DUPLICATION OF THE INTESTINAL TRACT

*Alex Simpson Smith Memorial Lecture delivered at The Hospital for Sick Children, London, W.C.1,
June 21, 1961*

ISABELLA FORSHALL, F.R.C.S., F.R.C.S.(Edin.)

*Consultant Paediatric Surgeon, Royal Liverpool Children's Hospital and Alder Hey Children's Hospital, Liverpool;
Senior Lecturer in Paediatric Surgery, University of Liverpool*

DUPLICATION is the current clinical term for certain accessory structures, with smooth muscle walls and epithelial linings, which usually lie in the dorsal mesentery of the digestive tract. Although this appellation has the demerit of being indiscriminating, it has the merit of being convenient and descriptive, and I therefore propose to retain it.

This paper is based on cases seen by my pædiatric surgical colleagues and myself during the last ten years, three patients treated at the Royal Liverpool Children's Hospital by members of the Chest Unit, and some autopsy findings. Some of these cases have been included in various previous publications. It is proposed to describe the material under two headings:

A—Duplications which appear to result from a localized aberration in development of the wall of the intestine; and

B—Duplications which are part of an abnormality in development; which in its complete form, affects in addition to the alimentary tract, the vertebræ, the spinal cord and the overlying dorsal coverings, including the skin.

Group A—Duplications which appear to result from a localized aberration in development of the wall of the intestine.

Abnormalities of this group occur as single thin-walled cystic structures intimately related to the bowel wall. They lie either within the lumen of the bowel, or between the layers of the mesentery. In either case the circular muscle coat of the intestine is split to provide a thinned-out covering for the cyst, so that it appears as if the cyst has formed within the circular muscle coat (Diagram 1). Intra-luminal cysts are covered for most of their circumference by the bowel mucosa. Extra-luminal cysts, which are usually large, expand both the circular and longitudinal muscle coats, so that they are thin-walled and translucent.

The cysts are lined by entodermal epithelium, usually arranged as a single layer, and often much

flattened; this epithelial lining may or may not be of the same type as that of the mucosa of the bowel to which the cyst is attached. Thirteen children in this series exhibited cysts which fulfilled the above criteria.

There were seven examples of *intra-luminal* cysts (6, 14, 16, 19, 29, 35, 45), all of which were small, varying in size from 0.5 to 2.5 cm. in diameter. They occurred in children whose ages ranged from 4 days to 13 years. The location of the cysts, the type of epithelium composing their lining, and the way in which the patients presented are shown in Table 1 (see also Fig. 1).

Six children (2, 8, 11, 31, 32, 48) aged from 3 days to 3½ months, had *extra-luminal cysts*, which lay between the layers of the mesentery; they were all readily felt on abdominal examination, the smallest being 4 x 2½ x 3½ cm., and the largest from 9 to 16 cm. in diameter, 30 cm. in length and weighing 3 lb. It occurred in an 8½-week-old baby which weighed 9 lb. (No. 48), Figs. 2, 3 and 4. Four of the children were either completely or partially obstructed, and in each of these, obstruction was due to stretching or 'ribboning' of the bowel over the convexity of the cyst, so that the lumen of the bowel was obliterated or almost obliterated (Fig. 5). In the remaining two cases, including that which had the largest cyst, though the bowel was 'ribboned', the babies were not obstructed. In each case the cyst with the involved length of bowel was resected. The types of lining epithelium are shown in Table 2.

These intrinsic bowel-wall cysts, whether intraluminal or extra-luminal, caused mechanical interference with intestinal function, none of them perforated, ulcerated, or became infected. In no case was more than one cyst found, and the children showed no other abnormality of the alimentary tract, apart from one case having a Meckel's diverticulum, and two cases abnormal mid-gut rotation, a point which will be referred to later; the spinal vertebrae of those cases X-rayed were invariably normal.

DIAGRAM 2.—In *A* the abnormality found in the prototype case (No. 21) is outlined. *B* and *C* represent the less extensive thoracico-abdominal duplications of Cases Nos. 27 and 24. *D* (Case No. 3), which had two posterior mediastinal cysts and a long tubular duplication of the jejunum, part of which was extra-mesenteric, shows near-persistence of the original abnormality. *E* (Case No. 28) shows a further stage in obliteration, a posterior mediastinal cyst and a mesenteric cyst remain. *F* (Case No. 20) shows a posterior mediastinal cyst only. *G* (Case No. 12, see overleaf) shows a tubular duplication and a more proximal mesenteric cyst. *H* (Case No. 12a), a tubular duplication, is the only remnant of the original structure. The condition of the spinal vertebræ is indicated.

DIAGRAM 3.—Shows position in which peptic ulcers developed, and their relationship to different types of mucosa in seven intestinal tubular duplications.

DIAGRAM 4.—Development of diverticulum from primitive entodermal tube, resulting from persistent adherence of entoderm to ectoderm. Division of notochords is shown.

A.V.—amniotic vesicle. EC—ectoderm. ENT—entoderm.
M.—mesoderm. N.C.—notochord. S.C.—spinal cord.
V.B.—vertebral body. E.N.T.T.—entodermal tube. DIV.—diverticulum.

DIAGRAM 5.—Shows sagittal section of hypothetical diverticulum from primitive entodermal tube, passing dorsally between hemivertebræ.

A—the diverticulum.
B and C—progressive caudal growth of parent channel and diverticulum.
D—release of diverticulum from vertebral body.
E—closure of communication between parent channel and diverticulum.

DIAGRAM 6.—Shows lesions in cases with intra-spinal entodermal tissue and accessory structures anterior to vertebral bodies.

T.A.D.—thoracico-abdominal duplication.
P.M.C.—posterior mediastinal cyst.
A.C.—abdominal or mesenteric cyst.

DIAGRAM 7.—Cases with intra-spinal lesions without evidence of accessory structures anterior to the vertebrae.

TABLE I
GROUP A—INTRINSIC INTESTINAL DUPLICATION CYSTS. (a) INTRA-LUMINAL

No.	Age	Situation	Size of cyst	Epithelium	Pres-entation	Operation
14	4 days	Distal ileum	2 x 1.5 x 1.5 cm.	Columnar	Intussusception	Reduction of intussus- ception. Excision of cyst
9	6 days	Cæcum	2 cm. diameter	Colonic	Obstruction	Resection
16	3 months	Ileum	1 cm. diameter	Columnar and pyloric gastric (Necrotic)	Intussusception	Resection
19	3 years	Ileum	1 cm. diameter	Columnar	Intussusception	Resection
6	5 years	Rectum	2.5 cm. diameter	Columnar	Prolapse	Excision of cyst
45	8 years	Terminal ileum	2 cm. diameter	Columnar	Intermittent abdominal pain	Resection
35	13 years	Distal ileum	3 cm. diameter	Cuboidal	Incidental finding	Excision

FIG. 1.—Photograph of artist's water colour of intra-luminal intrinsic bowel-wall cyst. Case No. 45.

These intrinsic intestinal cysts can be explained by the time-honoured theory that the original lumen of the primitive entodermal tube is obliterated or narrowed by proliferation of epithelial cells, which then form vacuoles. Should there be incomplete breakdown of the vacuoles to form the definitive intestinal lumen, isolated epithelial-lined hollow structures could result, and become sur-

FIG. 2.—Nine-pound infant with extra-luminal bowel-wall cyst weighing 3 lb. Case No. 48.

rounded by the mesoderm which forms the intestinal muscle (Johnson, 1910). Sequestration of islands of entodermal cells at the time of formation of the circular muscle coat is a possible alternative mode of origin. Ectopia and heteratopia of the epithelium lining the cysts can be explained by the early initiation of the abnormality, at a time when the cells of the primitive entodermal tube have the potential to develop into any type of intestinal or respiratory epithelium. It is, however, difficult to understand why the extra-luminal cysts were always found on the mesenteric aspect of the bowel, and why the extra-luminal cysts were so much larger than the intra-luminal cysts, though

TABLE 2
GROUP A—INTRINSIC INTESTINAL DUPLICATION CYSTS. (b) EXTRA-LUMINAL

No.	Age	Situation	Size of Cyst	Epithelium	Presentation	Operation
31	7 days	Ileum	12 x 8 x 8 cm.	Squamous, columnar pseudo-stratified, ciliated columnar	Obstruction, palpable mass	Resection
2	12 days	Ileo-cæcal angle	10 x 4.5 x 4.5 cm.	Columnar	Palpable mass	Resection
8	4 weeks	Jejunum	9 x 6 x 6 cm.	Pyloric gastric	Obstruction, palpable mass	Resection
48	9 weeks	Ileo-cæcal angle	4 x 4 x 3 cm.	Columnar, pyloric gastric, stratified ciliated columnar	Obstruction, palpable mass	Resection
5	9 weeks	Ileum	30 cm. in length, 9-16 cm. in dia.	Columnar	Palpable mass	Resection
32	3½ months	Jejunum	11 x 8 x 7 cm.	Small intestinal	Partial obstruction, palpable mass	Resection

the average age of the children with the former is less than that of those with the latter. Further, it is curious, if either of the suggested modes of formation of these abnormalities is correct, that in this series, at least, none of the cases exhibited more than one cyst.

Group B—Duplications which are part of an abnormality in development, which in its complete form, affects in addition to the alimentary tract, the vertebræ, the spinal cord and the overlying dorsal coverings, including the skin.

These are the accessory structures which Bentley (1960) has called 'Posterior Enteric Remnants'.

In discussing those accessory structures of this group which lie anterior to the spinal vertebræ, a case which had what will be regarded as the prototype abnormality, will be first described.

A 16-hour-old infant (21), which had appeared normal at birth, developed rapidly increasing respiratory distress and cyanosis. An antero-posterior radiograph of the chest (Fig. 6) showed a translucent right hemithorax, displacement of the heart and mediastinum to the left, and abnormal lower cervical and upper dorsal vertebræ. A presumptive diagnosis of emphysematous cyst was made and immediate thoracotomy decided upon. On opening the right chest a thick walled structure was found to be attached by a narrow stalk to the body of the second dorsal vertebra, whence it bulged to the right, filling the right side of the thoracic cavity and compressing the right lung; it narrowed distally to pass through the diaphragm to the right of the mid-line. It was not attached to the œsophagus, which, with the other mediastinal structures, was displaced to the left; aspiration yielded gas and bile under considerable pressure. After ligation of its upper and lower extremities, the thoracic portion of the

FIG. 3.—Same case as Fig. 2. X-ray film after barium (a method of investigation not advised). Note opaque material in large left inguinal hernia.

duplication was removed; the infant died soon after operation. At autopsy the ligated lower end of the thoracic structure was found to be continuous with a tube resembling small intestine, which passed across the duodenum and transverse colon, to run parallel with, and between the layers of the mesentery of, the jejunum for about 8 cm., 33 cm. from the pylorus the lumen of the duplication joined that of the jejunum. The mid-gut intestine was unrotated. The histology of the accessory structure was constant throughout; it

FIG. 4.—Same case as Figs. 2 and 3. The removed cyst. The 'ribboning' of the involved segment of bowel is well shown.

FIG. 5.—Photograph of artist's water colour. Extra-luminal intrinsic bowel-wall cyst of ilio-caecal angle—'ribboning' of bowel well shown. Case No. 39.

FIG. 6.—X-ray film of case No. 21. Showing lower cervical and upper dorsal hemivertebrae—translucent right hemithorax and displacement of mediastinum to left.

resembled that of normal intestine, having a mucosal lining thrown into villous folds and histologically typical of jejunum. The proximal stump of the duplication, which had been divided at operation, disappeared into a cleft in a wedge of cartilage which divided vertically the body of the second dorsal vertebra; this part of the abnormality is described on p. 576. The immediate cause of the death of this baby was bleeding into the thoracic cavity, the progressive respiratory distress with which the infant presented was due to respiration sucking bile and gas from the jejunum into the thoracic portion of the duplication. It is exceptional for a child to be born with such an extensive accessory structure, and it is postulated that the abnormalities shown by other children in this series (excluding those in Group A), can be explained by partial persistence of the abnormality exhibited by this case (No. 21) (Diagram 2).

In this series there were other thoraco-abdominal duplications, posterior mediastinal cysts, mesenteric cysts and tubular intestinal duplications.

The three last-mentioned occurred either singly or in various combinations.

All these remnants showed the same structure, a thick wall consisting of two muscle layers, with the fibres of the two layers at right angles, muscularis mucosæ, Meissner's and Auerbach's plexuses and a lining epithelium of entodermal origin.

Those accessory structures which occur within the abdomen, whether applied to the gastro-intestinal canal or separated from it, usually lie between the two layers of the mesentery (dorsal mesentery); and share a common blood-supply with the gastrointestinal canal. In the thorax where the dorsal mesentery is lost, and the œsophagus has a segmental blood-supply from the aorta, duplications are usually separated from the œsophagus and obtain their blood-supply locally. The upper abdominal portion of thoraco-abdominal duplications and the proximal extremity of tubular intestinal duplications may follow the thoracic pattern by being extra-mesenteric.

Like other parts of the abnormality shown by the prototype case (21), the vertebral abnormality may or may not persist, or may or may not be radiologically demonstrated to persist. Hemivertebrae is the typical abnormality, but more extensive bony defects are found, as in cases which also have spina bifida; on the other hand, some may show only broadening of the vertebral pedicles (butterfly vertebrae).

Thoraco-Abdominal Duplications

Our cases include two further thoraco-abdominal duplications; neither communicated with the intestine and a spinal abnormality was not demonstrated in the chest films, although in one case the upper end of the duplication was found to be firmly attached to the body of the sixth cervical vertebra.

One of these two cases (27) was a 5-week-old male infant who from birth had had respiratory distress and intermittent dysphagia. This child had an accessory structure which was narrow in the lower cervical region, expanded in the right thorax to displace the mediastinum to the left, and at the œsophageal hiatus, passed to the left behind the œsophagus, to join the greater curvature of the stomach to which it was widely attached, and with which it shared the longitudinal muscle coat; it ended blindly in relation to the second part of the duodenum. The cervico-thoracic portion was lined by intestinal epithelium, the abdominal portion by duodenal epithelium.

The other case (24) was a 6½-year-old boy who was admitted with a head injury; he had never had any respiratory or digestive symptoms, but on routine examination a large cystic tumour was found in the left upper abdomen. The radiologist suggested the diagnosis from the plain films, and confirmed it, in both this and the preceding case, after barium. Except that it did not reach so high in the chest, and was not attached to the vertebrae, the thoracic portion of this duplication was much the same as those of the two preceding cases. It narrowed to pass through the œsophageal hiatus posteriorly to the œsophagus, and then expanded into an enormous cyst-like structure with a very wide muscular attachment to the greater curvature and posterior surface of the stomach. It displaced the transverse colon downwards and the stomach to the right (Fig. 7), the spleen sat on its upper left aspect like a little cap. The thoracic portion was lined by stratified squamous epithelium, the abdominal portion by small intestinal epithelium. In each instance the gastric portion of the accessory structure lay between the layers of the great omentum (dorsal mesentery of the stomach).

FIG. 7.—Case No. 24. X-ray film after barium showing stomach displaced to right and rotated by very large cystic dilation of abdominal part of thoracico-abdominal duplication.

Both cases were dealt with by opening the chest and freeing the duplication down to the diaphragm; the child was then turned on to the back, and the abdominal portion removed through a long transverse incision. Contrary to the usual description of gastric duplications, little difficulty was encountered in freeing the abnormal structure from the stomach.

Posterior Mediastinal Cysts

Posterior mediastinal cysts are seldom attached to the œsophagus, but are not infrequently attached to a vertebral body, and occasionally there is a strand of tissue or hollow structure, which runs from the cyst dorsally between hemi-vertebræ.

In this series there are ten cases which had posterior mediastinal cysts (3, 18, 20, 25, 26, 28, 30, 36, 38, 39). Four of the children (20, 25, 26, 30) had symptoms referable to their cysts; pallor, respiratory difficulty (dyspnœa, cyanosis, stridor), and poor appetite. Pallor was the only complaint common to all four.

One case (26), an 11½-month-old female child, was moribund on admission; a peptic ulcer in a

cyst lined with gastric mucosa had perforated into the mediastinum approximately four days previously (Fig. 8). She died while efforts were being made to resuscitate her.

The cyst of one other child (20) was lined by gastric mucosa, and this cyst showed a single small peptic ulcer.

The other two cases (25, 30) had cysts which were lined by small intestinal epithelium.

In two children, posterior mediastinal cysts were incidental X-ray findings. One child (3) admitted with a perforated ulcer in a long jejunal duplication, was discovered to have a cyst in the right upper chest, and a second in the left lower chest (Fig. 9). All three accessory structures were lined by gastric mucosa. The other case (39) was X-rayed because he had a small lower cervical meningocele, a cyst in the left middle chest was revealed, which proved to be lined by tall columnar epithelium.

As has been said, one child died on admission. The six cysts of the remaining five children were successfully excised.

Four cysts were found at autopsy, including one lined by mucus-secreting columnar epithelium in

FIG. 8.—Perforated peptic ulcer in gastric-mucosa-lined posterior mediastinal cyst. Case No. 26.

a six-day-old female infant (18), who died after resection of a gangrenous volvulus containing a tubular duplication of the ileum. Another was found in a baby (28) who died after staged operations for a large meningo-myelocele; this child, in addition to a post-mediastinal cyst, had a mesenteric cyst; both were lined by pyloric gastric and columnar epithelium. Two cases (36 and 38) in addition to posterior mediastinal cysts, had intra-spinal abnormalities, and will be discussed in this context (p. 586).

Of these ten cases, seven had the characteristic cervico-dorsal, or upper dorsal, vertebral abnormality demonstrated radiologically, the child with the myelo-meningocele had a more extensive spinal abnormality. In one case the chest films showed no abnormality, and the films of the tenth case have been destroyed; no comment on the spine was made in the radiologist's report. In short, of ten children with posterior mediastinal cysts, at least eight showed hemivertebrae. These cases are summarized in Table 3.

Abdominal or Mesenteric Cysts

Abdominal or mesenteric cysts are the least common manifestation of group B abnormality.

In this series there were four thick-walled enterodermal, epithelium-lined, abdominal cysts (12, 10, 28, 34) which lay in the mesentery, and were

FIG. 9.—Case No. 3. Chest X-ray film. Posterior mediastinal cyst in right upper chest and second one in left lower chest. Abnormal upper dorsal vertebrae.

similar in structure to posterior mediastinal cysts.

Two children (12, 34), had cysts which lay cephalad to tubular duplications. In one of these the cyst and tubular duplication were resected with the related small intestine. The cyst of the other child had been removed, before he came under our care with bleeding from a peptic ulcer in an ileal tubular duplication. The third case (34) had a cyst of a similar structure, related to the caecum; the cyst was removed without resection of the bowel being required: this child had a normal spine, and no other abnormality. The fourth cyst (28) was found at autopsy in a child with a meningocele, and a posterior mediastinal cyst. These four cases are summarized in Table 4.

Abdominal Tubular Duplications

These abnormalities have circular and longitudinal muscle coats at least as thick as those of normal intestine. For some part, and often the whole of their length, they are applied to the intestinal canal, the accessory structure lying between the layers of the mesentery and the longitudinal muscle coat being common to it and the intestine. The two channels have individual circular muscle coats, so that the two lumina are separated by a septum composed of two layers of circular muscle with mucosa on either side.

TABLE 3
POSTERIOR MEDIASTINAL CYSTS

No.	Age	Sex	Presentation	Location of Cyst	Size of Cyst in cm.	Epithelial Lining	Spinal Abnormality	Other Abnormalities	Result
3	1½ years	M	Cysts X-ray findings	1. Right upper zone	3 x 2 x 2	Gastric	Hemivertebrae D3-4	<i>Tubular duplication of jejunum</i>	Cysts excised, recovery
18	8 days	F	Cyst found at autopsy	2. Left lower zone	4 x 2.5 x 2	Gastric	No comment	<i>Claspatus tubular duplication of ileum</i>	Died post-operatively of peritonitis
20	1½ years	M	Pallor and anorexia	Left upper zone	3 x 2 x 2	Mucous secreting gastric (fundal)	Hemivertebrae C5-D1 scoliosis	Spiegel's snail-er	Cyst excised, recovery
25	6 mths.	F	Pallor, cyanosis, dyspnea, stridor	Right upper zone	4 x 2 x 1.5	Peptic ulcer	Nil	Nil	Cyst excised, recovery
26	11½ mths.	F	Pallor, poor appetite —3 mths.	Left lower zone	'Large'	Fundal gastric	Hemivertebrae C6-D3	Nil	Perforation of peptic ulcer into left pleural cavity. Death
28	1 day	M	Cyst found at autopsy	Right middle zone	1.5 x 1 x 1	Pyloric gastric and simple columnar	Gross spina bifida (dorso-lumbar)	Meningo-myelocoele, mesenteric cyst — <i>intra-spinal enteric remnant</i>	Died after operation for myelo-meningocele
30	1½ mths.	M	Pallor, cyanosis, vomiting	Left middle zone	Diameter 7.5	Columnar	Hemivertebrae	Pyloric stenosis	Cyst excised, recovery
36	1 day	M	Cyst found at autopsy	Right lower zone	Diameter 2	Columnar, mucous secreting	Hemivertebrae most of dorsal spine, spina bifida, lumbar spine	Omphalocele, meningo-myelocoele	Died after operation for omphalocele and myelo-meningocele
38	3 mths.	M	Cyst found at autopsy	Mid-line upper mediastinum	Diameter about 2	Various types of alimentary epithelium, pancreatic tissue	Upper dorsal hemivertebrae	<i>Intra-spinal cyst</i>	Died—bleeding into subarachnoid space
39	3½ mths.	M	Cyst X-ray finding	Right middle zone	3 x 2	Tall columnar	Upper dorsal butterfly vertebrae	Healed upper dorsal meningocele	Cyst excised, recovery

TABLE 4
MISENTERIC CYSTS

No.	Age	Sex	Location of Cyst	Size of Cyst in cm.	Epithelial Lining	Spinal Abnormalities	Other Abnormalities	Procedure
10	16 mths.	M	Ileo-cecal angle	4 x 2½ x 2½	Gastric, small intestinal and colonic	None	None	Simple excision
12	10 weeks	M	Jejunal mesentery	3½ x 3 x 2	Low columnar	No comment	<i>Long ileal tubular duplication</i>	Simple excision
28	1 day	M	Proximal duodenum	3 x 2 x 2	Pyloric gastric and small intestinal	Gross dorso-lumbar spina bifida	Meningo-myelocoele, posterior mediastinal cyst, <i>intra-spinal enteric remnant</i>	Autopsy finding
34	18 mths.	M	Ileal mesentery	3 x 3 x 2	Various alimentary types and squamous	Hemivertebrae mid-dorsal region	<i>Tubular ileal duplication, ectopia vesicae</i>	Resection of cyst with tubular duplication and segment of ileum

Portions of tubular duplications which are not applied to the gut, whether intra- or extra-mesenteric, have their own longitudinal muscle coat.

The duplication usually communicates by its caudal extremity with the intestine; less commonly both extremities are blind; double colons commonly communicate proximally, but a tubular duplication of the stomach, jejunum, or ileum rarely communicates with the intestinal canal cephalically.

The length is variable, the longest accessory segment in this series was 60 cm.

Of our 11 tubular duplications (3, 4, 5, 9, 12, 12a, 13, 17, 18, 33, 34), one involved the duodenum, one the jejunum and eight were related to the ileum, whilst the position of the eleventh was not determined, apart from 'small bowel'. The epithelial lining was partly or entirely gastric, fundal in type, with oxyntic cells in all the nine cases in which a histological report is available. Acid pepsin secretion from this ectopic gastric mucosa caused ulceration in seven cases, the ulcer or ulcers developing in gastric mucosa or in intestinal mucosa adjacent to gastric mucosa, and either in the accessory structure, or in the normal bowel adjacent to the stoma of a gastric-mucosa-lined duplication. The positions at which ulcers were found, and their relationship to different types of mucosa are shown in Diagram 3.

Of the seven children who developed ulcers, one was a 10-year-old girl (5) with pain and a palpable abdominal mass. At operation, she had a duplication, the distal third of which was applied to, and communicated with, the ileum. The proximal two-thirds lay in the mesentery and was distended into a large (13 x 6 x 5 cm.), stomach-like pouch, lined with gastric mucosa, which showed eight acute ulcers.

The ulcers of six children perforated. One of these occurred in a 13-year-old girl who gave a five years' history of episodes of epigastric pain. At operation there was a healed perforation in a short duplication of the duodenum.

In five children (3, 4, 9, 18, 33), perforation caused general peritonitis, one was a 5½-year-old boy (4) admitted with abdominal pain, diarrhoea, and anaemia: the test for occult blood in the stools was positive. He was removed from the hospital before he had been fully investigated, only to be re-admitted a few days later with general peritonitis; he died following resection (Fig. 10). A 5-week-old female infant (18) died of peritonitis following resection for a perforation in an ileal duplication, whilst the other three cases (3, 9, 33) made satisfactory recoveries.

One or more episodes of severe bleeding from an ulcer occurred in three children (3, 12, 33); in two

FIG. 10.—Large perforated ulcer of ileum opposite stoma of gastric-mucosa-lined tubular duplication. Case No. 4.

cases the blood was described as red, or bright red, in one as melæna. A 6½-month-old boy (12) had had episodes of bleeding, for which he had required transfusions, since 9 weeks of age; he had had a palpable mesenteric cyst removed. On admission, there was obvious blood in the stools and the hæmoglobin was 41%. This is the only case in which operation was performed for bleeding, although in two other cases (3, 33), operated upon for peritonitis due to perforation of an ulcer, a history of bleeding was obtained.

Two cases were admitted with intestinal

FIG. 11.—Case No. 13. Long tubular duplication of ileum (60 cm.) closed at both ends. Obstruction due to pressure of distended proximal end of duplication.

obstruction (12a, 13). One, a five-week-old infant (12a), died following resection of a gangrenous volvulus which included a tubular duplication of the ileum. The other (13), an 11-day-old child, had a 60 cm.-long ileal duplication, closed at both ends, it was distended with secretion and had, by pressure, occluded the lumen of the main channel (Fig. 11). The eleventh case (34) complained of pain and vomiting, but was not obstructed; he had a short ileal duplication, adjacent to which there was a mesenteric cyst. It was thought that his symptoms had possibly been due to intermittent volvulus.

In this series there is a high mortality in cases with tubular duplications, four of our eleven cases died, two deaths followed resection in children with peritonitis due to perforation of an ulcer, and two occurred in babies in which long resections were required for gangrene due to volvulus.

The contention that these eleven cases exhibit the distal part of a more extensive abnormality,

is supported by a number of them having more proximal remnants, in addition to a tubular intestinal duplication. Two had mesenteric cysts, two posterior mediastinal cysts; one cyst and two cysts respectively.

Six children had a vertebral abnormality; in five of these (4, 5, 12a, 33, 34) there was no radiological evidence of a cyst in the chest. This is at variance with the oft repeated statement that abdominal duplications, in contra-distinction to thoracic duplications, are not associated with hemivertebrae.

The dorsal spine was reported normal, or no comment was made by the radiologist in the five remaining cases. Some of the films have been destroyed and cannot, therefore, be checked. These cases are summarized in Table 5.

As these duplications lie between the layers of the mesentery, and have a common blood supply with the involved bowel, it is not possible to remove them without prejudicing the blood supply

of the main intestinal channel, so that resection of both channels is performed. None of our cases had an accessory channel of such a length that its resection with the involved segment of bowel left the child with an inadequate length of intestine, but very long gastric-mucosa-lined duplications which perforate or bleed may pose a difficult problem. Waterston (1961) states that he has successfully cored out the gastric mucosa of a tubular duplication, leaving the muscle coats in place; this manoeuvre would avoid a dangerously long resection.

Embryology

Correlation of normal human embryology with experimental animal embryology, examination of abnormal human embryos, and of autopsy and clinical material, has left little doubt that these Group B abnormalities and their more dorsal extensions are determined at the blastocyst stage of development. They are due to varying degrees of persistence of continuity between the entoderm of the yolk sac and the ectoderm of the future neural plate. Persistence of a neurenteric opening results in a complete spinal cleft and dorsal fistula. Persistent adherence of entoderm and ectoderm without an opening leads to dorsal herniation of gut through either an open neural tube or a diastematomyelia. Less intimate adhesion produces a diverticulum from the gut passing backwards towards the spinal cord (Diagram 4).

Adhesion between entoderm and ectoderm of whatever degree will prevent development of the notochord in its normal mid-line position. The ectodermal cells from which the notochord is developed will be displaced to either side, and two notochordal centres will result. This is the developmental basis of the hemivertebrae which are such an important feature of group B abnormalities. The level of the spinal abnormality is variable, and one or many vertebrae may be affected. In our cases a heavy incidence of abnormality falls in the region of the second dorsal vertebra. This level appears to correspond with the twelfth somite, and the twelfth somite probably corresponds with the level at which, in the pre-somite stage, Henson's node starts to lay down the dorsal axis, and from which it begins its migrations caudally.

It is possible to explain Group B abnormalities of the alimentary canal by envisaging a diverticulum from the primitive entodermal tube passing dorsally between double notochords or hemivertebrae (Diagram 5).

At the time of the formation of the diverticulum, the future of the entodermal cells at point X has been determined; they will become the lining epithelium of oesophagus, stomach, duodenum,

jejunum or ileum and the primitive cells will be influenced or organized accordingly. The entodermal cells lining the diverticulum are the same primitive cells with the potential to develop into any type of alimentary epithelium, or into respiratory epithelium; perhaps because they line an abnormal or accessory structure, they fail to be influenced or organized, with the result that ectopic or heterotopic epithelial linings in duplications are common. As the gut grows rapidly caudally it carries the diverticulum with it; mesenchyme invests both tubes; the circular muscle coat, laid down first, is individual to each, though there will be some intermingling of fibres where the two channels communicate, and for a variable distance proximally. The longitudinal muscle coat, a later development, invests both channels where they are in apposition, individually where there is separation of the two structures. Separation of the two channels, as the point X is carried away from the level at which the diverticulum was formed, explains why thoracic remnants are not usually adherent to the oesophagus. It also explains why, if the point X is destined to travel only to the stomach, oesophagus and duplication pass through the same opening in the diaphragm (24, 27), whereas if it is ordained that point X will become jejunum or ileum, there will be a longer and wider separation between the two channels, and they will pass through separate openings in the diaphragm (case No. 21), or perhaps more correctly, the diaphragm will form about them after they have become separated. The apex of the diverticulum may become obliterated, so releasing the accessory structure from the vertebrae. Presumably, the diverticulum may close off at X, its original point of origin from the intestinal tube; this would explain those duplications which do not communicate with the intestine; such closure would not preclude the duplication being carried caudally, as there is intermingling of the fibres of the circular muscle coats of the two channels.

Colon and Rectum

Duplication of the colon is less common than duplications of the small bowel. Ravitch (1953) was able to find only 20 cases in the literature. Though the condition is essentially the same as, or nearly related to, more proximal duplications, there are significant differences, possibly the more caudal abnormalities are determined a little later in embryonic life. It is also possible that comparable degrees of doubling of the cephalic end of the embryo would lead to its non-survival.

When duplication of the colon is associated with a spinal abnormality, the lumbo-sacral spine is affected; spina bifida, hemivertebrae and partial or complete double spinal column from L.2,

TABLE 5

TUBULAR DUPLICATIONS

No.	Age	Sex	Presentation	Location	Length in cm.	Epithelial Lining	Spine	Other Abnormalities	Procedure and Result
3	3½ months	M	Bleeding and perforation	Jejunum	60	Gastric	Hemivertebrae D3 and 4	2 posterior mediastinal cysts	1. Perforation closed 2. Resection six weeks later, recovery
4	5½ years	M	Anemia and diarrhea perforation	Ileum	30	Gastric			Resection, died peritonitis
5	10 years	F	Pain, palpable abdominal swelling	Ileum	25	Gastric and small intestinal	Fusion of D3 and 4		Resection, recovery
9	2 days (weight 4 lb. 10 oz.)	M	Intestinal obstruction volvulus, perforation meconium peritonitis	Small bowel	10	Bowel necrotic	No comment		Resection two-thirds small bowel (gangrenous), death
12	6½ months	M	Bleeding	Ileum	20	Gastric	No comment	Mesenteric cyst	Resection, recovery
12a	5 weeks	M	Intestinal obstruction volvulus	Ileum	20	Gastric	Dorsal hemivertebrae and spina bifida	Meckel's diverticulum	Resection (gangrenous bowel), death
13	11 days	F	Intestinal obstruction	Ileum	60	Gastric	No comment		Resection, recovery
17	13 years	F	Epigastric pain, healed perforation	Duodenum	5	Gastric	Normal	Double gall bladder	Resection of duplication and gall bladder, recovery
18	8 days	F	Perforation	Ileum	16	Gastric and small intestinal	No comment	Posterior cyst	Resection, died peritonitis
33	10 weeks	M	Bleeding and perforation	Ileum	45	Gastric and duodenal	Hemivertebrae and spina bifida occulta D2, 3, 4, 5 and 6	Dorsal dermal sinus communicating with dura	Resection, recovery
34	1½ years	M	Pain and vomiting	Ileum	15	Various alimentary, including gastric	Mid-dorsal hemivertebrae	Mesenteric cyst, ectopia vesicae	Resection, recovery

caudally, have been described. There is also a high incidence of doubling of the genital and urinary organs. The two colons communicate by their proximal extremities, though a second distal communication is sometimes present. There may be two separate ani, either placed side by side or one in front of the other; alternatively one or both colons end blindly, or by fistulous communications to urethra or posterior vaginal fornix. There may be a true doubling of the entire colon, so that it is impossible to decide which channel is the accessory structure. Another difference from more proximal tubular duplications, is that double colons are invariably lined by colonic mucosa, ectopic epithelium has not been described.

In a few instances double colons have been associated with double terminal ileum, two cæcums and two appendices. In our eight tubular duplications of the ileum, the accessory structure never involved the terminal ileum. It seems possible that duplications associated with cervico-dorsal or upper dorsal spinal abnormalities, which are determined by an error in development towards the cephalic end of the embryo, may extend distally as far as, but no further than, the original site of the omphalomesenteric duct, whilst those accessory structures which are associated with lumbo-sacral spinal abnormalities, and in which the embryonic error is towards the hind end of the embryo, may reach proximally as far as, but no further than, the same point.

Group B duplication of the colon is very poorly represented in our series, which includes only two rather atypical instances of the abnormality (37, 42).

A newborn male infant (42) had vesico-intestinal fissure, diphallus and a sacral meningocele. The ileum and cæcum opened on to the ventral surface between the two elements of the exposed bladder. At operation it was found that there was a single cæcum, but that the colon, which was only $2\frac{1}{2}$ in. in length, was double, both channels ending blindly above the pelvic floor. The double colon and the cæcum were excised and the ileum brought down to the perineum. The baby died of pyelonephritis at $2\frac{1}{2}$ months of age. The second case (37) was a newborn female infant, with a radiologically normal spine; a segment of bowel, lined by typical rectal mucosa was applied to the vulva. One end was open to the surface, the other was blind; the structure was attached to the vagina and rectum, but did not communicate with either.

Accessory intestinal segments which lie externally are very uncommon, those described in the literature, unlike that of this case, have usually been covered with skin.

A Primitive Type of Mesentery or Non-Rotation of the Mid-Gut Loop

Abnormal rotation is common in both Group A and B abnormalities. A large cyst could hamper return of the intestines from the physiological hernia in the base of the body stalk, or simply by its bulk, impede rotation. Cases in which there is, or has been, a long duplication, part of which is extra-mesenteric (prototype case No. 21), must have non-rotation, as the duplication acts as a tether, and prevents rotation.

In this series abnormalities of rotation are mentioned in a high proportion of autopsy reports, whilst in operation notes, references to the abnormality are fewer.

Intra-Spinal Entodermal Tissue

It is to be expected that as remnants of the original entodermal diverticulum anterior to the vertebræ are not uncommon, more dorsal situated entodermal tissue would occasionally be found, and lately it has been realized that these are by no means rare. It is, however, unusual for there to be continuity between accessory structures anterior and those posterior to the vertebral bodies. The growth of the rigid structures of the spinal column tends to obliterate the portion of the diverticulum which originally separated the two notochordal centres.

Seven cases (21, 38, 28, 36, 44, 47, 41) with entodermal tissue posterior to the vertebral bodies are included in this series, these abnormalities at least cannot be called duplications and the term 'Posterior Enteric Remnants' (Bentley, 1960) is entirely appropriate.

Four children (21, 38, 28, 36) had accessory structures both anterior and posterior to the vertebral bodies and in two of these (21, 38) there was continuity of entodermal tissue between hemivertebræ; in two cases no evidence of the original communication was found.

Three children (44, 41, 47) had intra-spinal entodermal tissue, but no evidence of accessory structures anterior to the vertebræ; however, only one of these came to autopsy.

Intra-Spinal Lesions with Accessory Structures Anterior to the Vertebræ

The upper end of the thoracico-abdominal duplication of the prototype case (21) disappeared into a dura-lined cleft in a wedge of cartilage which divided the body of the second dorsal vertebra into hemivertebræ (Diagram 6 (1)). The stump of the duplication had a central lumen, lined by squamous epithelium; ventrally or superiorly the wall was composed by connective tissue, blood vessels, and scattered bundles of smooth muscle, dorsally or inferiorly the wall was of

nervous tissue continuous with that of the spinal cord. The hollow structure ran into the central canal of the cord where it ended blindly, blocking the canal but not communicating with it.

A 5-day-old infant (38) had vomited for two days. He continued to vomit and gradually developed quadriplegia, muscular wasting and purely diaphragmatic respiration. X-ray films showed lower cervical hemivertebrae, lumbar puncture gave a sterile fluid, a subdural tap showed xanthochromia and ventriculography showed dilatation of the ventricles. The child died at 2½ months of age.

At autopsy (Diagram 6 (2)) there was a cyst in the posterior mediastinum with a thick stalk which passed between hemivertebrae to communicate with a cyst lying in an expanded spinal canal: it compressed the cord which lay dorsal to it. The subarachnoid space above the lesion was obliterated by blood clot. The portion of the cyst anterior to the vertebrae was lined by alimentary epithelium of various types and a large wedge of pancreatic tissue; the walls were of smooth muscle with ganglia. The intraspinal portion had largely fibrous tissue walls: the lining showed traces of alimentary epithelium. It was considered likely that the bleeding into the subarachnoid space had followed erosion of blood vessels by pancreatic secretion (Fig. 12).

A child (28), aged 10 days, died following operation for a large meningo-myelocoele. At autopsy he had a thick-walled cyst applied to the duodenum, lined with intestinal and pyloric gastric epithelium, and a cyst in the posterior mediastinum lined with simple columnar and pyloric gastric epithelium. There was no evidence of a dorsal track on coronal section of the vertebrae, but a hollow cord, the lumen lined by oesophageal epithelium, was attached to the dorsal surface of a vertebral body (Fig. 13 and Diagram 6 (3)); it ran dorsally and caudally, almost dividing the spinal cord into two parts; the structure fused with the tissue of the cord as the latter opened out into the myelocoele.

A child (36) died on the day of birth following closure of an exomphalos and excision of an extensive lower dorsal and upper lumbar meningo-myelocoele. At autopsy a posterior mediastinal cyst was found, lined by a single layer of columnar mucus-secreting cells (Diagram 6 (4)). There was also an intraspinal cyst which expanded the spinal cord tissue, the lining was almost unrecognizable, but was probably of entodermal origin.

Cases with Spinal Lesions, but no Evidence of Accessory Structures, Anterior to the Vertebrae

A baby (44) became irritable and pyrexial at 48

FIG. 12.—Photograph of artist's water colour. Case No. 38. Posterior mediastinal cyst and intra-spinal cyst communicating between hemivertebrae. Pancreatic tissue in upper anterior wall of posterior mediastinal cyst.

hours of age; she resented flexion of the spine, especially of the neck and the fontanelle was flat, but lumbar puncture showed xanthochromia. She developed paralysis of the right 7th and 12th cranial nerves and of the arms; the legs were hypotonic, later nystagmus to the right, dysphagia and hiccup developed. X-ray showed widening of the pedicles of the lower cervical vertebrae and abnormal arches. The baby died at nine weeks. Autopsy disclosed a cyst within the spinal cord tissue (Fig. 14 and Diagram 7 (1)) and attached to the dorsal surface of the flattened cord there was a crescent of intestinal, squamous and respiratory epithelium. In the right antero-lateral wall of the cyst was a solid nodule, 0.7 cm., in diameter, with a rugose surface; on section it was found to have a

FIG. 13.—Photograph of artist's water colour. Spinal cord held in forceps—accessory structure between cord and vertebral body. Case No. 28.

FIG. 14.—Photograph of artist's water colour. Cyst within spinal cord before and after removal of part of the cyst wall. Case No. 44.

central core of vessels and smooth muscle in two layers, and Auerbach's cells, and to be surrounded by epithelium, principally gastric; it resembled intestine turned inside out.

The last two cases both presented with lower motor neurone lesions and stiffness of the back (Diagram 7 (2) and (3)). One (47), a boy of two years, had a healed meningocele and sacral spina

bifida, the other, a girl (41) of two and a half years, had a healed cutaneous sinus in the mid-dorsal region. In both a soft tissue tumour was shown by air myelography. Laminectomy revealed cysts lying posterior to the cord and compressing it. The cyst of the boy was lined by columnar epithelium, ciliated in some areas, and the wall contained smooth muscle. In the girl the cyst was lined by ciliated cuboidal, transitional and

squamous epithelium; there was an incomplete layer of smooth muscle in its wall.

Both these children did well after removal of their cysts.

Forty-four cases have been included in this review, many of which have appeared under more than one heading. Two further cases had undoubted duplications, but their case sheets do not include the requisite data for their inclusion.

I acknowledge, the kindness of Mr. P. P. Rickham, Mr. J. H. Johnston and other colleagues, who have allowed me to review their cases, my indebtedness to Dr. Norah M. Walker, senior radiologist to the Royal Liverpool Children's Hospital and Alder Hey Children's Hospital, and to Dr. Edward Hall and Dr. J. S. Elwood, pathologists to the two hospitals. Dr. Hall has given me much help and advice, and to him I am especially grateful.

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