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INTRATHORACIC DUPLICATIONS OF THE BOWEL

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Intrathoracic duplications of the bowel, though rare, are important on account of the serious complications which they may cause.

Two main types of intrathoracic duplication of the bowel are recognized, those which are completely within the thorax, and the more uncommon group which are subdiaphragmatic in their attachment to the bowel and appear to herniate into the thoracic cavity.

AETIOLOGY

Several theories have been put forward to explain their origin, two of which are worthy of consideration. The first was formulated by Lewis and Thyng (1908) and suggested that evaginations of the foregut are partially or completely sequestrated off and give rise either to diverticula or to cysts. The other view enunciated by Keith (1933) points out that in the early weeks of intra-uterine life the epithelium of the gut proliferates to such an extent that the lumen is occluded. Later vacuoles appear between the cells and re-canalization takes place. Should this vacuolation take place in an irregular manner many of the forms of atresia, cyst formation, and duplication can be accounted for. This theory has been further elaborated by Bremer (1944) and appears to explain the abnormalities arising within the thorax, though the former conception seems more probable for the rare lesions arising below the diaphragm. Fallon (1953) points out that both types of duplication are not infrequently associated with abnormalities of the cervical vertebrae. He suggests that such cases arise in very early intra-uterine life by a local disturbance of growth which affects adjacent areas of ectoderm and entoderm.

PATHOLOGY

The main features in the pathology of the first type of duplications, whether arising within the thorax or not, have been pointed out by Ladd and Gross (1941) and are as follows: (1) Duplications resemble, and are usually contiguous with, the normal alimentary tract of the region in which they are situated. (2) The musculature and blood supply of the duplication and the adjacent normal gut are usually shared and may make resection difficult. (3) The duplication is lined by epithelium which is similar to that found in some part of the alimentary tract or an immediate derivative of the epithelium of primitive gut. In other words, the epithelium lining a duplication is either alimentary or respiratory in type but not necessarily similar to that of the adjacent gut. These duplications may or may not communicate with the gut, but if they do there may be more than one communication.

The second type of duplication arises from the duodenum or jejunum and passes up the posterior abdominal wall and through the diaphragm either into the mediastinum as in Gross's three cases (Gross, Neuhauser, and Longino, 1950) or through a hiatus which directly connects the pleural and peritoneal cavities. This type resembles a giant diverticulum of the bowel.

Secondary pathological changes may be produced in either of these main types or in their surroundings by pressure and inflammation.

SYMPTOMATOLOGY

The sex incidence is roughly equal in both groups. Gross, Holcomb, and Farber (1952) mention some 16 cases; the average age of onset of symptoms was 16 months, but it must be remembered that this series is from the Harvard Medical School Paediatric Service. It appears from a quick review of the literature, which, however, does not include findings of symptomless cases at mass radiography, that symptoms appear early in life.

Large duplications produce pressure symptoms soon by causing respiratory distress and dysphagia. The former is shown by grunting respirations, cough, pulmonary collapse, and infection, and the latter by froth in the pharynx, regurgitation, and vomiting. An acute onset or acutely recurring episodes may be produced by a rapidly expanding lesion such as a duplication lined with gastric mucosa containing pepsin and hydrochloric-acid-

secreting cells which lead to peptic ulceration and diffuse inflammation. Such cases present as dysphagia and pain and sometimes as haematemesis or haemoptysis due to spread of the inflammation to the oesophagus or lung, and even as perforation of the duplication or sloughing of its wall together with that of the oesophagus. Symptoms are not so acute, according to Valle and White (1946), in duplications which do not contain gastric mucosa or if gastric mucosa should be present in which there are no pepsin and hydrochloric-acid-secreting cells. These milder symptoms, due to the effects of a slowly expanding intrathoracic lesion, are commoner than the acute catastrophes mentioned above.

The diverticulum type of duplication which has herniated through the diaphragm presents in two main ways (Gross, Neuhauser, and Longino, 1950; Gross, Holcomb, and Farber, 1952). (1) As intermittent dyspnoea, cyanosis, and dysphagia which arise from the variable distension of the herniated gut pressing on the chest contents. Recurring attacks of "bronchitis" are common. (2) As inflammation from peptic ulceration which causes chest pain and sometimes severe haemorrhages into the lumen of the oesophagus and anaemia.

Diagnosis

The diagnosis depends on considering its possibility when the somewhat bizarre symptoms outlined above are found.

The diagnosis can and should be made by radiological examination. The type I duplication usually shows as a shadow in the right lower chest displacing the oesophagus to the left and causing an indentation of its right border which is easily shown by swallowing a contrast medium. This picture is said to be diagnostic by Valle and White (1946). However, it should be pointed out that the position of the duplication and of the oesophagus may be reversed, or the duplication may be higher up in the thorax. Whatever the site within the chest, the association of a tumour with a long and fairly even indentation of the oesophagus is suggestive.

When haematemesis is the presenting feature peptic ulceration from reflux oesophagitis due to chalasia of the cardia or a sliding oesophageal hiatus hernia must be excluded, as well as a continuation upwards from the stomach into the oesophagus of pepsin- and hydrochloric-acid-secreting mucosa. Here, as well as biopsy and estimation of the *p*H of the mucosal secretion, oesophagoscopy is of value, excluding by direct vision foreign bodies and oesophageal varices. Bronchoscopy may show displacement by the lesion and verify the site of the rarely occurring haemoptysis (Figs. 1, 2, and 3).

Type II duplication presents radiologically as an intrathoracic inflammation with fluid levels which are usually mistaken for lung abscesses and empyema, though the changing amounts and positions of the fluid may suggest an alternative diagnosis. No record has been found of radiographs showing filling of the diverticulum with contrast medium (Figs. 4 and 5). Finally, as pointed out by Fallon (1953), the association of such intrathoracic findings together with abnormalities of the cervical vertebrae are very suggestive of the diagnosis of duplication.

In both the recorded cases given below a firm pre-operative diagnosis should have been made.

Treatment

The general principles of the treatment of any intrathoracic mass apply to these cases. These are exploration and excision, if possible, in order to prevent pressure symptoms and inflammatory changes, or, in cases of tumour, to prevent local spread or metastases. Exploration is still indicated, unless the condition is obviously hopeless, to try and alleviate such complications should they unfortunately have arisen.

Considerable care is needed in the resection of duplications because of the association of their blood supply and the continuity of their muscle coat with that of the adjacent oesophagus.

Case Reports

Type I Duplication.—The following case history (No. 267453) of a woman admitted to the City General Hospital, Sheffield, is illustrative.

K. G., a woman aged 44 years, a housewife, was referred to me as a "pick up" by mass radiography. The history was that for two years she had suffered from a slight morning cough and dyspnoea on going upstairs. Also she had felt a "cold patch" in the chest wall posteriorly just below the inferior angle of the right scapula for the same period. There was no other relevant history. The patient appeared to be a fit, healthy woman. She was admitted to hospital on February 21, 1952, and the general clinical examination, including bronchoscopy and routine blood and urine examinations, revealed no abnormality.

A radiological report dated February 23 stated that a cystic-looking shadow was present at the right base adjoining the postero-lateral cardiac border. It appeared on screening to be intimately connected

with the latter, and to pulsate, but that might be transmitted pulsation. The lung fields were otherwise clear (Figs. 1 and 2).

A barium swallow showed a slight pressure by the mass which was approximately 2 in. in height and 2½ in. in depth. It was not thought to be a hernia and the diagnosis was most probably a pericardial or pleural cyst, but a neurofibroma or dermoid was a possibility (Fig. 3).

A right-sided bronchogram was normal. The cervical vertebrae appeared normal. At operation on March 10 the anaesthetic was pentothal and tubarine and intratracheal gas and oxygen. The chest was opened by a right postero-lateral incision with resection of the sixth rib. No adhesions were present, and a cystic swelling the size of a tennis ball was situated under the mediastinal pleura just above the diaphragm. The cyst was deliberately opened to facilitate its dissection from the oesophagus to which it was intimately adherent, as the muscle coat of the oesophagus was thinned out over the cyst although the mucosa and submucosa were intact. The oesophageal wall was covered with pleura, and the pleural cavity drained. Convalescence was uneventful, and the patient was discharged on March 15. She was seen as an out-patient on September 24, 1952, and on September 26, 1953, symptomless, and the radiograph of the chest and a barium swallow were normal.

Pathological Report.—Dr. H. E. Harding reported that the lining of the cyst was respiratory epithelium,

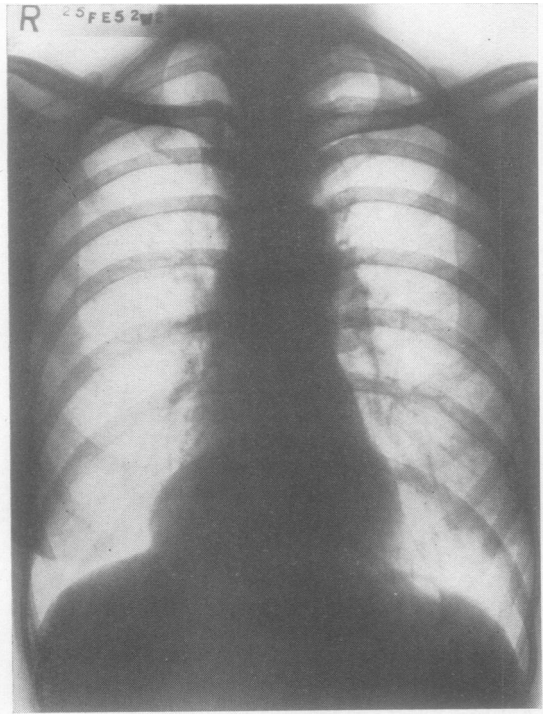

FIG. 1.—Typical postero-anterior view of a type I duplication.

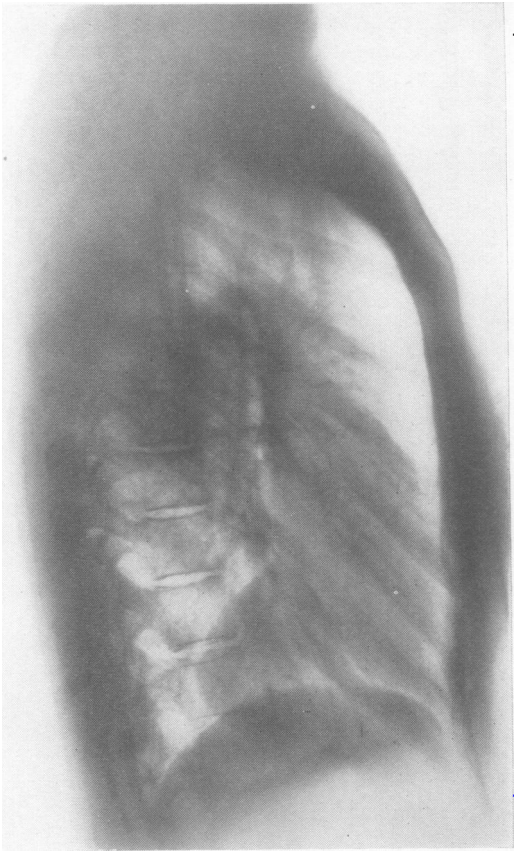

FIG. 2.—Typical right lateral view of a type I duplication.

and the gland in muscle was salivary. Dr. A. J. N. Warrack said further that the cyst was lined with a ciliated type of epithelium. There were a few cells in the muscle which could not be definitely identified. The picture suggested that it was ectopic glandular tissue.

TYPE II DUPLICATION.—This case (No. 52/7492), admitted to the City General Hospital, Sheffield, is an example of the second type.

P. C., a boy aged 5 months (birth weight 6 lb. 2 oz., three weeks premature), developed right-rided pneumonia when 6 weeks old, with vomiting immediately after feeds. The child improved, but still had signs of bronchitis until May 24, 1952, when it became much worse and signs of consolidation appeared in the right chest.

The child was admitted to the Doncaster Royal Infirmary under Dr. A. L. Hawkins on May 31. He was shocked, irritable, crying, cyanosed, and vomiting. He was placed in an oxygen tent and given large doses of antibiotics. During June and July his condition varied but vomiting subsided. Radiographs suggested a hydropneumothorax, and later fluid level

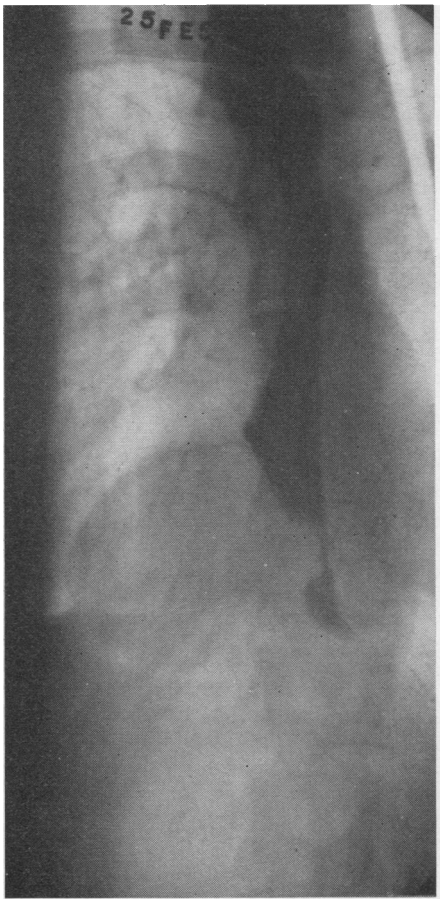

FIG. 3.—Typical result of a barium swallow in a type I duplication.

appeared in the upper part of the chest. The oesophagus appeared normal on barium swallow. Repeated aspiration did not obtain any fluid (Figs. 4 and 5).

The child was admitted to the City General Hospital, Sheffield, on July 27. Radiographs showed displacement of the mediastinum to the left and a right-sided pleural effusion. In addition there was the radiological appearance of a spina bifida in the lower cervical region. The effusion was aspirated and a little straw-coloured fluid obtained which showed numerous pus cells and cell debris and scanty Gram-positive cocci on direct examination. The culture was sterile. The child's general condition again improved until August 29, when all the previous symptoms returned. The child was seen during my absence by several colleagues, and on my return exploration was agreed upon and carried out on September 9 under intratracheal cyclopropane and oxygen anaesthesia (Dr. J. Johnston). The right chest was opened after resection of the seventh rib. A large, thick-walled cyst presented, and, as it did not seem possible to explore the chest, the cyst was opened and appeared to be of a gastric nature. The apex of the cyst was freed from the subclavian vessels with some difficulty, and it was traced to a pedicle which came through the posterior part of the diaphragm behind the inferior vena cava. The pedicle appeared to be normal small intestine, though the main blood supply of the cyst came in branches from a large vessel which ran down into the mediastinum by the oesophagus. The pedicle was cut and sutured and the cyst excised. The lung expanded immediately in spite of having been compressed antero-medially by the cyst. No attempt was made to close the hiatus. The chest was closed without drainage. The operation took one hour.

Two and a half hours later the child went into convulsions and died four and a half hours after the end of the operation.

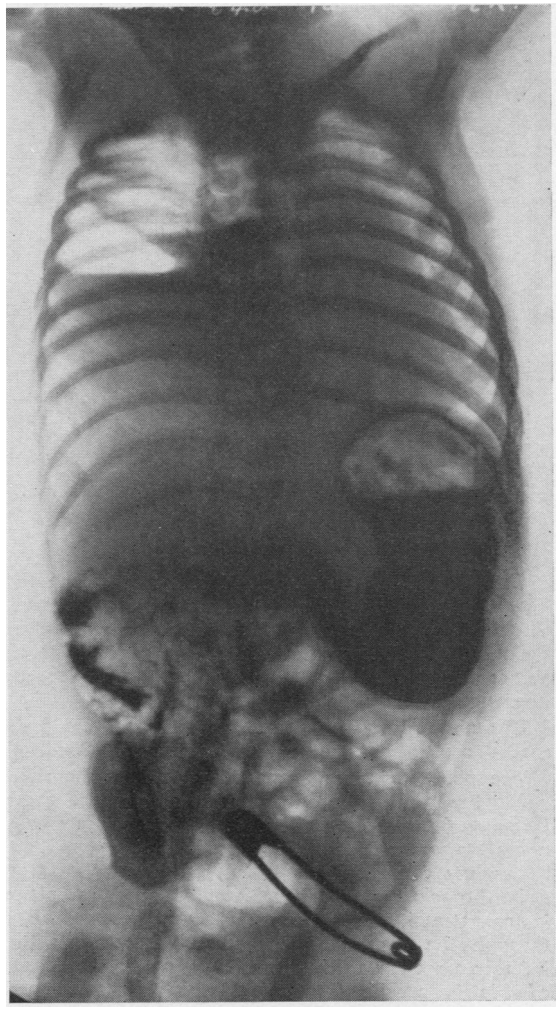

FIG. 4.—Postero-anterior view of a type II duplication.

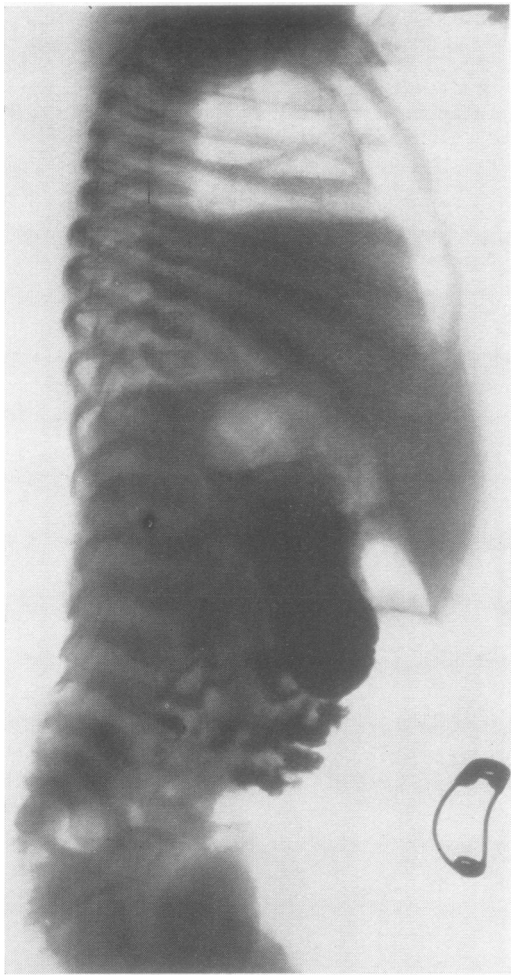

FIG. 5.—Right lateral view of a type II duplication.

Dr. J. L. Emery performed a necropsy at the City General Hospital at 10 a.m. on September 10. The body was that of a thin, small, rather wasted male child. There were transfusion sites at both ankles and at the left ante-cubital fossa, and a recent operation site over the right chest.

The abdominal cavity was normal, and the pericardial cavity contained a small amount of fluid.

The pleural cavities appeared normal. The right cavity contained a certain amount of air, and there was a certain amount of blood. There was a recent operation wound and the oesophagus appeared to be unduly dilated, if anything, ballooning into the right thoracic cavity. There was an aperture in the lower extremity of the pleural cavity passing down through the diaphragm immediately medial to the inferior vena cava, the vena cava passing in front of it in the middle of the canal, which communicated directly with the peritoneal cavity. In other words, the pleural cavities and the peritoneal cavity were continuous.

The nose contained some mucus, as did the larynx, trachea, and bronchi, with a small amount of milky curds, presumably inhaled.

The left lung showed small areas of collapse, but most of the lung was distended and did not collapse on opening the chest, apparently because of the inhaled material. The right lung was almost completely collapsed, and contained small firm nodules, apparently bronchopneumonic, which were not then dissected.

The heart externally showed no gross abnormality. On cross section the only abnormality suggested was some small clear vegetation along the mitral valve—possibly a minor congenital abnormality. The great vessels showed no gross anatomical distribution, but the aorta in the region of the abnormal tumour contained an abnormal number of small vessels. Three of these had been ligated during operation. The lowest had a small fibrinous adherent clot on the inner surface, and beside this a small slit in the intima, but not penetrating the other layers of the aorta.

The tongue was normal. The oesophagus showed no gross abnormality. The stomach appeared normal and in its normal situation. The cardia, pylorus, and duodenum appeared normal. Eight centimetres from the duodenum there was a small pouch, approximately 2 c.mm. long, which had been ligatured, and continued with the intestine, which, at this point, appeared as if it were two, the pouch being on the mesenteric side. The rest of the intestine close to the appendix appeared normal. The large bowel appeared normal and contained normal faeces. The mesentery to the whole of the large and small intestine was extremely short in its attachment to the posterior wall, the attachment being entirely in the mid-line and measuring approximately 3 c.mm. The mesentery was long and tortuous, and the appendix and caecum were situated almost in the mid-point of the abdominal cavity. The large bowel was largely on the right side. No other abnormality was found.

The mass removed from the mediastinum consisted of an irregular twisted mass of intestinal tissue superficially looking rather like stomach having a neck approximately 1.5 c.mm. in diameter which penetrated through the diaphragm. The mass appeared to be completely covered by a membrane which was thicker and rather more opaque than either the pleural or the peritoneal, and might possibly be a combined layer of both pleura and peritoneum. Many blood vessels and lymph nodes were attached to this tissue which appeared to have been lying within the posterior mediastinum and projected into the pleural cavities behind the lung (Figs. 6 and 7).

The primary lesion in this child was a gross congenital abnormality of the intestinal tract. The immediate cause of death was probably inhalation of regurgitated stomach contents. The convulsion and cyanosis were probably precipitated by some bio-

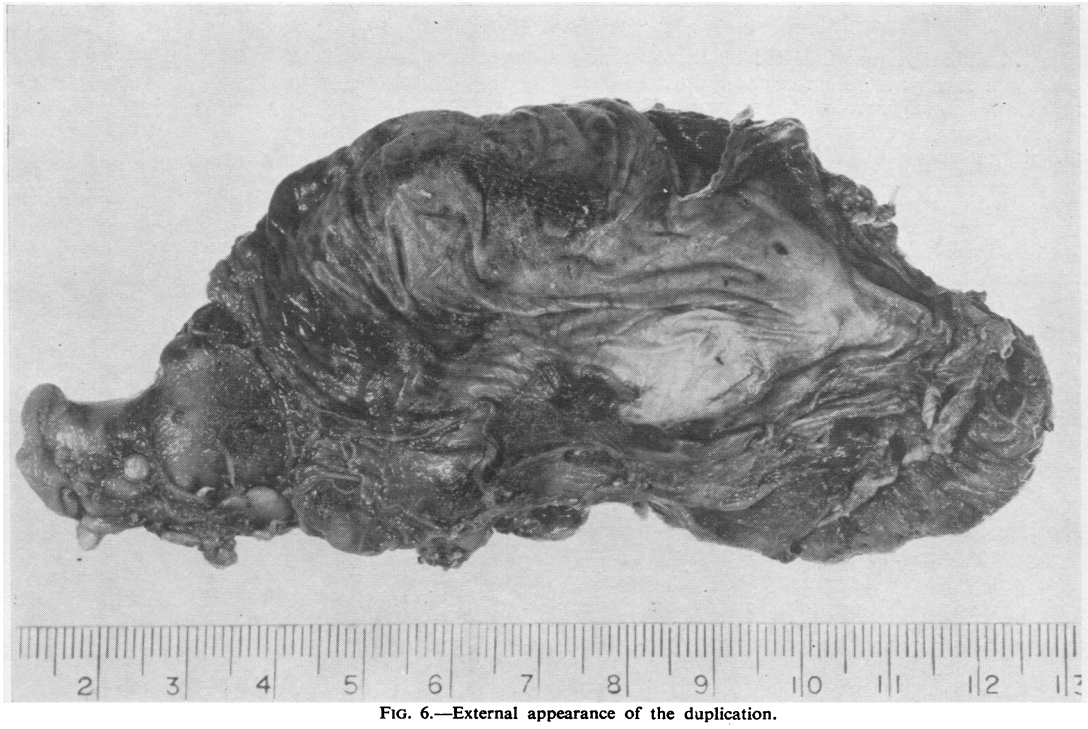

FIG. 6.—External appearance of the duplication.

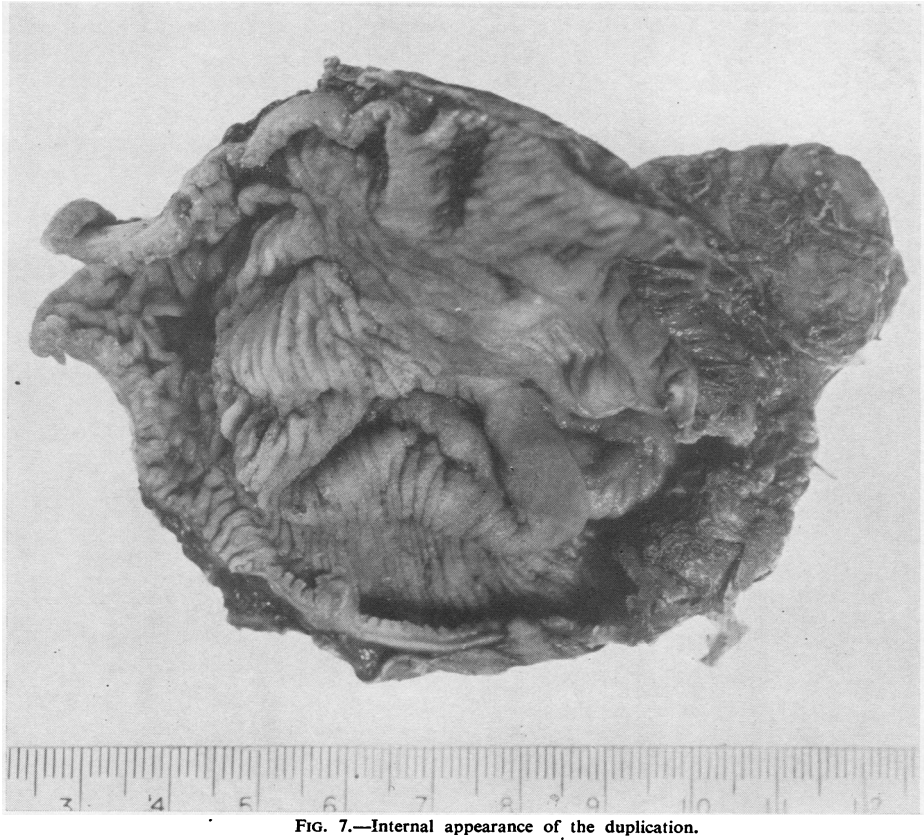

FIG. 7.—Internal appearance of the duplication.

chemical upset in its turn initiated by the operation. There was a chronic infection of the right lung.

All viscera, which were examined, were normal.

The blood vessels of the tumour appeared to arise in the mediastinum and not from the abdomen. The tissue and epithelium near the tip of the sac was smoother than that near the neck, and the muscular layers appeared very prominent near the tip.

Sections from the neck of the sac and transverse sections from the mid part of the sac showed gastric mucosa with fairly prominent lymph tissue.

The tip of the sac and the fragments near the tip showed a stratified epithelium and structure fairly typical of oesophagus. Beneath the epithelium were small ducts lined with stratified epithelium such as are seen in bronchial remnants. There appeared to be some ulceration of the oesophageal epithelium. Cells in the gastric mucosa appeared to be actively secreting.

The immediate cause of death in this child was of a respiratory nature due to multiple small areas of collapse, possibly due to anaesthetic, possibly due to inhalation. The primary anatomical lesion appeared to be the reduplication of the oesophagus and stomach with a foregut remnant. There appeared to be some ulceration of the squamous (oesophageal) epithelium within the diverticulum.

Summary

The types, aetiology, pathology, diagnosis, and treatment of intrathoracic duplications of the bowel are discussed with illustrative case reports.

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