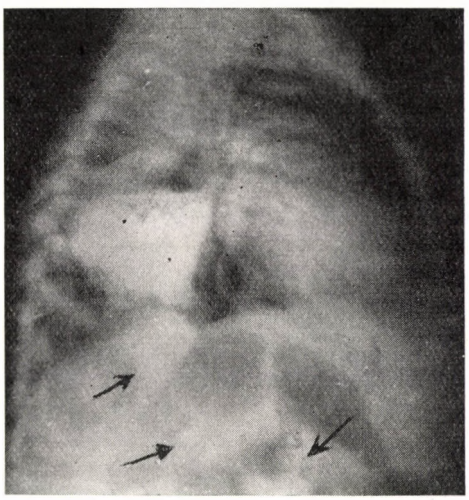

FIG. 4. CASE 2. Passage towards the small intestine of contrast material injected into the thoracic enterocyst

part of the duodenum (Fig. 5). Microscopic inspection revealed a longitudinal and a circular layer of muscles in the wall of both the cyst and the duct; some portions of the mucosa consisted of stratified squamous epithelium like the oesophageal mucous membrane, but the greatest part, containing narrow glandular passages lined with mucous columnar epithelium, was exactly similar to small intestinal mucosa.

Case 3. J. A., a 9 months old male infant was admitted with pyrexia and recurring melaena. The baby was moderately anaemic. Haematology and coagulograms showed no change. X-rays revealed a walnut-sized homogeneous shadow on the right side before the spinal column, at the height of the fifth thoracic vertebra (Fig. 6). Hemivertebrae and anterior spina bifida were observed along the first to eighth thoracic vertebrae. The upper digestive tract appeared to be normal. The thoracic shadow, the vertebral anomalies and the repeated melaena pointed to thoracic duplication. The presence of blood in the stools was attributed to a mucosal bleeding of the duplication communicating with the upper part of the digestive canal. On blood transfusions and vitamin K treatment the melaena disappeared after a few days. Dextrolateral thoracotomy revealed in the posterior mediastinum before the fifth thoracic vertebra a walnut-sized, thick-walled cyst; this was closely attached to the posterior wall of the chest, and from its lower part a narrow duct ran downward. The duct was ligated, the cyst extirpated and the thorax closed. Microscopic inspection showed a double wall; the outer contained longitudinal, the inner transverse smooth muscles. Certain portions of the cyst were lined by stratified squamous epithelium suggestive of oesophageal mucosa and the submucosa contained an abundance of lymph follicles. Recovery was uneventful.

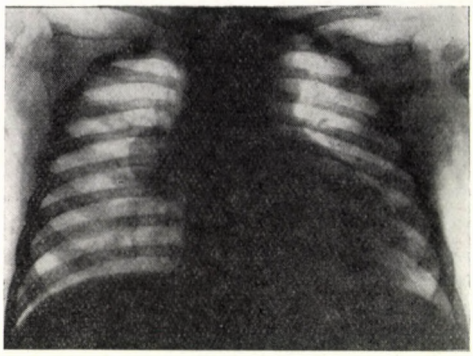

FIG. 6. CASE 3. Radiogram of thoracic enterocyst

Nine months later the child was admitted to another hospital with an acute abdomen; laparotomy revealed a perforated tubular duplication of the ileum. The opening was closed with sutures and, in a second session, the duplication was extirpated. One end of it communicated with the ileum. On its mucosa there were two ulcers, one was responsible for the perforation, the other penetrated into the abdominal wall [11]. This raised retrospectively the suspicion that the original melaena may have been due to the ulcer communicating with the intestine. The problem could not be settled reliably as the blood in the stools may have originated from the stomach or from the duplication lined by gastric mucosa.

Case 4. P. K., a male infant was referred to us from the obstetric ward at the age of 10 days. Six days before a resistance

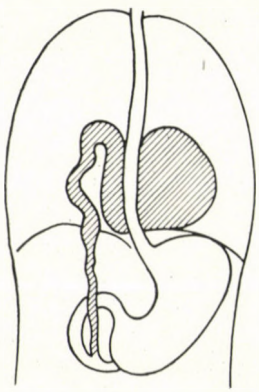

FIG. 5. CASE 2. Diagram of thoracic enterocyst communicating with the duodenum

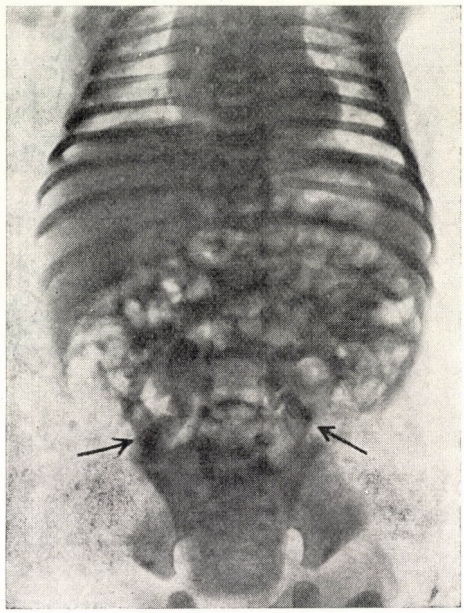

FIG. 7. CASE 4. Postoperatively photographed bilateral megaureter

was palpable in the abdomen and an elastic mass was observed between the anus and the right ischial tuberosity. Catheterization produced 130 ml urine; the abdominal resistance was reduced but did not disappear. Its puncture yielded 5 ml of protein-rich yellowish fluid. The baby discharged urine regularly through the indwelling catheter, but when it was removed, spontaneous voiding ceased. The abdomen was distended from time to time, and faeces were only discharged and flatuses escaped on rectal tubage. Abdominal X-rays revealed in the projection of the pelvis a soft shadow; arching upward, it extended to beyond the ileal crest and pushed the intestines upward. In the cystogram the urinary bladder appeared to be pushed upward and forward. Rectal examination revealed in the minor pelvis a palpable elastic structure which was compressing the rectum. The contrast material injected into the structure accumulated in the lower part. With an alternative diagnosis of a dermoid cyst originating from the pelvis or a rectal duplication, lower median laparotomy was performed. In the minor pelvis an apple-sized cyst was found; it pressed the bladder forward and upward. The latter was distended, and its wall hypertrophic. Both ureters had the width of a little finger (Fig. 7). After puncturing the cyst it was dissected out as far as the minor pelvis. Except for a moderate dilatation, the intestines (examined as far as the lower part of the pelvic colon) appeared to be normal. After draining the cyst it was put back into the pelvis and peritonized, and the abdominal cavity was closed. Then the baby was brought in a lateral position and from a curved incision between the anus and the right ischial tuberosity the lower part of the cyst was exposed and removed with the exception of a small mural portion closely attached to the lateral wall of the rectum. The structure had, thus, the shape of a dumbbell: its larger half was situated above, its smaller one beneath the inlet of the minor pelvis (Fig. 8). The narrow middle portion, situated between the symphysis and the coccyx, compressed the urethra and the rectum. The baby tolerated the operation well; spontaneous bowel movements and diuresis started on the first postoperative day. The wall of the cyst consisted of fibrous connective tissue, and its lining of endothelial cells. The baby now is well and developing normally.

Case 5. J. C., a male child of 6 years, was admitted with abdominal complaints

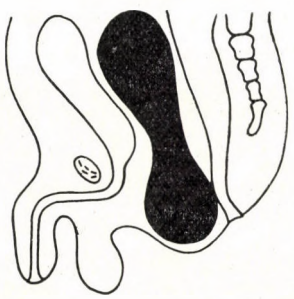

FIG. 8. CASE 4. Diagram showing position of cyst in the minor pelvis

of four days standing, vomiting and intestinal obstruction. There were no bowel movements or escape of flatus, the abdomen was tense, bulging and tender, the tongue dry. After adequate preparation laparotomy was performed. The abdominal cavity yielded a large amount of turbid fluid and revealed a volvulus of the middle part of the ileum, caused by a fist-sized cyst closely attached to the intestinal wall. The small intestine was considerably distended above, and collapsed beneath, the cyst, the resection of which involved the removal of the intestinal segment adhering to it (Fig. 9). After side-to-side anastomosis the abdomen was closed. The child died a few hours after the operation with circulatory failure despite antishock treatment. The wall of the cyst was found to consist of smooth muscles, its lining of cuboid and columnar epithelium.

Case 6. J. H., a male child of 8 years, was referred to us after five days of abdominal complaints. A fist-sized elastic mass was palpable in the lower left abdominal quadrant. X-rays revealed a 10 cm long impression at the boundary of the pelvic and descending colon, outside the large intestine. Intravenous pyelography showed intact kidneys, pelvis and calyces. Laparotomy exposed at the root of the mesentery a baby-head sized, thin-walled cyst. It was enucleated and removed without injuring the mesenteric vessels. Microscopic inspection showed that the wall of the cyst consisted of fibrous connective tissue and smooth muscles; its cavity was lined by endothelium and contained greenish-brown detritus and fluid. The child recovered without complications.

Case 7. E.N., a girl of 3 years, had had abdominal complaints for two days when admitted. An apple-sized resistance was palpable to the right of the navel. The alternative diagnosis was periappendicular infiltration or abdominal tumour. Laparotomy exposed 50 cm from the ileocoecal orifice a fist-sized cyst; it was removed without injuring the mesenteric vessels. Microscopic examination showed that the wall of the unilocular cyst contained fibrous connective tissue and smooth muscle bundles, and was lined with flat epithelium. Recovery was uneventful.

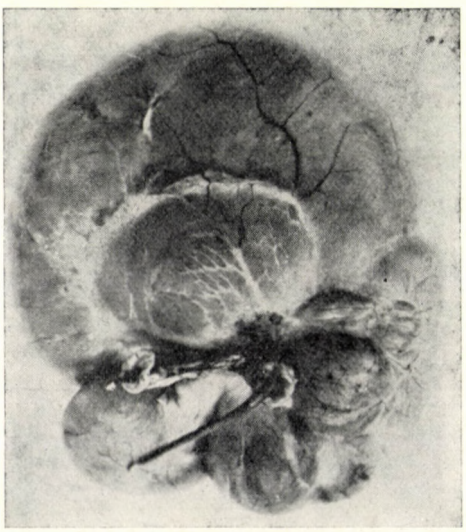

FIG. 9. CASE 5. Abdominal cyst extirpated together with a segment of the small intestine

DISCUSSION

Cases Nos 1, 2 and 3 were undoubtedly intestinal duplications, Nos 6 and 7 mesenteric cysts, Nos 4 and 5 transitional forms. The cyst in Case No. 1 which had a thick wall containing smooth muscle bundles and communicated with the small intestine, was certainly a duplication even if its mucosa — consisting of several rows of columnar epithelium — was not characteristic of duplicated small intestines. Case No. 2 was an intrathoracic oesophageal-gastric duplication, communicating with the duodenum, it had a typical double-layered wall of smooth muscles and a lining suggestive of oesophageal or small-intestinal mu-

cosa. The prolonged septic-toxic condition made it impossible to save the atrophic baby's life. Case No. 3 was likewise one of thoracic duplication accompanied by typical vertebral malformation which facilitated the preoperative diagnosis. The wall of the structure consisted of double-layered smooth muscles, its lining of stratified squamous epithelium. The subsequently detected abdominal reduplication of the small intestine was a rarity; literature contains but a single similar case [5].

Appearance of the structures in Cases Nos 6 and 7, as also the histological findings qualified them as mesenteric cysts although, in addition to fibrous connective tissue, bundles of smooth muscle were also demonstrated in their walls. Both cysts had endothelial lining, and their contents were serous. They could be removed easily and without injuring the mesenteric vessels.

Case No. 5 represented a transitional form. Although the cyst-wall was not thick, it contained smooth muscles and the lining consisted of cuboid and columnar epithelium. The cyst was closely attached to the wall of the small intestine so that the latter too had to be excised. The operation as such was successful but the patient died with postoperative shock.

Case No. 4 was the most puzzling. Situation and shape of the formation pointed to a rare form of rectal duplication which seemed strikingly similar to the anomalies encountered by PILASZANOVICH and HALMOS [10] in two cases. Yet, microscopic examination revealed a structure characteristic of mesenteric cysts and not of intestinal duplications; the wall was composed of fibrous connective tissue, and the cyst was lined by a single layer of flat endothelial cells. It was so large as to compress both the urethra and the rectum of the newborn baby, thus impeding urination and defecation. Compression of the urethra must have occurred *in utero*, hence the extensive distension of bladder and ureter. The structure was no chyle cyst, for its contents were serous and not chylous and because it did not contain fat but protein. It could have been a mesenteric cyst only if it had communicated with the mesentery of the pelvic colon. The topography of the cyst and its close attachment to the rectal wall were more characteristic of duplication, and this the more so as a similarly situated cyst protruding between the anus and the ischial tuberosity, provided with wall and mucosa characteristic of duplications, had been observed by PILASZANOVICH and HALMOS [10] in their first case. In the same authors' second case, of similarly one of supposed intestinal duplication, the cyst wall was likewise composed only of fibrous tissue, while the necrotized lining was unidentifiable.

The histological features of true duplications and mesenteric cysts cannot be as sharply differentiated as was done by GROSS [4]. The epithelial coating in our Case No. 1 did not, for instance, conform to the usual characteristics of the diges-

tive mucosa, although the structure was in all other respects a typical example of intestinal duplication. Neither the wall nor the epithelial lining were typical of any form in case No. 5. Moreover, the cyst in case No. 6 was lined by endothelium, and the wall contained bundles of smooth muscle also. Case No. 4 was one of duplication if on cross examination, but resembled mesenteric cyst under the microscope.

It would follow from the foregoing that there is no essential difference between intestinal duplications and mesenteric cysts either aetiologically or embryologically. It has already been noted that BENTLEY and SMITH [2] grouped under the term split notochord syndrome all cases of anterior and combined spina bifida, diastematomyelia, diplomyelia, postvertebral cysts and fistulae, further vertebral and prevertebral intestinal duplications and mesenteric cysts, and even certain forms of malrotation and diaphragmatic hernia. They rely in this respect on common aetiological factors, splitting of the chord, and the partial or total persistence of the neuroenteric canal (Fig. 1). Such anomalies are really similar to anomalies (persisting omphaloenteric duct, umbilical fistula, Meckel's diverticulum) caused by a complete or partial patency of the omphaloenteric duct.

The different microscopic findings in these conditions may be explained in various ways. The structure of the wall and the lining of cysts depends on whether their differentiation is complete or else has stopped at a certain stage of development, that of the monolayered epithelium of the foregut. It may further happen that, owing to excessive accumulation of the contents, the cyst wall becomes thin and the mucosa flat and endothelioid. These considerations would illuminate the common embryological background of all malformations currently regarded as duplications, mesenteric cysts or transitional forms, making it unnecessary to differentiate between these congenital anomalies. There will, of course, always occur unilocular or multilocular mesenteric cysts containing chylous substance which will have to be considered lymphangiomas [7].

We feel safe in concluding from the foregoing that all disorders grouped by BENTLEY and SMITH under the split notochord syndrome (including intestinal duplications and certain mesenteric cysts) form an aetiological unit; the fact that a vertebral cleft is not always demonstrable need not invalidate this theory considering that the organism tries to close the split cord; if the attempt is successful, the anomaly cannot be demonstrated after birth.

SUMMARY

In connection with 7 own cases the aetiology, symptoms and treatment of thoracic and abdominal intestinal duplications have been discussed. All these conditions should be classified under one aetiological entity, the split notochord syndrome.

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References

1. Abrami, D., Dennison, W. M.: Duplication of the stomach. Surgery **49,** 794 (1961).
2. Bentley, J. F. R., Smith, J. R.: Developmental posterior enteric remnants and spinal malformations. Arch. Dis. Childh. **35,** 76 (1960).
3. Bremer, J. B.: Diverticula and duplications of the intestinal tract. Arch. Path. **38,** 132 (1944).
4. Gross, R. E.: The Surgery of Infancy and Childhood. Saunders, Philadelphia, 1953.
5. Johnston, J. B., Hallenbeck, G. A., Ochsner, A., Dushane, J. W.: Duplications of two parts of the alimentary tract. Arch. Surg. **68,** 390 (1954).
6. Ladd, W. E.: Duplications of the alimentary tract. Sthern med. J. (Bgham, Ala.) **30,** 363 (1937).
7. Léb, J., Dénes, J., Dömötör, L.: Multizystisches Lymphangiom des Mesenteriums im Kindesalter. Z. Kinderchir. **2,** 492 (1965).
8. Lewis, F. T., Thyng, F. W.: The regular occurrence of intestinal diverticuli in embryos of the pig, rabbit and man. Amer. J. Anat. **7,** 505 (1907).
9. McLetchie, N. G. B., Purvis, J. K., Saunders, R. L.: The genesis of gastric and certain intestinal diverticula and enterogenous cysts. Surg. Gynec. Obstet. **99,** 135 (1954).
10. Pilaszanovits, I., Halmos, L.: Rectum duplex eredményesen műtött két esete. Orv. Hetil. **99,** 205 (1958).
11. Strehlinger, L.: Personal communication.

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