C O N G E N I T A L O C C L U S I O N S

of the

O E S O P H A G U S and L E S S E R B O W E L

In the Human Subject

by

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P R E F A C E.

The following research was undertaken with the object of attempting to elucidate by dissection and microscopic examination the causation of Congenital Occlusions of the Oesophagus and Lesser Bowel, in the Human Subject.

The specimens on which the research was based had come under my observation during the past few years. In several cases other malformations co-existed; but only those which involved the above-mentioned portions of the alimentary canal are here described, and each bears a reference to the case in which it occurred.

The Table of Contents not only enumerates the individual malformations described, but indicates the general scheme of the paper.

The Histological Examination of the specimens was carried out in the Embryology Laboratory of the University, and I wish here to record my indebtedness to Dr.James F.Gemmill for much friendly help and advice.

Geo.H.Edington.

January, 1913.

C O N T E N T S

CONGENITAL OCCLUSIONS OF THE OESOPHAGUS AND LESSER BOWEL.

INTRODUCTORY.

The following series of specimens illustrates some of the congenital malformations of the oesophagus and lesser bowel.

I have arranged them in three classes according as they affect A.Oesophagus: B. Duodenum: and C. Ileum.

Following the description of the specimen or specimens in each of these classes there is a review of the principal literature on the subject. This is succeeded by remarks on the specimen or specimens under consideration. Each section is followed by a list of bibliographical references.

Summaries of the cases from which the specimens were obtained are given in an Appendix. In these summaries each malformation described in the paper is underlined, and the reference page given.

The illustrations referred to in the paper are bound separately.

A. OESOPHAGUS.

Malformation of the Tracheo-oesophageal Septum.

Two specimens of this malformation occur in my series of cases (Cases I. & VIII.)

1. (Case I.). There was complete atresia of the oesophagus. The upper segment, continuous with the pharynx, terminated below in a blind pouch, the lower end of which reached to the level of the 7th.ring of the trachea. The lower segment of the oesophagus on being traced upwards from its cardiac end became markedly contracted as it neared the level of the bifurcation of the trachea. Almost immediately above this level (19th.ring) it joined the trachea, its posterior wall forming the posterior wall of that tube, and continuing upwards as a muscular layer ventral to the ventral wall of the blind upper segment. There was no appearance of any band joining the segments of the oesophagus, but some thin fascia stretched continuously over both.

The appearance of the parts is shown in fig.1, which represents the trachea and upper segment of the oesophagus split antero-posteriorly in the median line, and laid open.

2. (Case VIII.) There was complete atresia of the oesophagus. The pharynx terminated in the blind, pouched upper segment (fig. 2), which extended down to the level of the 12th.ring of the trachea, 1.5cm. below the cricoid cartilage. The upper segment appeared dilated - measuring 9mm. across - as compared with the lower segment, which was only 4 mm. across. On tracing it up from the stomach the lower segment of the oesophagus was found to join the trachea at the level of the 19th. cartilaginous ring. Above this level there was a common tracheo-oesophageal tube, the posterior wall of which was continuous with the original posterior wall of the lower segment of the oesophagus. There were no communicating bundles passing up from it to the blind end of the upper segment.

HISTOLOGY. Antero-posterior longitudinal sections showed that the wall of the upper segment was substantial, consisting of outer longitudinal and inner circular muscle-bundles, and a well-marked layer of fibrous tissue, and was lined by stratified squamous epithelium. The longitudinal fibres showed striation: the circular fibres having been cut transversely, striation was not seen in them. The posterior wall of the tracheo-oesophageal tube was well supplied with muscle-fibres: for the most part these were smooth, but some striated fibres were met with here and there. Mucous glands were numerous, some of them lying external to the muscle-fibres. The wall was only about half the thickness of that of the upper segment. In the portion of tracheal wall in contact with the ventral wall of the pouch there were a few scattered muscle-bundles. Below this level the arrangement of the muscle-fibres corresponded pretty closely with that of the blind upper segment, viz:-

external longitudinal, and internal circular. As the lower part of the section was approached, however, there were in addition longitudinal bundles present between the circular muscle and the mucous membrane. These bundles varied in thickness and could not be looked upon as normal muscularia mucosae. At the junction of the ventral wall of the lower segment of the oesophagus with the trachea there was a short septum, the upper edge of which was thin, and on a level with the 19th.cartilaginous ring.

The septum was a combination of trachea and oesophagus. It contained cartilage, on the anterior surface of which was tracheal mucous membrane, and sub-mucosa. Posterior to the cartilage were numerous transverse sections of muscle-bundles. These were found as high as the free edge of the septum. Between them and the oesophageal mucous membrane were longitudinal bundles of muscle-tissue, of no great bulk. In the region of the free edge were numerous mucous glands, between the circular muscle-bundles (oesophageal) and the tracheal surface of the septum.

Epithelium of lower segment of the oesophagus. The posterior wall was lined by columnar ciliated epithelium, which, a little below the level of the free edge of the septum, gave place to stratified squamous non-ciliated cells. The ventral wall of the oesophagus, where it formed the oesophageal surface of the septum was covered by columnar ciliated epithelium.

R E M A R K S.

On comparing these two specimens they are found to agree in several points. (1) The upper segment of the oesophagus ends as a blind sac which projects downwards beyond the level of the larynx. (2) There is a free communication between the lower

segment and the trachea, close above the bifurcation of the latter. (3) The absence of connecting bands between the upper and lower segments. (4) The foetuses from which they were obtained present other malformations (see Case-Summaries in Appendix).

The specimens differ from one another in the position of the tracheo-oesophageal fistula, and in the nature of their associated malformations. Thus, in Case I. the lower segment traced up from below is found to become incorporated with the trachea at or just above its bifurcation. In Case VIII. there is a short septum between the two tubes, which septum extends upwards to the level of the 19th. tracheal ring. As regards associated malformations, Case I. was the subject of transposition of the thoracic and abdominal viscera, septal occlusion of the duodenum, and atresia ani. In Case VIII. there was malformation of the recto-vesical region - atresia recti, stenosis of urethra, abnormality of left vas deferens, and microscopic communication between the rectum and the bladder.

On referring to the literature it is therein stated that congenital malformations of the oesophagus are not very frequent. A considerable number of examples have, however, been published since 1838, at which time SCHOLLER (cited by Giffhorn, v. infra) found only one case reported. Scholler framed a classification of oesophageal anomalies, and subsequent writers have done the same; but observation has shown that the form which occurs in a great preponderance of these cases is that in which there is complete interruption of the oesophagus, the upper segment ending blindly and the lower communicating with the trachea.

MACKENZIE[1] writes:- "Deficiency of a part of the oesophagus, generally affecting the middle third, together with an abnormal communication between the gullet and the trachea or one of the bronchi, is the most common deformity, and though met with in monsters and still-born children, is most frequent in infants who are born alive, but survive only a few days". The opening of the lower segment as viewed from the trachea is generally a small aperture, sometimes oval, round, or crescentic with its concavity downwards. "The two separate portions are generally connected by a small band of muscular or tendinous fibres" (p.223). Other deformities frequently co-exist. Mackenzie gives a long list of references. He believes that by far the larger number of malformations of the oesophagus are due either to disease or to a "displacement of formative material at a very early period of embryonic life..... some slight morbid deflection of the normal".

SHATTOCK[2] reported in 1890 a case to the Pathological Society, and referred to four cases in the Museum of the Royal College of Surgeons, and to three cases reported in the Central-blatt für Laryngologie.

BALLANTYNE[3] describes as the common form "termination of the oesophagus in a cul-de-sac, the lower rudiment of the canal communicating with the trachea or bronchi". From the lower part of the oesophagus a "small opening establishes a communication with the trachea" (loc.cit. p.463). Associated malformations are uncommon. He gives a number of references to the literature, and an interesting explanation of the fact that meconium is sometimes passed in these cases, possibly from the foetus in utero swallowing liquor amnii.

KEITH[4] in 1906 reported three cases and referred to a fourth. His communication was entitled Malformation of the Tracheo-Oesophageal Septum, and he recorded the cases "not...because of their rarity...but because in the first place the nature of these malformations is not generally understood by those who describe them in the medical press: secondly, because in all three cases there was present a right aortic arch or its representative, "and thirdly, because recorded cases in literature are usually unaccompanied by explanatory illustrations.

He explains (l.c. p.54) the malformations as evidently formed by the "lateral tracheo-oesophageal ridges and fold" proceeding "obliquely backward (?downwards) and dorsalwards so as to meet on the dorsal wall of the foregut". If, however, there is a partition between the lower segment of the oesophagus and the trachea - the common form of the malformation - the tracheo-oesophageal septum must have been formed in two parts. The posterior (lower) part forms the partition: the anterior (upper) part, obliquely placed, forms the occluding agent, i.e. the floor of the blind end of the upper segment, if the oesophagus continues back from the occlusion, undivided from the trachea, then the whole of the tracheo-oesophageal septum has been obliquely placed.

GIFFHORN[5] published in 1908 an account of two cases, in both of which the lower segment of the oesophagus communicated with the trachea by a slit-like opening, and fibrous bands connected the upper and lower segments. He found that in the literature of the subject the majority of cases resembled his two, and he mentions amongst others the frequent co-existence of other malformations (especially atresia ani). and pneumonic patches in the lungs, and redness of the mucous membrane

of the trachea and oesophagus. As a variation of the above he found that sometimes the lower segment of the oesophagus communicated with the trachea by a wide opening so that the two tubes, alimentary and respiratory, formed a common passage (zusammenhangende Kanal).

He enters at some length into the question of the etiology of the malformation. He finds the explanation of the fistula in the incomplete union of the lips of the tracheal outgrowth from the foregut. The atresia of the oesophagus is, however, more difficult to account for. He quotes Forster's theory that the atresia depends on "einem Stehenbleiben des Osophagus auf der Stufe der Entwickelung, in welcher es noch solid war"; hollowing of the solid tube has taken place above and below, but not in the middle portion. While he considers that Forster's theory has something to recommend it, he advances his own opinion that there is an adhesion (Verklebung) between the upper portion of the lips of the tracheal outgrowth and the posterior wall of the foregut which is squeezed in between these lips. This explanation is akin to that of Shattock (loc.cit. p.93), viz:- that the outgrowth of the diverticulum from the anterior wall of the foregut drags forwards and kinks the posterior wall. Shattock finds support for this view in the narrowing of the oesophagus which normally occurs in this situation. At the same time Giffhorn (loc.cit. p.125) thinks that the type of case where the tracheo-oesophageal opening is wide is to be explained by Mackenzie's theory of an abnormal situation (Verlagerung) of the lips of the tracheal opening, or as Keith(v.supra.) puts it, the tracheo-oesophageal septum has been placed obliquely.

SCHNEIDER[6] in a recent work thinks it probable that in this displacement of the lips of the orifice of the tracheal diverticulum a defect of formative material plays a part. He bases this opinion on the case, reported in 1845 by Levy, in which the lower segment of the oesophagus arose from the root of the right bronchus, and in which there was defective development of the right lung.

FORSSNER[7] considers that the primary cause of the malformation is disturbance in the separation of the trachea from the oesophagus. He thinks that the alterations in the capacity (Raumverhältnisse) of the oesophagus, depending on the persistence of the fistula, are a predisposing factor in the origin of an epithelial occlusion of that tube, which occlusion, again, may be the reason for the frequent co-existence of atresia of the oesophagus with oesophago-tracheal fistula.

LEWIS[8] draws attention to an appearance in the model (which he figures) of Bremer's embryo. This is an oblique sulcus on the outer aspect of the lateral wall of the oesophagus, with corresponding elevation of the epithelial surface. It "seems correlated with the shape of the adjacent body-cavity"; but "apparently" it "has not previously been described," and Lewis refers to its potentially etiological relationship to oesophageal atresia. It is worthy of mention that in the model there is, in addition to the lateral oesophageal groove just mentioned, a notch between the lung-bud and the oesophagus, and, further, a certain degree or approximation of the lateral walls of the fore-gut immediately above the notch to form the tracheo-oesophageal septum.

From a consideration of the specimens described above, and of the literature of the subject it seems highly probable that the malformation is due to an abnormal relationship of the tracheo-oesophageal ridges to the posterior wall of the oesophagus.

These ridges, which in normal circumstances separate the trachea from the oesophagus - the "Trennungsleisten" of Giffhorn (loc.cit. p.125) - and so form the tracheo-oesophageal septum, are probably placed obliquely so that their hinder ends come in contact in the posterior wall of the oesophagus. This malposition of the ridges would account for the absence of separation of the trachea from the oesophagus lower down, as in Levy's case (v.supra).

It is not common to meet with this high degree of malformation. More frequently there is a partition forming the lower margin of the tracheo-oesophageal communication, which, as we have seen, Keith explains as due to the tracheo-oesophageal ridges having been formed in two parts. This explanation of Keith's is, I think, supported by the appearances in Bremer's embryo. In this embryo it will be remembered that there was, in addition to the notch between the lung-bud and the oesophagus (and also the beginning of a septum), an oblique infolding of the lateral and postero-lateral wall of the oesophagus.

Whether we have to deal with ridges (in part or wholly obliquely placed) approaching the hinder wall of the gut, or with a dragging forwards of the gut-wall to meet the upper part of normally-placed ridges, contact is brought about. The contact becomes permanent, and occlusion results. The ridges in contact with the posterior wall of the gut unite with one another in normal fashion to form the posterior wall of the larynx.

R E F E R E N C E S.

1. MACKENZIE, Diseases of the Throat & Nose, 1884, Vol.II. p.216.

2. SHATTOCK, Trans.Path.Soc.of London, 1890, Vol.XLI. p.87.

3. BALLANTYNE, Antenatal Pathology & Hygiene - The Embryo. 1904, p.462.

4. KEITH, Journal of Anatomy & Physiology, 1906, Vol.XLI. pp.52-55.

5. GIFFHORN, Virchow's Archiv. Bd. 192, 1908, s.112.

6. SCHNEIDER, in Schwalbe's Morphologie der Missbildungen des Menschen und der Tiere, III. Teil, VIII. Lieferung, 2 Abteilung, 8 Kapitel, 1912, s.777.

7. FORSSNER, Anatomische Hefte. Band 34, Heft 102. 1907. s. 1-163.

8. LEWIS, in Keibel & Mall's Manual of Human Embryology, Vol.II. 1912, p.313.

B. DUODENUM

Occlusion of lumen by septa.

This specimen, the only one of its kind in my series, is from Case I. The case occurred in the practice of Dr.David Glen, of Glasgow, and I was enabled by his kindness to examine the body after death. Birth was premature (6 and a half months) and the patient survived only one day. Besides atresia ani there was no other external malformation.

On opening the body, which was that of a male infant, it became evident that it was the subject of transposition of the viscera.

Abdominal Viscera.- The umbilical vein passed into the liver, 1.5 c.m. above its lower margin, and at the upper end of a vertical groove. The appendix and caecum occupied their usual position in the right iliac fossa; but, on following up the ascending colon it was found to pass backwards under the right lobe of the liver and to enter the descending colon which coursed down, to the right of the mesentery, and behind the ileo-caecal junction, to end in the rectum. The rectum ended as a blind pouch close to the perineum. On turning up the liver the stomach was found coming from behind the posterior edge of the right lobe, above the right kidney and coursing forwards for nearly 3cm. to end at the right extremity of a bulky duodenum, distended with greenish material and overhung by the left lobe of the liver (fig.3). At its cardiac end the stomach measured nearly 1cm. vertically; but before reaching the duodenum it narrowed to half of this measurement. The

stomach, and still more the distended duodenum, contrasted with the small intestine which was about 6mm. in diameter: the colon was 7-8mm. From the left (distal) extremity of the cyst-like duodenum the small intestine passed, its coils lying over the front of the left kidney. The line of attachment of the mesentery extended from the front of the upper pole of the left kidney, along the lower margin of the distended duodenum and as far as its right extremity, whence it ran down to the ileo-caecal junction, springing from the right of the vertebral column,and continuous with the mesocolon.

On removing the liver, the gall-bladder was found embedded in the under part of the left lobe, and passing towards the right extremity of the distended duodenum. After removal of the liver, the cardia of the stomach was seen under the right side of the dome of the diaphragm. The distended duodenum had a maximum diameter of about 2.5cm., and compression failed to drive its contents back into the stomach or forwards into the distal bowel. The spleen was in its normal relationship to the cardiac end of the stomach, and measured 1.5cm. in length.

On cutting open the duodenum it was seen that the distended portion was shut off from the remainder of the bowel by two septa. The proximal of these was situated just above (i.e. on the pyloric side of) the gall-bladder, and from the fact that the contents of the distended portion were inky in colour and its mucous membrane was stained almost black, while that of the bowel between the septum and the pylorus was of a pale green colour, it was presumed that the septum was higher up in the bowel than the entrance of the bile-duct.

HISTOLOGY. The distal portion of the cyst-like duodenum, with the adjoining undistended bowel, was removed en bloc and hardened. On trimming the specimen previous to embedding in paraffin it was observed that, at the seat of its junction with the undistended bowel, the wall of the cyst was thickened over an area much greater than the diameter of the latter.

An interrupted series of vertical longitudinal sections was made which showed the occlusion between the two portions of gut to be formed of their respective mucosae placed back to back and separated from one another by a scanty layer of submucosa. There was no appearance of any inflammatory process. The muscular tunics of the bowel passed uninterruptedly from the dilated to the contracted portion. The proximal end of the latter was spread out on the former in both antero-posterior and vertical directions, giving rise to the thickening of the wall of the cyst, mentioned above. The epithelium of the mucous membrane generally was not well preserved, and was best seen in the spread-out proximal end of the contracted gut.

A series of longitudinal sections of the proximal (pyloric) end of the cyst was made. These showed that the septum between the cavity of the cyst and the lumen of the bowel on the pyloric side of the cyst differed in structure from the distal septum. It showed two layers of mucous membrane, each with its submucosa. Between the two submucous layers was a layer of muscularis, derived from and continuous with the deeper layers of the intestinal muscularis. The septum was not solid; but was traversed by a channel, lined by columnar epithelium,

with here and there a clear, or "goblet", cell. This channel was continuous below with the lumen of the proximal bowel: above, it was continuous with ducts in the pancreatic tissue. These, again, presumably after union with the bile-duct (n.b. the bile-duct was not found as a separate structure, probably owing to incompleteness of the series of sections) communicated with the cavity of the cyst by a single large opening. At the seat of communication between the septal channel and the pyloric part of the duodenum another pancreatic duct (? duct of Santorini) coursed down the anterior wall of the cyst and opened into the pyloric part of the duodenum. (A reconstruction of the sections is shown in the diagram fig.4).

The epithelium lining the ducts was pure columnar; but as the opening of the ducts into the lumen of the gut was approached goblet cells were superadded in increasing numbers.

L I T E R A T U R E

It is impossible to ascertain from literature the actual frequency of congenital malformation of the intestine. There must be many cases which are not recognised, or in which, even if recognised, no opportunity is given for detailed examination of the lesion. Besides, allowance must be made for cases where the lesion may have been examined but the result not published.

Excluding for the present any consideration of the malformations of the

rectum and anus, I shall for convenience divide intestinal malformations into three classes, according as they occur in (1) the duodenum; (2) the jejuno-ileum; and (3) the colon.

I wish first to consider the comparative frequency of malformation affecting the duodenum, taking up afterwards the nature and seat of the malformation, abnormality of ducts, and lastly the etiology of the condition.

(a) Comparative frequency.

The literature of the subject is rather confusing. Thus JOHNSON[1] writes that next in frequency to the rectum and anus comes "the site of the omphalo-mesenteric duct in the ileum. Atresia or stenosis of the duodenum has been observed in a fairly large number of cases. The seat of the closure or stenosis has been in most cases above the papilla...Congenital stenoses of the large intestine are, except at its lower end, rare".

MURPHY[2] states that congenital occlusion of the small intestine is "most frequent in the duodenum; the next most common site is the ileo-caecal region".

BALLANTYNE[3] does not definitely state the frequency, but remarks that "obstruction of the duodenum has been noted.... The ileum is apparently a common seat of intestinal obliteration".

If we go back a little into German literature, however, we find more definite statements. Thus SILBERMANN[4] tabulated 57 cases of congenital occlusion of the small bowel. The frequency was as follows:-

Duodenum, 24 cases (42.1%)

Jejuno-Ileum, 31 cases,(54.38%)

Jejuno-Ileum & Colon 2 cases (3.5%)

GÄRTNER[5] gives particulars of 65 cases of congenital atresia and stenosis of gut. In the seventh group ("Fälle von rein casuistischem Interesse") there are 38 cases. Analysis of these shows the following incidence:-

Duodenum, 16 cases (42.1%)

Jejuno-Ileum, 18 cases (47.36%)

Colon, 4 cases (10.55%)

SCHLEGEL[6], in 1891 reported as follows:-

Duodenum, 29 cases (32.5%)

Jejuno-Ileum, 54 cases (60.6%)

Colon, 6 cases (6.6%)

These figures, if added together, give the average percentage of frequency.

Duodenum, 38.9%

Jejuno-Ileum, 54.11%

Colon, 6.88%

FORSSNER[7], as the result of his investigations gives the following numbers (p.89):-

Duodenum, 79 cases (35.9%)

Jejuno-Ileum, 103 cases (46.8%)

At Bauhin's Valve, 16 cases (7.27%)

Colon, 22 cases (10%)

It will be seen, on comparing Forssner's percentages with the combined percentages of Silbermann, Gärtner and Schlegel, that there is not a great discrepancy so far as the duodenum is concerned. The discrepancy in the case of the jejuno-ileum is probably to be explained by the inclusion, in the former figures, of malformation at Bauhin's Valve with those of the jejuno-ileum. If we add Forssner's percentage for valve-malformation (7.27) to that for jejuno-ileum (46.8) we get 54.07 as compared with 54.11 of combined figures. If we combine the percentages of Forssner's cases with those of the previous table we get:-

Duodenum,	37.4%
Jejuno-Ileum,	54.09%
Colon,	8.44%

These figures show that malformations of the jejuno-ileum are actually the most frequent. Those of the duodenum come next, and lastly and a long way behind come those of the colon. But, as Tandler (Forssner, loc.cit. p.131) pointed out, the relative frequency, or frequency per centimetre of gut-length, is quite another matter. From this point of view the duodenum is very much the most frequent seat of malformation. Forssner (loc.cit. footnote, p.135) quotes Dreike's figures for the length of the different parts of the bowel: duodenum, 12cm., jejuno-ileum 253cm. and colon 58cm. Malformation occurs in the following proportions:-

Duodenum,	6.6 per centimetre
Jejuno-Ileum,	.4 per centimetre
Colon,	.38 per centimetre.

I do not consider that this point is of importance; but I mention it

because it is apt to lead to misunderstanding unless the difference between comparative frequency and per-centimetre frequency is kept in mind.

(b) Nature of Malformation.

In considering this point I shall refer only to Forssner's figures. He was able to find reported 220 cases in all; but in only 134 of these was the anatomical form of the malformation described. He divides the cases into three classes according to nature of the malformation.

In Class I. the malformation takes the form of a membrane stretched across the lumen of the gut. The membrane may be complete, causing an atresia, or incomplete (iris-like), in which case a stenosis is produced. Of 49 cases of congenital obstruction due to a membrane, 34 were atresias, and 15 stenoses. Of the 49 cases, in 17 the duodenum was the part affected: atresia occurred in 9, and stenosis in 8. In Forssner's own cases the membrane was composed of two epithelial coats with a layer of submucosa between them. But in some cases mucous membrane alone has been found, while in others a layer of muscularis in addition has been noted.

In Class II. there is an actual interruption of the gut, and there are formed two blind ends connected by a cord of varying thickness and length. Of this variety there were 40 cases - 35 atresias, and 5 stenoses. In the atresias the cord was solid; in the stenoses it was tunnelled, having a small canal-shaped lumen. These 5 cases of stenosis all occurred in the duodenum. In addition, this part of the gut was in 11 cases the seat of atresia.

In Class III. the interruption of the continuity of the gut is complete, and the blind ends are free. Of this variety, in 13 out of 45 cases the duodenum was affected. As a rule in the duodenum the blind ends are in apposition although no tissue connects them.

To summarize: in the duodenum the obstruction takes the form of

Membrane	(complete...9 (incomplete.8	17	(36.9%)
Interruption, with blind ends,	(atresia, 11 (stenosis, 5	16	(34.77%)
Free blind ends		13	(28.26%)
		46	

According to these figures there is not a great difference in the frequencies of classes I. & II. The variety showing free blind ends is less common than either of the others. The figures show further that atresia is more common than stenosis. This fact is more strikingly brought out in Cordes' paper (v.infra) where of 57 cases there were 48 showing atresia, and only 9 stenoses.

It is quite evident, in going over lists of recorded cases, that in the duodenum the occurrence of more than one obstruction is quite exceptional.

(c) Seat of Malformation.

Reference to the literature brings out the fact that the malformation in the duodenum is usually situated close to the point of entrance of the ductus communis.

PERRY & SHAW[8] investigated 7 cases. In 4 the obstruction was at or near the biliary papilla; in 3, at the lowest part of the duodenum; and in 1 it was close to the pylorus.

CORDES[9] reports 1 case, and summarizes 56 found in the literature. She finds that "occlusion of the duodenum is most frequently very near the opening of the common bile-duct into the duodenum, either above it or below it". It was above the duct in 20 cases (in 12, immediately above), and below in 13 (in 2 immediately below).

SPRIGGS[10] describes 1 case where there was dilatation of the stomach and duodenum, the latter structure ending abruptly at the "point of entrance of the bile-duct". There is no note of whether the bile-duct opened above or below the stricture.

KEITH[11] examined 8 specimens. "Immediately above the entrance of the common bile-duct the duodenum is occluded by a septum, which is covered on each side by mucous membrane, and contains muscular strata continuous with the muscular coats of the bowel."

(d) Abnormality of Ducts.

In connection with the relationship of the occlusion to the bile-ducts it is worthy of mention that several authors report the occurrence of accessory ducts. In the case reported by Cordes (loc.cit.) there was an anomaly of the common bile duct - "a branch of the same having been found leading into the dilated duodenum above the atresia...An accessory pancreatic duct is not present." This writer quotes Forster, that "sometimes the ductus choledochus divides into two branches, of which one opens into the stomach or even into the large intestine." She also judges that, in two cases reported by Theremin as stenoses, what was

supposed by him to be stenosed bowel was probably a branch duct. She states further that in "four other cases" in her series the clinical history and autopsy findings together render it very probable that a branch duct existed.

Keith (loc.cit) mentions two cases in which "an accessory pancreatic duct can be seen to open in the duodenum above the septum". Lewis (KEIBEL & MALL[12]) mentions Tandler having found in an 8.5mm.embryo a complete obliteration between the outlets of the dorsal pancreatic duct and the ductus communis.

(e) Etiology.

The etiology of congenital atresia or stenosis of the bowel has been the subject of much speculation. Various causes - developmental, peritonitis, volvulus, intussusception, post-ulcerative contraction, embolism or thrombosis of mesenteric vessels, enlarged valvulae conniventes, (Spriggs, loc.cit) - have been invoked, and different writers have gone, more or less fully, into the pros and cons. It is interesting to notice how authors have come more and more to regard the condition as depending on some error of development. "A number of facts, such as the repeated occurrence of duodenal occlusion near the site of the papilla, the existence, in probably more than one case, of anomalies of the duct system, the absence of Brunner's glands (in one case at least) at the point of and near the atresia, the striking absence of any signs of disease to which the condition might be traced and the occurrence of other malformations than that of the duodenum in a certain number of the cases, lead me to think that an error of development not yet explained may underlie this condition." (Cordes, loc.cit. p.423). Again,

"Most if not all malformations in the duodenum are associated in some way with the development of the large glands from this region". (Clogg[13].) Credit is due to BLAND-SUTTON[13] for drawing attention to the occurrence of malformations at the seat of "embryological events". He supports his statement by reference to imperforate pharynx occurring "at the spot where fore-gut and stomodaeum come into contact. An imperforate or septate duodenum occurs just above the bile papilla, the region where the diverticulum issues to form the liver and pancreas. Imperforate rectum and anus are due to imperfect union of the hind-gut and proctodaeum. Lastly, imperforate ileum occurs in the region where the primitive alimentary canal is in communication with the yolk-sac by means of the vitelline duct."

The observation by TANDLER[14] of the obliteration of the lumen of the duodenum as a phase of the normal development of this part of the bowel has certainly paved the way to a more accurate conception of duodenal malformation. It may or may not be invoked in the case of obstruction in the neighbourhood of the biliary papilla; but it certainly helps to an understanding of those cases where the obstruction is situated some little distance from the entry of the ductus communis. Following on Tandler comes Forssner (loc.cit.), who holds that an ingrowth of the mesenchyme, before the re-establishment of the duodenal lumen has been effected, offers a solution of the difficulty. The obstruction, according to him, is composed of connective-tissue embedded in epithelium. If the connective-tissue bridge be thin, a membrane is formed: if thicker, the interruption may take the form of a cord connecting the two blind ends of the bowel. Should the connective-

tissue be incomplete, or so weak that at one spot it tears, there will be an incomplete membrane - crescentic or like an iris. Through subsequent changes, occurring during foetal life, a thick obstruction may be thinned down to form a membrane, or a stenosis may be transformed into a valve.

Before leaving this subject I would draw attention to GRAY'S[15] suggestion as to the etiology in a case of complete interruption of the duodenum, which he investigated some two years ago. He advances the theory that during the third month the passing across the upper abdomen from left to right of the distal ileum and the caecum, drawing with them their fold of mesentery, may result, if the mesentery be sufficiently taut, in nipping through the duodenum across which they pass.

The only other theory of origin requiring mention here is that of foetal peritonitis. I shall have again to refer to this, when considering the subject of occlusion in the jejuno-ileum. But it has been brought forward in duodenal cases. The chief arguments in its favour, as advanced by Spriggs (loc.cit), are (1) the obstruction may occur at a part of the bowel which is not the seat of one of Bland-Sutton's "embryological events". Thus, of Perry and Shaw's 7 cases in 3 only was the obstruction at the site of the papilla. (II) The presence of adhesions. That these evidence a precedent peritonitis is almost beyond doubt; but, as Spriggs says, "it may be that the obstruction was the cause of the slight signs of localised peritonitis found in some cases, and not the effect, as has been generally assumed".

My dismissal of peritonitis as an etiological factor in duodenal

malformation may seem rather summary; but I think that it is hardly necessary to consider it at length, when the production of the malformation is so eminently capable of explanation by the facts of normal development. Bland-Sutton's suggestion is valuable, and Gray's theory as to the etiology in his case is ingenious: but it seems to me that the normal obliteration during development is the important fact. The discovery of this by Tandler, and its further investigation and application by Forssner, have resulted in placing in our hands a working theory which for simplicity and probability it will be hard to better.

REMARKS

If the specimen which I have described be now considered it is seen that there is (1) a septum-formation proximal to the biliary papilla; and (II) a second septum near the distal end of the duodenum. Between these septa the discharge from the biliary papilla is confined, and by its pressure the portion of bowel concerned has been distended to form a cyst.

So far, this seems simple enough. Microscopic examination shows, however, that these two septa markedly differ in structure from one another.

The distal septum is of simple construction - two layers of epithelium, separated by a submucosa. The proximal is traversed by a passage lined by epithelium and communicating dorsally with the ducts of the pancreas (? and liver):

ventrally, it communicates with the lumen of the pyloric part of the bowel.

It might be held that this canal in the substance of the septum represents an abnormal branching of the biliary ducts; but the presence of goblet-cells in its epithelium and of muscular tissue in the substance of the septum opens up the possibility of its being originally a stenosed portion of the duodenal lumen. The accumulation of bile distal to the stenosis is favoured by the septum lower down in the bowel, and the resulting distension of the bowel, has, instead of opening up the stenosis in a direction towards the pylorus, tilted up the narrow portion so that it comes to form a valve. Pressure, on this valve, from the distal (biliary) side serves to make the obstruction more complete.

The occurrence of the double obstruction is distinctly unusual (v.supra p. 19). The microscopic characters of the proximal septum are in support of Theremin's views already quoted (v.supra, p.20) as having been adversely criticised by Cordes. In the drawing (fig.3) the pancreas is represented as clumped together and practically encircling the bowel at the proximal pole of the cyst. It was not noted whether the gland was prolonged at all behind the stomach. This disposition of the pancreas may have some relation to the stenosis of the neighbouring portion of the bowel. It seems to me that Forssner's theory of ingrowth of mesenchyme satisfactorily explains the etiology of both proximal and distal occlusions in this specimen.

REFERENCES

1. JOHNSON, Surgical diagnosis, Lond. 1909, vol.II. p.72.

2. MURPHY, in Kelly and Noble's Gynecology and Abdominal Surgery, Lond.1908,vol.III. p.383.

3. BALLANTYNE, Manual of Antenatal Pathology and Hygiene. The Embryo. Edin.1904 p.532.

4. SILBERMANN, Jahrbuch für Kinderheilkunde und physische Erziehung, N.F. Band XVIII, s.420 et seq. Leipzig, 1882.

5. GÄRTNER, Ibid. Band XX. s. 403 et seq. 1883.

6. SCHLEGEL, quoted by BRAUN in Bruns Beiträge, Bd. 34, s.995, 1902

7. FORSSNER, Anatomische Hefte, Wiesbaden, 1907. Bd. 34, Heft 102.

8. PERRY & SHAW, Guy's Hospital Reports, 1894, Vol.L. (3rd.Series XXXV.) p.174.

9. CORDES, Archives of Pediatrics, 1901, Vol.XVIII. p.401 et seq.

10. SPRIGGS, Lancet, 1910, Vol.I. p.94.

11. KEITH, British Medical Journal, 1910, Vol.I. p.303

12. KEIBEL & MALL, Manual of Human Embryology, Lond. 1912, Vol.II. p.382.

13. CLOGG, Lancet,,1904, Vol.II. p.1773.

14. TANDLER, quoted by FORSSNER, loc.cit.

15. GRAY, Glasgow Medical Journal, 1910, Vol.LXXIV. pp.116 et seq.

C. I L E U M.

There are 5 specimens (and probably a 6th. specimen) of obstruction of the ileum in my series.

1. (Case II.) Obstruction 37cm. above ileocaecal junction; volvulus; adhesions-visceral, omental, and parietal.

A female infant, aged six days, was admitted to the Royal Hospital for Sick Children on June 23rd. 1907, with a history of vomiting from the day after birth. No meconium had been passed; but there was a discharge per anum of whitish slimy material. The anus was well-developed, and the anal canal was patent, admitting the little finger as far as the middle of the second phalanx. There was no feeling as of a septum with distended bowel above; but rather of a channel gradually narrowing so that its termination could not be reached by the finger-tip. An obstruction high up in the bowel was diagnosed, and the abdomen was opened in the left iliac fossa. A finger, passed into the wound, was directed to the left along the parietal peritoneum and into the iliac fossa, where a cord-like, undistended portion of bowel was encountered in the situation of the sigmoid flexure. The abdomen was closed, and the child died 5 days later from progressive weakness, with constant vomiting.

At the post mortem examination the right flank was occupied by greatly distended bowel extending from the iliac fossa up to the margin of the right lobe of the liver, to which it was adherent below and internal to the fundus of the gall-

bladder. The great omentum was adherent, high up, to the mesial surface of this piece of bowel.

Further examination of the parts showed the condition depicted in the diagram (fig.5). On referring to the diagram it will be seen that in the iliac fossa the lower (proximal) end of the distended bowel is directed horizontally towards the left (middle line) and joins a coil of small intestine. In front of the junction a piece of cord-like, undistended bowel runs upwards and backwards into the flank and terminates on the mesial aspect of the upper end of the above-mentioned dilated gut. This undeveloped bowel, on being followed down, is seen to course backwards round the horizontal junction, and then to the left. The distended gut on being freed from the liver, and turned down is found to be adherent on its outer side for a distance of 1.75cm. to the parietal peritoneum. This adhesion contains large blood-vessels ramifying in a backwards direction on the abdominal wall. On the mesial aspect of the dilated bowel its mesentery stretches between it and the undeveloped portion. On following the mesentery down it is found to accompany the undeveloped bowel over the horizontal piece to the under aspect of which it is attached, and then to pass to the left and upwards behind. At this point the undeveloped gut courses backwards and to the right to join the caecum.

This somewhat confused arrangement of the bowel was due to <u>volvulus</u> which was untwisted by turning downwards and forwards (a half turn), and then from right to left (one complete circle and a half) the dilated gut. This manipulation showed the very great degree of dilatation of the distended gut - nearly 4cm. in diameter.

The volvulus had occurred about 12cm. above the distal end of the dilated portion of gut, and had not produced any obstruction of the lumen of the bowel. There was nothing abnormal in the arrangement of the mesentery, and the portion corresponding to the undistended segment of the ileum contained well-developed lymphatic glands. The whole of the great intestine was undistended. The caecum lay on the psoas muscle, to which it was firmly attached a little below the iliac crest.

Measurements. The great intestine, from caecum to anus, measured in length 43cm. The small intestine had a length of 188cm., and the distended portion joined the undistended at a point 37cm. above the ileo-caecal valve.

Details of Obstruction.- Attempts, made post mortem, to force fluid from the narrow into the dilated gut were unsuccessful, and resulted in rupture of the end of the contracted gut, and extravasation under the peritoneum of the dilated.

On the anterior surface of the junction of the dilated and contracted portions of the bowel, a fibrous adhesion stretches between these portions (fig.6a). The adhesion is situated a little in front of the mesenteric line, and the continuity of the latter can be displayed only when the adhesion is lifted up. After removal of this adhesion and of the mesentery behind it, the termination of the dilated portion of the gut is seen to be doubled on itself so that the ballooned blind end is directed downwards on the mesial aspect of the ascending piece of gut, behind the line of attachment of the mesentery. The doubling is maintained by fibrous adhesions stretching between the apposed peritoneal surfaces. Two of these adhesions are situated one behind the other (fig.6,b. & ~~fig.7,d~~ c.), the anterior being just

behind the line of mesenteric attachment, which passes upwards and to the left to be continued into the mesenteric line on the contracted bowel beyond the obstruction. These adhesions form stout bands, and between them is a shorter adhesion, to be again referred to.

On cutting away the anterior wall of the distended bowel the doubling is seen to have produced a kinking so that the lumen of the bowel is very much encroached upon by a spur. This spur, and the short, substantial, fibrous adhesion (mentioned above) which assists in its maintenance are well shown in fig.7 *d*. The inner surface of the wall of the gut is marked by transverse ridges. The seat of junction with the contracted gut is indicated by a slight swelling, of circular outline, with a blind depression in its centre. A probe introduced into the contracted bowel was forced through the junction at this spot; but, as was afterwards found on microscopic examination, the communication was artificial. The junction was bisected in the vertical direction, and it was then found that in the posterior half the contracted gut run upwards and outwards on the surface of the distended gut, from the lumen of which it was separated by a thin septum. Extravasation of some of the debris from the contracted gut was seen under the peritoneum of the distended gut. In the anterior half, the septum showed an oval opening allowing of communication between the dilated and contracted portions of gut; this was the opening made by the probe.

HISTOLOGY.- Sections made in the long axis of the bowel showed that a complete mechanical obstruction existed, the continuity of the lumen of the gut being interrupted by a septum, formed of mucosa and submucosa (see diagram, fig.8).

Owing to faulty preservation of the specimen the mucosa of the distended gut was not demonstrable; but this layer was better preserved in the contracted gut. The thickness of the occluding layer of tissue was markedly lessened at the point corresponding with the depression in the wall of the distended gut. This depression was situated nearly opposite to the commencement of the lumen of the contracted gut.

The muscularis was much more marked in the contracted than in the distended gut. It did not form part of the occluding septum; but although it passed continuously from distended to contracted bowel on the concavity (i.e. mesenteric aspect) of the angle of junction, it apparently formed here a spur. On the convexity the muscularis was not nearly so well marked, and near the junction it apparently disappeared from the contracted and was sparsely represented in the distended gut. It was not clear whether the spur was a secondary formation, due to kinking, or indicated an attempt on the part of the muscularis to take part in the obstruction.

In none of the sections was the mucosa of the contracted bowel intact. In all it had been ruptured by the fluid injected into the contracted bowel to distend the latter, and the rupture opened into an artificial cavity formed by separation of the layers of the wall of the congenitally distended gut.

The next specimen belongs to a different type; and is an example of complete interruption, the malformation being multiple.

2. (Case III.) Multiple interruption of ileum, high up; coiled-up mass of undistended bowel.

A female child, aged 4 days, was admitted into the Royal Hospital for Sick Children on 13th.July, 1910, with a history of no passage *per anum* of meconium. The anus was well formed, and admitted a probe a distance of 6.25cm.

The abdomen was opened and an artificial anus made; but the child died in the course of 24 hours.

On *post mortem* examination it was found that the artificial anus had been made in the small intestine, a distance of 46cm. below the pylorus. There was complete interruption of the bowel 25.5cm. further down (71cm. from the pylorus). Beyond this there was spherical mass of bowel coiled on itself, and beyond this again the bowel recommenced and continued, without interruption, to the caecum, a distance of 91cm. The great intestine measured 39.5cm., and, like the distal portion of the small bowel, was of normal shape but undistended.

Closer inspection of the parts at the seat of obstruction showed that the proximal portion of the bowel only apparently terminated abruptly: it was sharply kinked at a spot 1.25cm. from its actual extremity, so that the part beyond the kink was collapsed, and was at right angles to the rest of the bowel. The short portion distal to the kink tapered to a termination which was in close apposition to the upper (mesenteric) extremity of the coiled mass next to be described. The kink was maintained by adhesions stretching from the mesentery over the right side of the collapsed bowel beyond. The distended bowel, after hardening, measured 2cm. in diameter; that of the portion distal to the kink was .75cm. On cutting open the bowel the kink is seen as a spur of the bowel-wall, on its mesenteric side.

In close contiguity with the termination of the kinked bowel lay the convoluted mass of undistended bowel. In the hardened specimen it was conical in shape (fig.9), and attached by its apex to the mesentery. It roughly resembled a cochlea with three turns, and it was so bound together in peritoneum that it could not be straightened out. On bisecting the cone, the cut surface showed various sections of coils held firmly together. From apex to base the cone measured 3.5cm.; and 2.5cm. across its base.

Following the free edge of the mesentery an ovoid body was reached at a distance of 2cm. from the attachment of the conical mass. This body measured 1.25cm. in length, and 6.25mm. transversely, and, on being bisected longitudinally, presented the naked-eye appearance of a piece of bowel closed at both ends.

It was immediately succeeded by the upper end of the collapsed bowel, to which it was attached by adhesions independent of the mesentery. The collapsed bowel had a diameter of 7-8mm.

The arrangement of parts is shown in the figure (fig.9) which was drawn after hardening. The specimen is viewed from the right side.

HISTOLOGY.- Two parts of the malformation were examined microscopically:-

(1) The kinked termination of the distended bowel:

(2) The conical mass of convoluted, contracted bowel.

(1) Longitudinal sections of the kinked termination of the distended bowel showed that the kink was formed by an infolding of the whole thickness of the bowel-wall to form a "spur". The structure of the bowel-wall above the kink was normal,

save that the epithelium of the villi had disappeared. It was present, however, in the deeper recesses of the mucous membrane. The muscularis consisted of two layers - inner (circular) and outer (longitudinal). As the region of the kink was approached certain well-marked changes in the muscularis were observed. On the convexity, the circular bundles rapidly became smaller in area at a spot where the bowel began to curve in to the narrow (a little above the level of the "Spur" formed by the infolding of the opposite wall), and from here on to the commencement of the contracted terminal piece of bowel they formed a thin, but distinguishable layer. On the other hand, the longitudinal muscularis was continued to the extremity of the contracted gut. The muscularis was differently arranged in the concavity of the kink. Here the circular bundles were continued into the proximal limb of the spur, for about two-thirds of its length. They remained of large sectional area, till about halfway towards the apex of the spur. They then diminished, but were of good size till they altogether ceased at a point short of the apex by one-third of the length of the spur. Just about the spot where the thick bundles diminished, the longitudinal ceased, rather suddenly, and were not recognisable in the remainder of the spur, or in the contracted-gut-wall continuous with it. The mucous membrane of the contracted gut was much destroyed, and, with the exception of some remains in the debris, was unrecognisable. (Fig. 9α).

In one section, two spaces lined with cubical epithelium occurred in the thickness of the wall, and just beyond the termination of the cavity of the contracted gut. These spaces were in close relationship to the terminal strands of the longitudinal muscularis.

(2) Longitudinal sections of the conical mass showed, to the naked eye

six or seven transverse or oblique sections of gut. The arrangement of parts is shown in fig.10. Of these the peripheral seemed to stain better than the two larger ones in the centre of the mass. Under the microscope, five of the seven sections show some muscularis: in none is there any trace of epithelium. In one section the muscularis was succeeded by areolar-like submucosa; but generally this was not represented, and we passed directly into the debris which filled the lumen of the gut. This debris showed as deeply-stained granules and masses of various sizes, arranged peripherally, and succeeded by very faintly stained material of similar character. The masses resembled clumps of epithelium, and in some cases suggested crystalline substance. The different sections were bound together by tissue which was in some parts areolar, but in most fibrous, and which not only formed a wrapping for the mass, but also was present between the various sections of bowel.

As regards the muscularis: in none of the sections was there a complete circle of muscle-fibres. In the five which showed muscularis the muscle was the longitudinal one. In none did it extend more than half the circumference of the gut, and, except in the case of the most central of the sections, it was confined to the outer halves of the peripheral sections.

It should be added that, while the circular muscularis was not very evident, traces of it were found here and there under the longitudinal bundles.

The next in the series resembles No.2, inasmuch as there is complete interruption of the bowel. It differs from that already described, in respect of the

presence of peritonitic adhesions which may have had a causal relationship to the malformation.

3. (Case IV.) Interruption of Ileum, high up and multiple; omental band associated with interruption.

A female child, aged 4 days, was admitted into the Royal Hospital for Sick Children on 21st.October, 1911. There was a history of there having been no movement of the bowels since birth. A rubber catheter could be passed into the rectum to a distance of 1 or 2 inches, and on withdrawal was observed to be covered with thick white pasty material. The abdomen was opened in the left iliac region, and an artificial anus made in a loop of distended bowel. The patient died, collapsed, 32 hours later.

On opening the body after death the great omentum in the pyloric region was found stretched out to form a band about 3.25mm. thick, and 7.5cm. long. This band passed downwards and to the left to be attached to a gland-like body in the mesentery at the seat of interruption of the continuity of the bowel.

The interruption had occurred in the small bowel, and was distant 91.5cm. from the pylorus, and 101.5cm. above the ileo-caecal junction. The caecum and appendix were situated high up on the front of the right kidney, so that there was practically no ascending colon. From caecum to anus the bowel measured 44.5cm.

The artificial anus had been made 15cm. above the blind end. The gut beyond the interruption was collapsed, its diameter varying between .5 and .75cm. The distended gut measured about 4cm. across, at its termination: higher up, it was

about 2cm. The duodenum was little more than 1cm. (These measurements are not true diameters, being made with gut lying flat). The stomach measured 6.25cm. in length, and 4.3cm. transversely.

The parts were fixed before a detailed examination of the seat of interruption was made. Inspection then showed the arrangement figured in the outline drawings (figs.11, 12 and 13).

From below upwards in the drawing (fig.11), are seen (1) the distended proximal bowel, with its blind extremity: (2) the collapsed bowel distal to the interruption: and (3) a piece of tissue, 2.5cm. long and 1cm. across.

The peritoneal coat of the distended gut was smooth until the apex of the blind end was reached. Here it presented (fig.14) an irregular surface over an area of about 1.25cm. in diameter. On cutting open the distended gut the mucous membrane was seen to be transversely ridged, save at the extremity, where it became granular. At the very apex was a pit-like depression of pinhole size, with raised edge. The portion of gut-wall corresponding with the area of granular mucous membrane was very thin and translucent, especially at the "pit", and the irregularity of the peritoneal coat here was suggestive of its having been forced up from the muscularis.

The collapsed gut which formed the distal limb of the interrupted loop was irregularly bent on itself. This was seen both from the right and the left side; but the arrangement was best seen when the bowel was viewed from its free surface (opposite to the mesenteric attachment). It was disposed so as to form a figure resembling an interrogation-mark, angular instead of rounded. The portion forming the head of the interrogation-mark was over 9.5cm.long, and its concavity was

towards the right side of the mesentery.

The third structure to be noticed is that shown in the figure (figs.11,12) as lying above the collapsed gut, and in close relationship with the omental band. This structure resembled a lymphatic gland, and had the appearance of an elongated bean, the hilum of which was turned towards the collapsed gut below it. It was firmly bound to the collapsed gut by fibrous tissue. This fibrous tissue passed into the concavity of the loop of collapsed gut; but some of the fibres passed over the convexity of the gut and blended with its mesentery. The omental band already mentioned passed behind the bean-like structure, and spread out, some of its fibres blending with the fibrous tissue joining that structure to the collapsed gut; other fibres of the band passed downwards in front of the loop of collapsed gut and ended in its mesentery. In the concavity of the bean-like body lay a small spherical node, embedded in the fibrous tissue.

HISTOLOGY.- Sections were made of

(1) The distended bowel: (a) some distance from blind end:
(b) blind end near "pit"
(c) "pit).

(2) The bean-like body, with attached node.

(1) (a) Longitudinal sections of the wall of the distended gut, some distance from its blind end, including a transverse ridge (valvula connivens), showed two well-marked layers of muscularis (fig.15). At the seat of the ridge there was some increase in thickness of the circular layer of muscle: here also there were very large vessels in the submucosa, which was slightly thicker than in the remainder of the section. There was well-marked arrangement of villi; but the epithelium over

them has not been preserved during hardening. Here and there, epithelium of glandular recesses was visible.

(b) Sections of the blind end, close to the "pit" showed mucous membrane with an occasional villus, but devoid of epithelium (fig.16). Here and there in the deeper parts of the mucous membrane cross-sections of Lieberkuhn's glands were seen, and immediately external to them was the well-marked muscularis mucosae. In the tissue external to the muscularis mucosae were numerous, irregularly-coursing bands of muscular tissue, which became thinner on being followed from the edge to the central portion of the section. There was in the outer part of the very vascular subperitoneal tissue an irregular cavity with deeply-stained crystalline contents which caught the edge of the razor. Part of the contents had come away: the remainder were adherent to the wall. The cavity was surrounded by fairly dense connective tissue of the subperitoneal coat, and close to the cavity were small calcareous foci, embedded in the fibrous tissue.

(c) Sections including the "pit" (fig.17) showed the latter to be formed by a depression affecting the mucosa and muscularis, and leading into an irregular cavity containing calcareous material. This cavity corresponded with that seen in (b), where it was situated in the subperitoneal tissue. Here, however, it lay rather in the deeper part of the mucous membrane and muscularis.

(2) Sections made in the long axis of the bean-shaped body, and including the node in its concavity, showed that both structures were really a portion of malformed intestine. In only one portion of the section, viz:- that corresponding to the "node", was there any approach to the normal structure of bowel-wall. In one section the appearance of this portion was that of a transverse section of bowel.

The mucosa was well-formed, but the epithelium very indifferently preserved. The submucosa was, except at one spot, very much compressed and flattened. At this spot, as well as at another where the submucosa approached its normal dimensions, the two layers of muscularis were well-marked. For about three-fourths of the circumference the longitudinal muscularis was easily distinguished: the circular, however, was clearly marked in only about one-third of the circumference. In subsequent sections this portion of bowel was found to be continuous with the larger mass, of which it was really a part.

Elsewhere in the section (particularly along the convexity) were well-marked bundles of muscularis - chiefly longitudinal - but nowhere was the appearance of a complete section of bowel. No mucosa was distinguishable, and the abnormally vascular and abnormally bulky submucosa filled the lumen of the gut.

The following specimen in my series differs from the preceding examples of malformation of the ileum, in that the abnormality affects the terminal portion of the bowel, and that the blind ends of the large and small bowel are connected by a "cord" running along the free edge of the mesentery.

4. (Case V.) Interruption near termination of Ileum: blind end connected by cord with ileo-caecal region of large bowel.

This specimen was obtained from the body of a siren foetus, born at full term, and given to me by Dr.Stirling Robertson, of Clydebank.

There was no trace of external genitals or anus, and the hinder end of

the body ended in a "tail" which was curled ventrally on the lower abdomen. On straightening out the "tail" a rounded band (? muscle) stood out prominently in the middle line of the hypogastrium. The tip of the "tail" showed posteriorly a sulcus in the integument.

The abdomen was opened and the viscera, much softened by decomposition, were removed.

The caecum and appendix were found in their usual situation, from which region the ascending colon ran upwards and ended blindly, under the liver. The remainder of the colon was absent.

The lower part of the ileum was distended with meconium, and measured from 2cm. to 2.5cm. across when filled with water. It seemed to end blindly about 2.5cm. from the caecum. From the end of the distended bowel a cord-like structure (fig.18), measuring about 2.5cm. long by 4cm. across, extended, along the free edge of the mesentery, to the caecum which it joined above and internal to the origin of the appendix (at a spot corresponding to the normal ileo-caecal junction). Further examination of this "cord" showed that in its proximal (ileal) half it was really stenosed bowel, being possessed of a lumen which communicated with the lumen of the ileum. The orifice of communication was narrower than the lumen of the stenosed bowel, and at the seat of junction there was some angulation similar to, but in a less degree than, that seen in specimen No.2 (fig.9). A probe, passed into the stenosed bowel, stopped short half way along the "cord".

The appendix, which was 2.3cm.long, arose from the inner and lower aspect of the caecum and coursed upwards and outwards in front of the bowel. Its origin

was infundibuliform. The meso-appendix arose from the front (right side) of the mesentery, and from it an ileo-caecal fold extended downwards over the front of the terminal part of the "cord".

The caecum and ascending colon measured 7.5cm. in length: its diameter as the bowel lay flat was 1.25cm. The mesentery in the ileo-caecal region was normally shaped.

The small intestine, from the pylorus to the blind end of the ileum, measured 188cm. in length.

HISTOLOGY.- Transverse sections were made of the distal half of the "cord" connecting the large and small intestine. The appearances seen under the microscope were as follows:-

Peritoneum could not be distinguished, (doubtless owing to post-mortem maceration). There was a scanty layer of sub-peritoneal tissue, not very vascular, contrasting with the richly vascular subperitoneal tissue of the attached mesentery. The longitudinal muscularis was well marked, except at the part of the circumference furthest removed from the mesenteric attachment. The circular muscularis was not nearly so well marked. It was best seen in the region of the attachment of the mesentery. From here it thinned off on both anterior and posterior surfaces of the "cord", and it was hardly distinguishable in the remaining half of the circumference. The submucosa was quite distinguishable, being specially bulky in the anterior wall at the part furthest removed from the mesentery. Succeeding this was the mucosa, a reticulum with the spaces for the most part empty, and in the centre of this tissue were masses of deeply-stained material, possibly desquamated epithelium, filling a

narrow lumen. A little nearer to the caecum, however, there was complete absence of both lumen & of deeply-stained mass (figs. 19 & 19a).

On being traced, in successive sections, towards the caecum the "cord" expanded and a lumen became apparent, communicating with that of the caecum at the origin of the appendix.

Briefly - the structure of the "cord" was that of intestine. It was impossible to say whether it was obliterated, or merely stenosed: but in its middle portion it certainly was not functionating as a tube.

I now come to describe a condition which may be looked upon as <u>want of growth</u> rather than defect of development.

5. (Case VI.) <u>Congenital non-expansion of part of Ileum and of Colon: absence of septum or interruption of continuity of Bowel</u>.

This condition occurred in a male infant, aged 3 days, who was admitted into the Royal Hospital for Sick Children, 22nd.April, 1910, with a history of there having been no passage of meconium <u>per anus</u>. The anus was pervious, admitting a "large rubber catheter". The abdomen was opened, and the colon was found to be in the form of a firm cord-like structure. No operative treatment was undertaken.

After death the stomach and intestines were removed for examination. The small intestine, from pylorus to caecum, was 180cm. long: the great bowel was 33cm. The small bowel was dilated in its upper part, and reached its maximum at a spot about 99cm. from the pylorus. At this spot its diameter was 4cm. (nearly). From here down it gradually narrowed till it became cord-like about 28cm. above the ileo-caecal junction. The cord-like character of the bowel continued from here onwards

into the colon, and persisted right down to the anus. The diameter of the cord-like portion of the ileum was a little over 6mm.: that of the colon was slightly less.

The bowel was cut open, to determine if any obstruction of the lumen existed. None was found. The contents of the large gut were quite unstained by bile, and the same condition was found in the lower part of the small gut. Higher up, the bowel contained well-stained fluid faecal matter.

HISTOLOGY.- Transverse sections were cut of (1) distended small gut; (II) non-expanded ditto; and (III.) colon.

The arrangement and form of the coats of the small gut in (I) and (II) were normal. In the distended bowel, however, the muscular tunics were of much larger size than in the non-distended, having a sectional area of generally three or four times the size of those in the latter. The epithelium of the mucosa, on the other hand, was of larger size and better preserved in the non-distended bowel.

In the section of the colon the different layers of the wall were well formed. The lumen was filled with structureless debris (? inspissated mucus).

In (II) and (III) there was apparently no structural malformation. The sections showed in miniature the normal appearances of the parts..

Closely allied to the specimen described above is the following:-

6. (Case VII) Congenital non-expansion of colon: rupture at hepatic flexure, by enema.

A male infant, aged 3 days, was admitted into the Royal Hospital for Sick Children on 19th.September, 1910, on account of obstruction of the bowels. An enema had been given on the preceding day, and another on the day of admission, but without result, and the patient was accordingly sent to hospital.

There was great distension of the abdomen. A finger introduced into the rectum felt the bowel-lumen narrowing like the finger of a tight glove. Both testicles were in the scrotum, and the hinder end of the body was well formed and without any external malformation. The abdomen was opened in the left iliac region, and from it a quantity of bloody fluid and gas escaped. So much was this the case that it was at first thought that a piece of bowel adherent to the anterior parietes of the abdomen had been opened; but on enlarging the wound it was found that the fluid and gas were in the peritoneal cavity. The sigmoid flexure was brought out for inspection. It was collapsed, but was more succulent than is usually seen in a case of congenital obstruction higher up in the bowel, i.e. there was less of the feeling as if the intestine were a fibrous cord. An artificial anus was made in a distended loop of small intestine: when it was opened, somewhat light olive green faecal matter escaped.

The patient died early on the following day. Post mortem examination showed faecal matter and fluid in quantity lying free in the peritoneal cavity. The small bowel was much distended in its whole length; but the distension was less marked in the terminal portion of the ileum. The caecum was of about normal size; but its wall was thicker than normal.

In the colon, on the anterior surface of the hepatic flexure, was a rupture, transverse in direction and involving half the circumference of the bowel. Meconium issued from this rupture, and, on water being injected upwards from the sigmoid region of the bowel, a cast about 2.5cm. long was expelled through the tear. This cast was stained green at the end next the tear; its distal portion was white.

There was no sign of a definite obstruction (septum, stenosis, or interruption) in any part of the lumen of the bowel.

The above series of specimens of malformation of the ileum presents a number of points for consideration, e.g.:- frequency of ileal malformation; nature of malformation; its situation (part of bowel affected); etiology.

Before taking up these, however, it will be convenient here to tabulate the specimens, giving their principal features, exclusive of their histology.

TABLE

giving the principal features of the specimens of malformation of the ileum.

x.	Nature of Occlusion	Number of ditto.	Seat of ditto	Remarks
?.	Septum of mucosa and submucosa.	Single	Junction of middle and lower thirds of ileum (37cm.above ileo-caecal region).	Volvulus, not causally related to occlusion; peritoneal adhesions - visceral, omental, and parietal.
?.	Interruption, with blind ends.	Multiple (3)	Highest in upper third of ileum.	At termination of distended bowel, kink, succeeded by small portion of stenosed bowel; beyond first interruption, coiled mass of undistended gut, succeeded by ovoid blind piece.
F.	Interruption, with blind ends.	Multiple (2)	Higher in upper half of upper third of ileum.	Omental bands associated with interruption.
?	Interruption, with "cord" joining blind ends.	Single	Terminal portion of ileum.	Upper part of "cord" had lumen, and was kinked on termination of dilated bowel, as in No.2.
M.	Nil; non-expansion.	Single-widespread	Probably 87cm. from duodeno-jejunal junction i.e. in upper third of ileum.	Partial expansion had taken place downwards as far as upper part of lower third of ileum.
M.	Non-expansion.	-----	Not determined	Probably near termination of ileum, as distension less marked here than higher up.

Coming now to consider the points mentioned above, the first of these is

(a) Frequency of Ileal Malformation.

The comparative frequency, in the various parts of the bowel, of congenital malformation has already (v.supra, p.15) been dealt with, and statistics show that malformation occurs most frequently in the jejuno-ileum. Following the rough and ready method of dividing the jejuno-ileum into fifths, and considering the upper two fifths as jejunum and the remainder as ileum (QUAIN[1]), it will be seen that in all the specimens in my series the malformation affected the ileum.

(b) Nature of Malformation.

The following conditions are illustrated by the series of specimens:-

1. Obstruction by a septum.
2. Stenosis, with kinking at commencement of narrowing, and with multiple interruptions.
3. Multiple interruptions, associated with omental band and peritoneal adhesions.
4. Stenosis, with obliteration and cord-like continuation in mesentery.
5. Non-expansion of lower ileum and colon.

Putting aside for the present non-distension of a large tract of bowel (specimens 5 and 6), the remaining specimens exemplify in its different grades of severity congenital obstruction of the bowel,- stenosis; ditto with communicating cord; septum; and interruption of the continuity of the bowel-wall.

These varieties are all well known to pathologists, and examples are to be found recorded by writers on the subject. I have already (v.supra p.18)

referred to the different forms, with special reference to the duodenum. Taking FORSSNER'S[2] figures for the jejuno-ileum, - this writer found in literature 103 cases. In only 58 of these, however, were the details given sufficient to formulate statistics. Of these 58, in 16 the bowel was obstructed by a complete membrane: in 4 the membrane was incomplete: in 19 there were two blind ends connected by a cord which was in no case tunnelled: in the remaining 19 the blind ends were free.

Expressed in percentages the frequency of the different forms of obstruction is as follows:-

Membrane	complete.....16		
	incomplete.....4	20 cases	.....34.4%
Interruption, with blind ends			
connected by a cord	19	"	.....32.75%
Free blind ends	19	"	.....32.75%

If the two varieties with blind ends be taken together, they are almost twice as frequent as the membrane-cases.

In the present series of 4 cases, three showed blind ends, and only one a membranous septum.

(c) Situation of Malformation.

As just mentioned above, the ileal portion of the jejuno-ileum was the part affected in all of my four cases. There were, however, differences as regards the part of the ileum. In two (specimens 2 and 3), where there were multiple interruptions, but all quite close together, the proximal interruption was situated in the upper third of the bowel. In specimen 1 it was at the junction of the middle and lower thirds, and in specimen 4 it was at the termination of the bowel.

It would seem from the literature that congenital occlusion of the jejuno-ileum is most frequently met with at the lower end of the bowel. Thus SILBERMANN[3] in his table of cases mentions it as having occurred at the lower end of the ileum, 10 times. In 12 cases the exact situation is not given. In 3 cases it was between the jejunum and ileum (in 2 of these there were multiple occlusions); in 1 case it was at the middle of the gut; in 8 cases there were multiple occlusions.

In the cases given by GÄRTNER[4] the preponderance in favour of the lower end of the ileum is more marked:-

Jejunum	1
Between jejunum and ileum	2
Middle of small gut	1
Multiple occlusions	3
Lower end of ileum	9
Not stated	2 (1 of these presumably in lower part of gut).

BLAND-SUTTON[5] almost certainly referred to the lower ileum, when he wrote "imperforate ileum occurs in the region where the primitive alimentary canal is in communication with the yolk-sac by means of the vitelline duct". (see below).

According to BALLANTYNE[6], "congenital intestinal obstruction may be situated in the jejunum.....The ileum is apparently a common seat of intestinal obliteration...Probably most of the cases reported as instances of congenital occlusion of the small intestine affect the ileum....The seat of obstruction is frequently the place where the small intestine becomes continuous with the large". He adds, however, that multiple constrictions have also been noted.

The importance of determining as accurately as possible the situation of the malformation depends upon the relationship of situation to one of the theories of the etiology of congenital occlusion. This will be referred to below. Meanwhile I would simply draw attention to the fact, observed by previous writers and here illustrated by specimens 2 and 3, that the malformation may occur in the upper part of the ileum. This fact was also illustrated by another specimen, which I recorded some years ago[7].

(d) Etiology.

As already mentioned (v.supra p.21) a large number of theories have been propounded with the object of explaining the etiology of congenital occlusion of intestine.

Of these I propose to consider three:

1. Embryological.
2. Foetal peritonitis.
3. Volvulus.

1. Embryological.- The view that the malformation is the result of a departure from the normal course of development is favourably entertained by many writers. Several theories fall to be considered under this heading.

(a) AHLFELD[8], in 1873, advanced the theory that the physiological hernia of the umbilical loop of bowel continued from traction by a persisting omphalomesenteric duct, and might later be separated from the abdominal cavity by the coming-together of the body-walls at the umbilicus. Such a process seemed to him a

likely explanation of the appearances which he found in one case,- a newly-born infant in which a tumour was present, attached to the umbilicus by a fine pedicle, and containing necrotic bowel filled with meconium. At the post mortem examination the blind ends of the ileum and colon were found adherent in the neighbourhood of the umbilicus.

This theory affords a likely enough explanation of cases in which the malformation affects the portion of bowel associated with the vitelline duct, viz: lower ileum.

b. BLAND-SUTTON (loc.cit.) in 1889, promulgated the theory attributing defects of the ileum "to excessive coalescence of the intra-abdominal section of the vitello-intestinal duct". The ground on which this author bases his theory is that "congenital obstruction and narrowing of the alimentary canal are always found in the situation of embryological events". He instances, in support of this, the occurrence of imperforate pharynx at the spot where fore-gut and stomodaeum come into contact; of imperforate or septate duodenum just above the bile-papilla,- "the region where the diverticulum issues to form the liver and pancreas", and, "lastly, imperforate ileum occurs in the region where the primitive alimentary canal is in communication with the yolk-sac by means of the vitelline duct".

This author, in the last edition[9] of his well-known work on Tumours (1911), writes "These curious defects are attributable to the influence of the vitello-intestinal duct, because they always occur in that portion of the ileum to which the duct, when persistent, is attached - that is, they do not occur within

30cm. of the ileo-caecal valve, and are rarely found at a greater distance than 1 metre from the caecum."

Bland-Sutton's theory has much to recommend it, and it offers a feasible explanation of the majority of cases of ileal occlusion, in which, as I have already pointed out (see Gärtner's figures, quoted above), the malformation is situated in the lower part of the bowel. Its weak point, however, is that it fails to explain those cases in which the occlusion occurs close to the ileo-caecal valve (e.g. Specimen 4, in my series), or those in which there are multiple occlusions, or, lastly, those in which there is occlusion situated high up in the small intestine.

(c) CLOGG[10] reported, in 1904, a case in which Ahlfeld's theory is supported. Referring to the etiology of congenital intestinal atresia, Clogg supports the embryological view. The vitelline duct may undergo excessive obliteration, or it may persist for an abnormal length of time and make traction on the growing bowel. Lastly, bowel so kept out may be snared by the umbilical ring. He rather inclines to snaring of bowel in an umbilical sac, with or without subsequent retraction within the abdomen.

(d) FORSSNER[11], in a lengthy paper on congenital atresias of bowel and oesophagus, expresses (p.135) the opinion that epithelial occlusion, shown by Tandler to occur normally in the duodenum, may exceptionally occur in all parts of the gut. This, followed by ingrowth of mesenchyme, will account for occlusion, single or multiple, in parts of the bowel unconnected with the vitelline duct. He considers that while this theory is applicable to some of the atresias of the jejuno-ileum, others are explicable by Ahlfeld's theory.

With considerable ingenuity Forssner shows, by means of diagrams, how the various forms (septum, blind ends &c.) of malformation may be brought about after the mesenchymal ingrowth has taken place.

(e) SPRIGGS[12], writing in 1910, expresses the opinion that no one theory will cover all cases. He considers that the majority are due to errors of development; others to foetal accidents (volvuli, torsions, or strangulations); and a minority to precedent peritonitis.

2. Foetal Peritonitis as a Cause of Occlusion.

There has been a good deal of discussion as to the role played by foetal peritonitis in the etiology of congenital occlusion. Ballantyne (loc.cit.p.533) does not actually refuse to admit it as a cause, "but", as he says, "there are difficulties in the way, for, in order to explain some of the reported cases, it must be supposed that the peritonitis is developed at a very early date in antenatal life, at a time in fact when it is not easy to understand how an inflammation can exist." Then again, as already mentioned (v.supra p.23), Spriggs reminds us that the obstruction may be the cause of slight signs of localised peritonitis, although he is of opinion that in a minority of cases peritonitis is the cause of the intestinal lesion. Forssner (loc.cit. p.129) does not favour peritonitis as a cause, he thinks it improbable that a peritonitis can disappear and leave no traces beyond a circumscribed atresia of gut; and further, he can not reconcile the peritonitis-view with the fact that the parts of the gut-wall which are nearest to the peritoneum are unaltered, while the innermost layer of the wall,- the epithelium - is the seat of the pathological process.

These opinions notwithstanding, I do not feel that we are justified in excluding peritonitis from the category of causal factors. In my paper already referred to (v.supra p.51) I have quoted the cases of W.Thomas[13] and J.Thomson[14] as examples of peritonitis causing occlusion; and in one of the cases in the present series (specimen 3) there can, I think, be no doubt of the occurrence. In another case (specimen 1) various adhesions were present, but did not seem at all to bear a close relationship to the occlusion, to which indeed it was evidently secondary.

3. Volvulus.

This would seem not to have been frequently observed, and it does not occupy a prominent place in the literature. It occurred in one of my cases (specimen 1); but it evidently had no causal relationship to the seat of the occlusion, from which it was quite distant. I am inclined in this case to view it as secondary to the occlusion, and immediately caused by the overloaded and unwieldy gut proximal to the occlusion.

e. Note on the Muscularis.

In specimen 2 (v.supra p.34) mention was made of defect in the muscularis. The circular bundles stopped short of the stenosed gut which was in direct continuity with the dilated gut above. The longitudinal coat on the convexity was continued to the termination of the stenosed gut; but on the concavity it ceased on the proximal limb of the "spur", just short of the point where the circular bundles terminated.

A similar defect was observed in the sections of the bowel coiled up to

to form the "conical mass" (v.supra. p.35) The circular bundles were represented only by traces here and there, and in none of the sections of the gut was the longitudinal coat present in more than half of the circumference.

In specimen 3 (p.40) and 4 (p.42) this deficiency of circular muscularis was also met with.

In specimens 1 (p.31) there was deficiency of longitudinal muscle of non-distended gut, where the latter joined the distended bowel. The muscularis of the distended bowel was so sparse that it could not be determined whether it represented either or both layers.

It is difficult to account for this defect of muscularis. It is in contradiction to Forssner's statement, just quoted, that in congenital occlusion the epithelium is the seat of the pathological process. At the same time, the higher degree of deficiency in the circular muscular coat shows that the inner coats of the bowel suffer more than the outer layers. This is quite consistent with the theory that the occlusion is brought about by pressure from without - either by Forssner's ingrowth of mesenchyme or by the contraction of post-peritonitic scar-tissue,- and is analogous to what may occur in strangulated hernia. In the latter lesion it is well known that the pressure exercised on the bowel-wall by the strangulating agent exerts a more marked effect on the mucous membrane than on the external layers. (SULTAN[15], COLEY[16].). It would seem that the obliteration, or, in the case of strangulated hernia, the ulceration begins and is most marked at the periphery of the circulation, and extends thence in a central direction.

f. Condition of the Gut distal to the Occlusion.

Spriggs (loc.cit) has drawn attention to the difference between the distal gut in congenital obstruction and that in acquired obstruction. In the former it is "not merely collapsed but firmly contracted up, having never functionated". The microscopic appearances of the distal gut in specimen 1 showed that the gut was of normal structure, reduced in size. They indicated that the contracted state of the gut was not caused by an active contraction of the muscularis: they pointed rather to a want of growth.

g. Obstruction without Structural Malformation.

In considering the "nature of the malformation" (v.supra, p.48) I purposely left out of account specimens 5 and 6. These two specimens did not represent any structural malformation. In specimen 5 (v.supra, p.43) the non-distended ileum possessed a normal structure, but in miniature, and the appearances indicated a want of growth rather than of development. The contents consisted of inspissated mucus, and it is probable that this acted as an obstruction to the onward flow of meconium. The obstruction had originally formed in, or had extended up to, the upper third of the ileum. That it had been partially overcome was shown by the gradual diminution in size from the greatly dilated upper part of the bowel to the point (28cm. above the ileo-caecal junction) where firmly-contracted gut began.

A similar condition, but with its upper limit in the lower end of the ileum, existed in specimen 6; but in this case attempts at forcible dilatation from below had resulted in laceration of the colon.

PEARCE GOULD[17], in 1882, reported to the Clinical Society of London a case in which "the caecum, the lower four inches of the ileum and the first four inches of the colon were filled with a firm whitish plug of inspissated mucus of the consistence of cheese". The plug was not adherent to mucous membrane but tailed off at each end into ordinary mucus, and at its upper end it was greenish in colour and softer in consistence than the rest. The wall of the lower end of the ileum showed on microscopic examination a normal structure. In the colon beyond the plug were found several masses of milk-white firm mucus. Below this the colon and rectum were empty and contracted to the size of a clay tobacco-pipe stem.

Mr.Gould pointed out that there had been no fault in development but obstruction from a plug which had probably been deposited sometime in the third month, before any bile had passed into the duodenum.

I must confess that the exact nature of cases like these is very obscure. There does not seem to be any answer to the question as to why an inspissated plug should be formed. The condition of the contracted bowel agrees so closely with that seen distal to a structural obstruction that it is extremely probably that the plug of mucus acts as an obstructing agent. That such inspissated mucus may be partially moved onwards by the pressure of meconium in the bowel above is shown in the first of my two specimens.

REMARKS ON SPECIMENS 1 - 4.

In this small series of specimens we have examples of the different forms in which congenital occlusion of the bowel is met with.

Specimen 1 is of simple construction as regards the occlusion; but it presents also volvulus and peritonitic adhesions. The volvulus has no direct causal relationship to the occluding septum. The twist is situated at the base of a loop of gut near the apex of which the septum occurs, and the twisted loop comprises both over-distended proximal and non-distended distal bowel. The volvulus has not caused any obstruction of the lumen of the gut; but it may, by producing congestion of the mesenteric vessels, have increased the distension of the already distended piece of gut between it and the septum. There can be little doubt that the volvulus has been induced by movements of the overloaded and unwieldy portion of bowel proximal to the septum. Either from the pressure of meconium alone, or aided by congestion, peritonitis followed by adhesions has occurred in places in the distended bowel after the volvulus has placed it in its final position.

With regard to causation, the position of the septum does not militate against Forssner's theory. But the malformation has occurred within the region of attachment of the vitelline duct in the embryo, and it is reasonable to attribute its formation to abnormality in the closure of the latter.

In Specimen 2 we have to deal with multiple interruptions. Their multiplicity and high situation characterise a condition which can hardly be attributable to anomaly in the closure of the vitelline duct. In this specimen the condition of

the coiled-up non-distended bowel points at any rate to a localised peritonitis, and in my opinion we are quite justified in attributing to the peritonitis an etiological role. The defect in the muscularis is comparable with that found in specimen 3.

In Specimen 3 the presence of a post-peritonitic band gives us a very clear indication of the etiology. The presence of a "pit" at the apex of the distended gut is interesting. It might be looked upon as a weakness of the wall at the place where a band has cut through the bowel. While this may be, this portion of gut certainly bears no evident relationship to any such structure, and the "pit" may, I think, be looked upon as the result of great pressure by meconium. It would, doubtless, had the child lived long enough, have ended in perforation. In this specimen, also, the greater deficiency of circular as compared with longitudinal muscularis, in the piece of gut closely related to the band, is to be noted.

Specimen 4 is an example of very marked stenosis, amounting practically to occlusion. The malformation is situated rather lower than the usual place of attachment of the vitelline duct. There are no signs of an old peritonitis, and the anatomical conformation of the mesentery and meso-appendix is normal. The presence, during embryonic life, of the vitelline duct in the near neighbourhood is undoubted, while Forssner himself (loc.cit, p.135) admits only that the occurrence of epithelial occlusion may exceptionally (ausnahmsweise) take place, in parts of the bowel other than the cranial portion. While, therefore, the production of the malformation in this specimen seems to me to be explicable by either Bland-Sutton's or Forssner's theories, I am inclined for the reasons just given to favour the former.

REFERENCES

1. QUAIN, Elements of Anatomy, 10th.Ed. London, vol.III. part IV. p.86.

2. FORSSNER, Anatomische Hefte, Wiesbaden 1907, Bd. 34, Heft 102.

3. SILBERMANN, Jahrbuch für Kinderheilkunde und physische Erziehung, N.F.Bd.XVIII. S.420 et seq. Leipzig 1882.

4. GÄRTNER, Ibid, Bd. XX. S.403 et seq. 1883.

5. BLAND-SUTTON, International Journal of the Medical Sciences, 1889, N.S. Vol.XCVIII. p.457.

6. BALLANTYNE, Manual of Antenatal Pathology and Hygiene. The Embryo. Edin. 1904 p.533.

7. EDINGTON, British Journal of Children's Diseases, Feby. 1906.

8. AHLFELD, Arch.f.Gyn, Bd. V.1873, p.230 (by Forssner loc.cit p.125).

9. BLAND-SUTTON, Tumours 5th.Ed. London, 1911, p.598.

10. CLOGG, Lancet, 1904, Vol.II.pp.1770-1774.

11. FORSSNER, loc.cit.

12. SPRIGGS, Lancet, 1910, Vol.I. p.94.

13. W.Thomas, British Medical Journal, 1886 Vol.II. p.925.

14. Thomson, Edin.Med.Journal, 1892, Vol.XXXVII. p.840.

15. SULTAN, Atlas & Epitome of Abdominal Hernias, Lond. 1902, p.88.

16. COLEY, Article on Hernia, in Keen's Surgery Vol.IV. Lond. 1908, p.43.

17. PEARCE GOULD, Lancet, 1882 vol.I.p.738; Clin.Society's Transaction 1882, Vol.XV. p.192.

CONCLUSIONS.

A. Oesophagus.

1. It is highly probable that imperforate pharynx is due to abnormal relationship of the tracheo-oesophageal ridges to the posterior (dorsal) wall of the fore-gut.
2. The abnormal relationship consists probably in the ridges being placed obliquely.
3. Should a partition exist below the tracheo-oesophageal fistula there is reason to believe that the ridges have been formed in two parts, of which the upper have been obliquely placed.
4. The lower ends of the obliquely-placed ridges having come in contact with the posterior wall of the fore-gut, a permanent union results, shutting off the upper from the lower segment of the oesophagus.
5. Meconium may be found in the bowel.

B. Duodenum.

6. There is frequently a close relationship between a duodenal occlusion and the point of entry of the bile and pancreatic ducts.
7. Occlusion without relationship to the ducts may occur.
8. While the former class may be related to the development of the biliary and pancreatic apparatus, both classes may result from the epithelial occlusion which normally occurs in the course of development of the duodenum.
9. A duodenal septum, although causing an actual obstruction, may be the seat of a

microscopic channel connecting the lumen above with that below the septum.

10. Such a channel has been erroneously looked upon as an abnormality of the ducts.

11. Abnormal disposition of the pancreas may accompany duodenal septum.

C. Ileum.

12. Occlusion of the ileum is usually caused by structural malformation - septum &c.

13. The malformation has a wide range of situation - from high up in the ileum to its lower end.

14. There may be no structural malformation, in which case obstruction is produced by a plug of mucus.

15. As regards etiology,- in two of my cases peritonitis was the causal factor: in two the cause was embryological - probably related to obliteration of the vitelline duct: in the remaining two no cause could be suggested for the mucous obstruction.

16. In three cases there was deficiency of circular muscularis - marked when compared with the longitudinal layer.

17. This deficiency is in keeping with the view that the occlusion of the bowel was produced by <u>pressure acting from without inwards</u>.

18. The gut distal to the obstruction is not merely empty, but unexpanded. Its structure is normal, but in miniature.

APPENDIX

Summaries of Cases from which Specimens obtained.

Case I. Male, premature (6 and a half months) still-born. Atresia ani: transposition of viscera: imperforate pharynx (p.1): duodenal septa (p.11)

Case II. Female aged 6 days. Obstruction of ileum by septum (p.27): secondary volvulus and peritonitis: otherwise normal.

Case III. Female aged 4 days. Multiple occlusions of ileum, high up (p.31); otherwise normal.

Case IV. Female aged 4 days. Multiple occlusions of ileum, high up, associated with omental band (p.36): otherwise normal.

Case V. Siren foetus. No trace of external genitals or of anus: occlusion lower end of ileum (p.40): blind ends of bowel joined by cord, partly hollow: large bowel represented by ascending colon only.

Case VI. Male aged 3 days. No structural obstruction, but gradual narrowing down of ileum: lower ileum and colon non-expanded (p.43): no other abnormality.

Case VII. Male aged 3 days. Distension of abdomen: gas and fluid in peritoneal cavity: colon and lower ileum not properly expanded: (p.44): laceration of colon near hepatic flexure: plug of inspissated mucus.

Case VIII. Male, full-time, dying immediately after birth. No trace of anus: scrotum empty: belly protuberant: atresia recti, and microscopic communication with urinary bladder: micro-urethra: distension of bladder and prostatic urethra: diverticulum of base of bladder: absence of prostate: irregular termination of vasa deferentia: imperforate pharynx (p.2): malformed limbs.

I N D E X

Dr. Symington D.Sc.

46) Nov 1913

List of Illustrations.

Fig. 1. Imperforate Pharynx
- 2. Imperforate Pharynx
- 3. Double occlusion of duodenum: transposition of viscera.
- 4. Diagram of duodenal septa.
- 5. Diagram of occluded ileum, with volvulus.
- 6. Details of occluded ileum
- 7. ditto: bowel cut open.
- 8. Histology at seat of occlusion.
- 9. Occlusion of ileum: coiled-up mass of bowel.
- 9a. Histology of same.
- 10. " " "
- 11. Interruption of ileum: omental band.
- 12. Different view of same.
- 13. " " " "
- 14. View of interior of distended gut, showing altered mucous membrane.
- 15. Histology of same.
- 16. " " "
- 17. " " "
- 18. Cord-like communication between blind ends of ileum & cæcum.
- 19. Histology of same.
- 19a. " " "

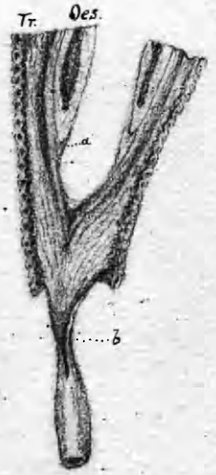


Fig. 1 (p.1)

a. Fascia stretching between upper + lower segments of oesophagus.

b. Narrowing of oesophagus below the junction with trachea.

The trachea + oesophagus split open from in front.

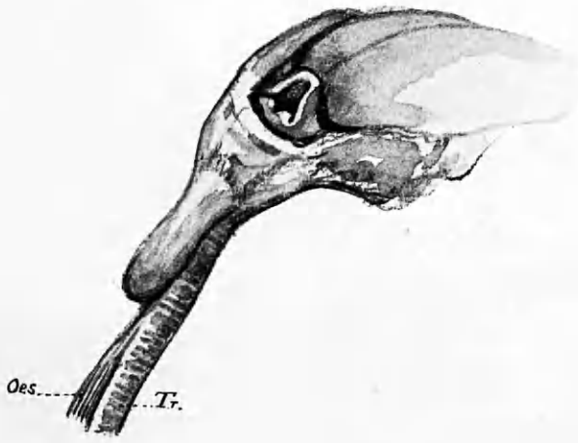


fig. 2 (p. 2).

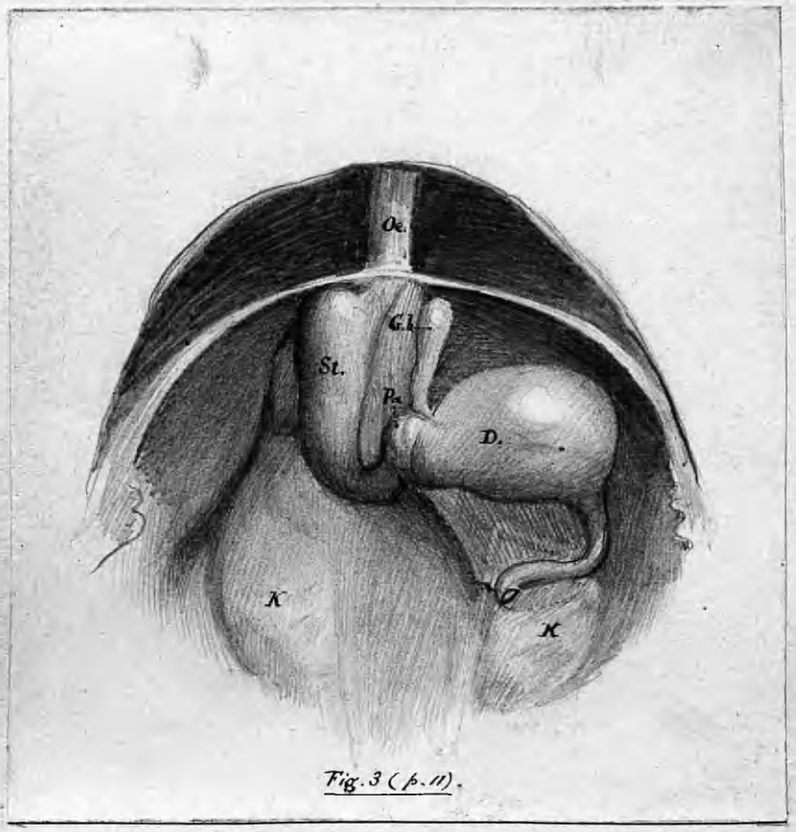


Fig. 3 (p. 11).

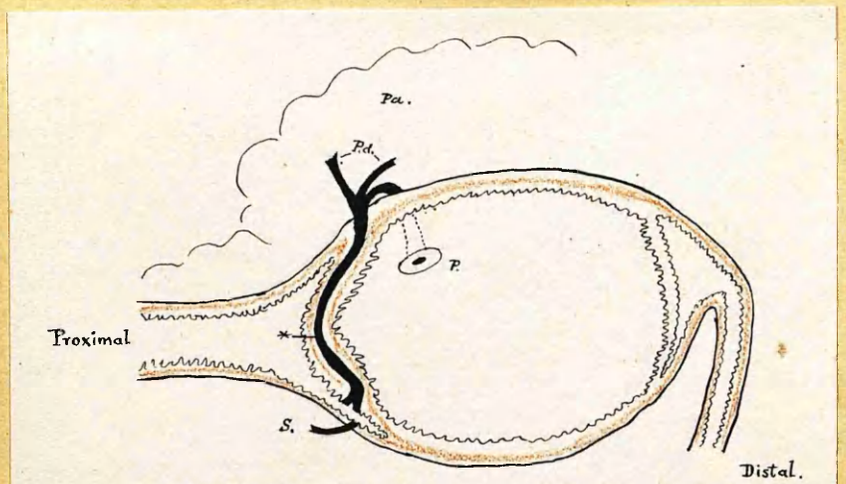


Fig. 4 (p. 14).

Diagram reconstructed from sections of duodenal Septa. The proximal is Funnelled by canal* leading from 1st part of duodenum up into communication with pancreatic ducts + or into Cyst via biliary papilla (P.) Pa. outline of Pancreas. P.d. ducts. S. duct of Santorini circular muscularis, in colour.

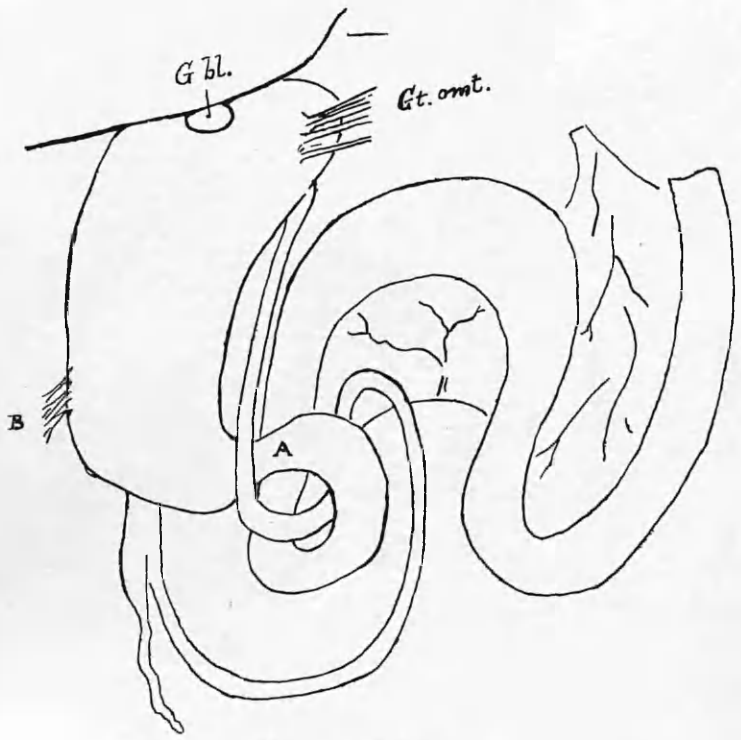


Fig. 5 (p. 28).

A. Seat of Volvulus. B. Adhesions – parietal; Gt. omt. – ditto to great omentum.

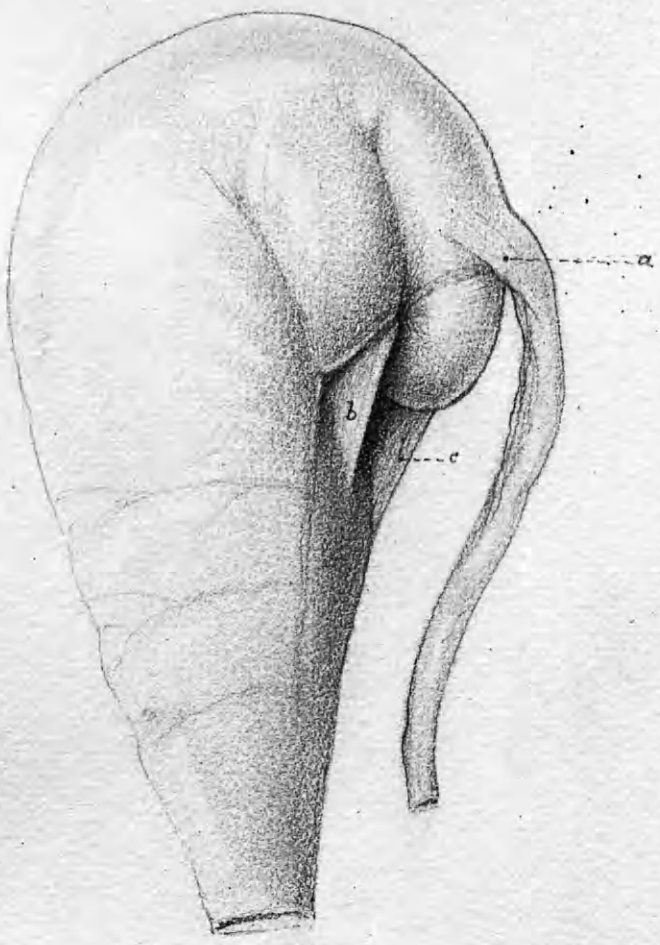


Fig. 6 (p. 29).

a. b. & c. - Adhesions. b is situated immediately behind the line of attachment of mesentery.

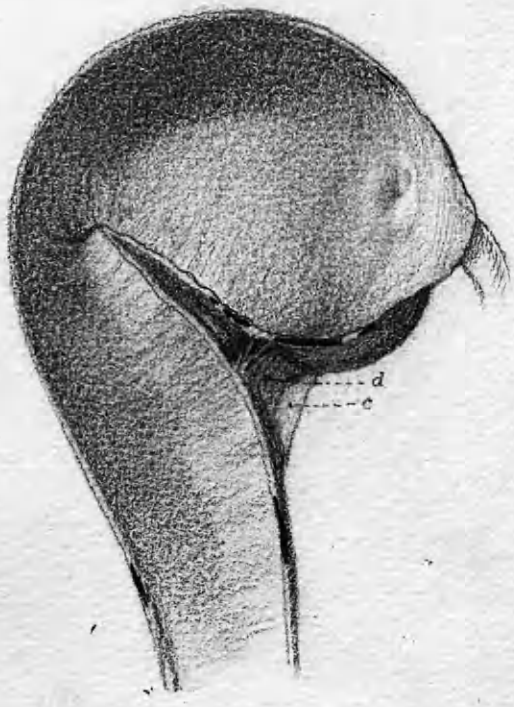
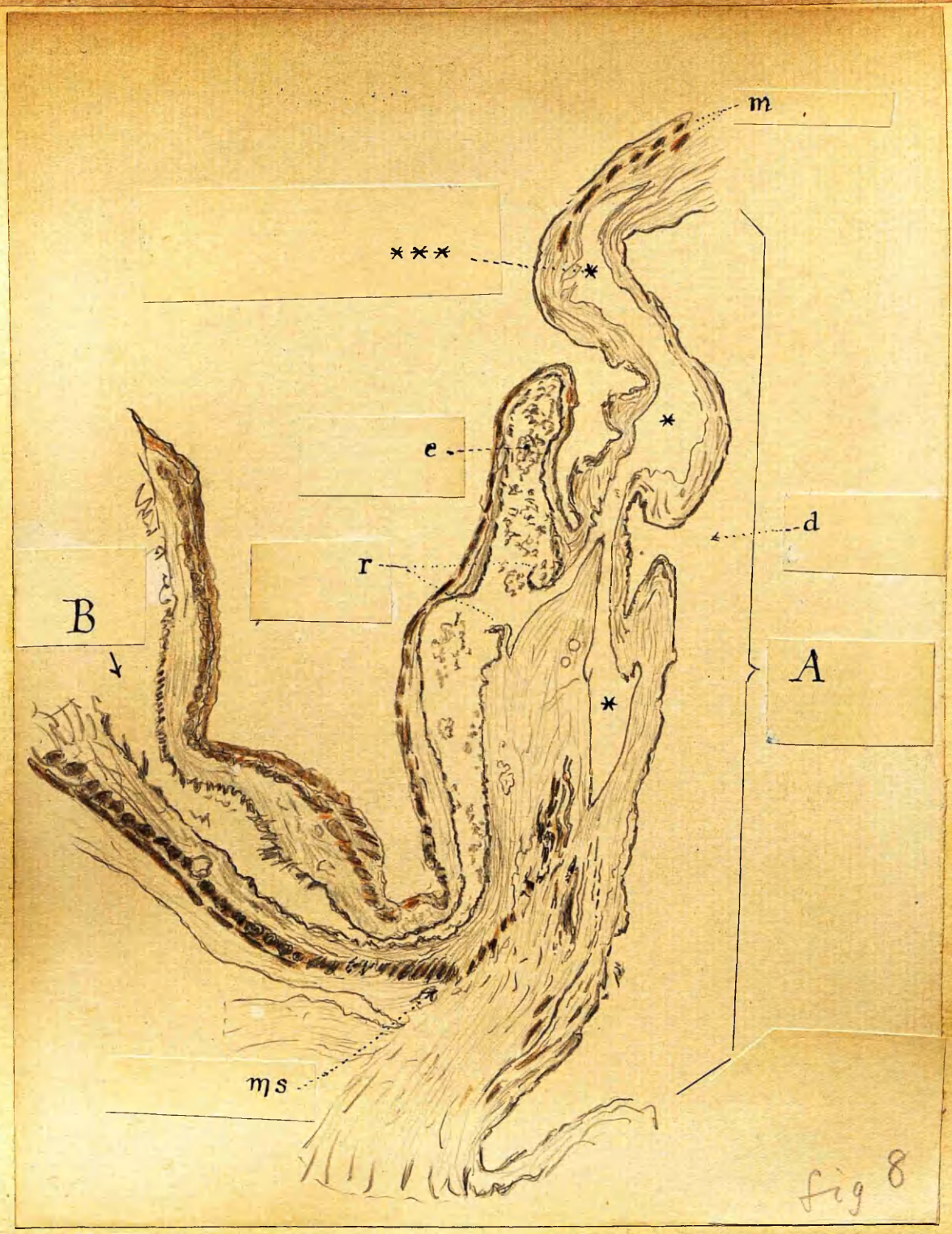


Fig. 7. (p. 30).

d. Adhesion assisting in maintaining spur. (Hidden by B. in fig. 6).

SEAT OF OCCLUSION, from behind; non-distended gut bent upwards;



A. Lumen of distended gut. B. Lumen of non-distended gut. ###. Cavity produced artificially by splitting of tunics of distended gut, when fluid injected into non-distended. d. Blind depression at region of the occlusion. e. Epithelial debris in cavity of non-distended gut. ms. muscularis entering spur. r. Rupture (artificial) of epithelium of non-distended gut.

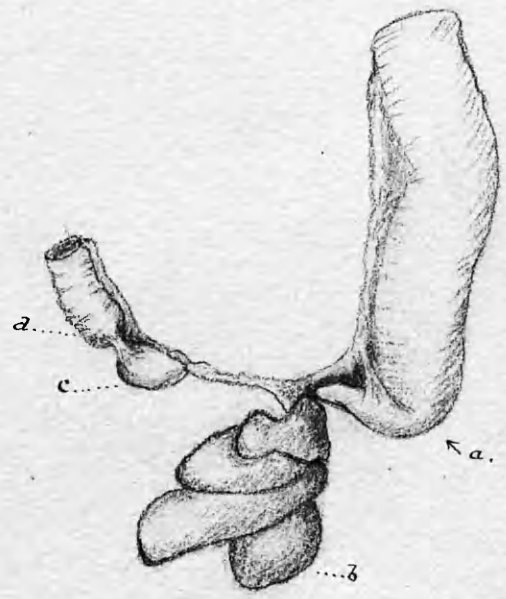


Fig. 9 (p. 33).

a. points to Kink,
b. Convoluted mass.
c. Ovoid body.
d. Upper end of non-distended bowel.

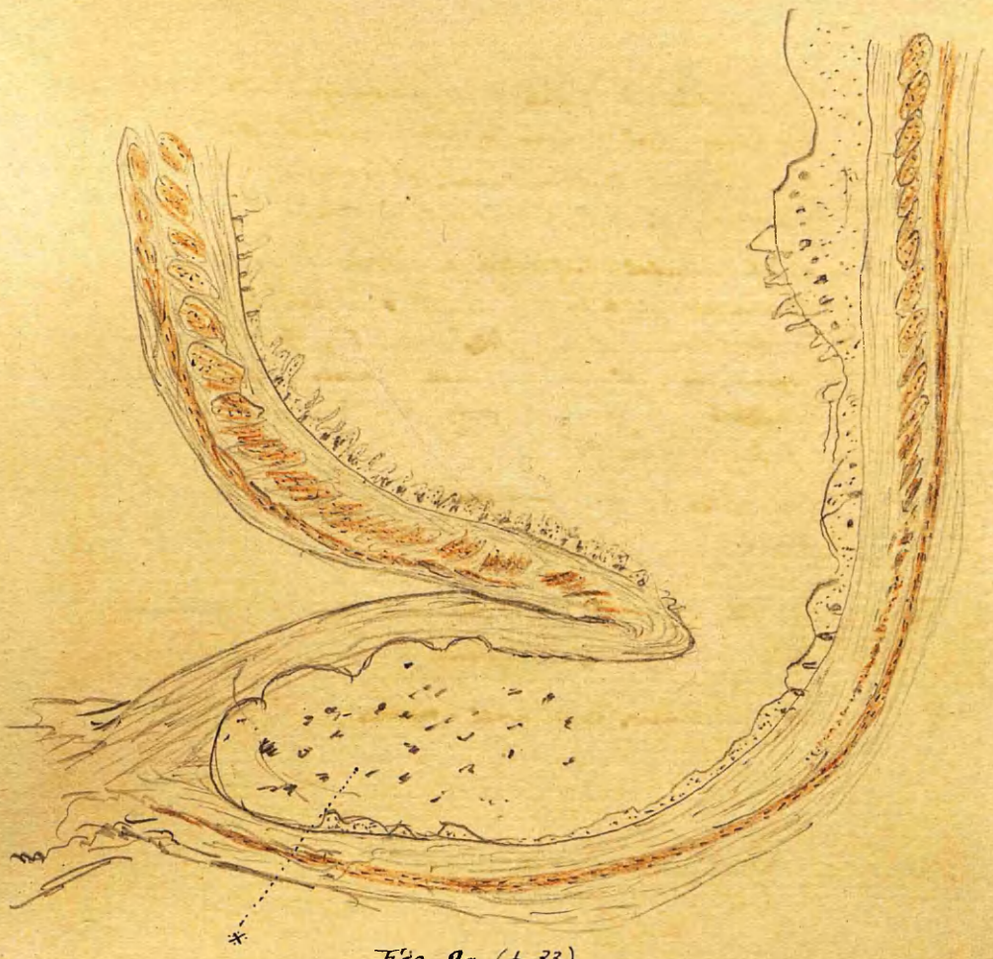


Fig. 9a. (p. 33).

Longitudinal Section at seat of kinking of gut. Shows defect of muscularis referred to on p. 34.
* Epithelial debris in stenosed gut, distal to kink.

Fig. 10 (p.35).
Showing occluded gut forming cervical mass. Note incomplete muscularis.

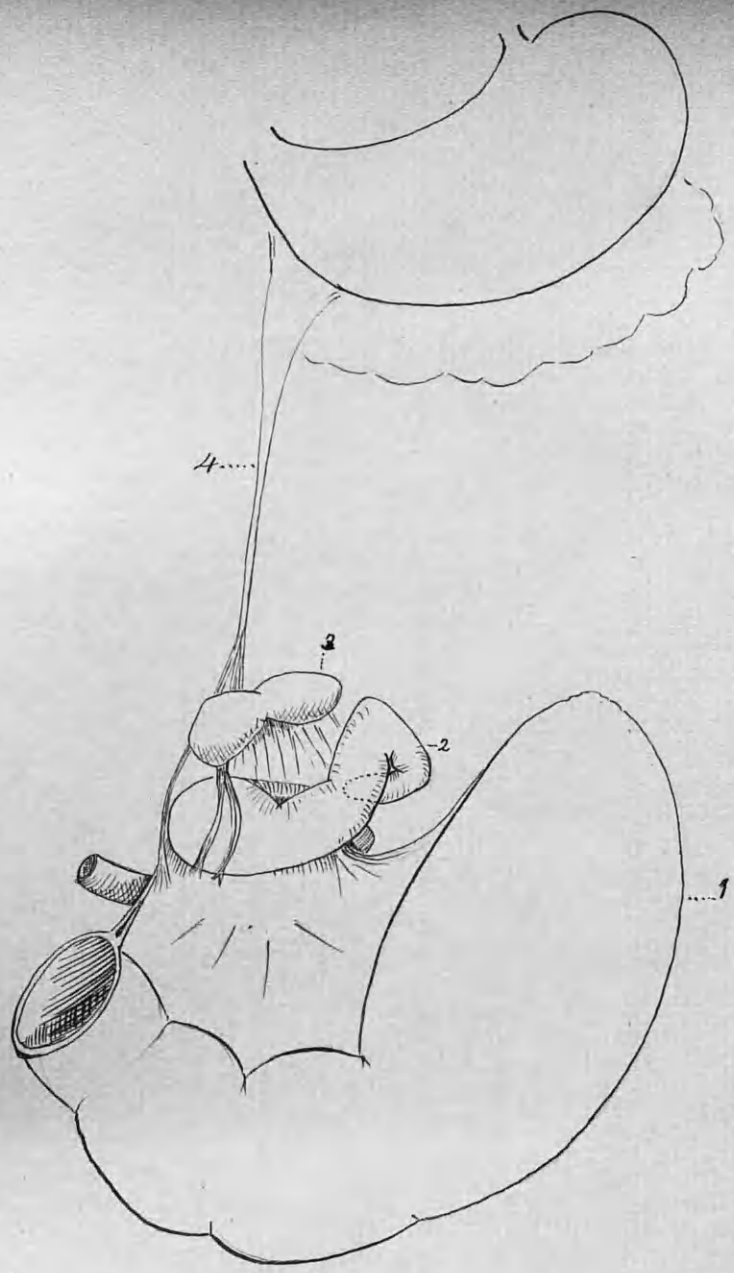


Fig. 11 (p. 37).

The relationship of the parts was distorted during hardening. The observer is looking at the Left side of the mesentery. The bowel-loop is inverted, so that the distal (collapsed) limb is between the stomach & the proximal (distended) limb.
1. Distended bowel; 2. non-distended bowel, distal to interruption; 3. beam-like body; 4. Omental band.

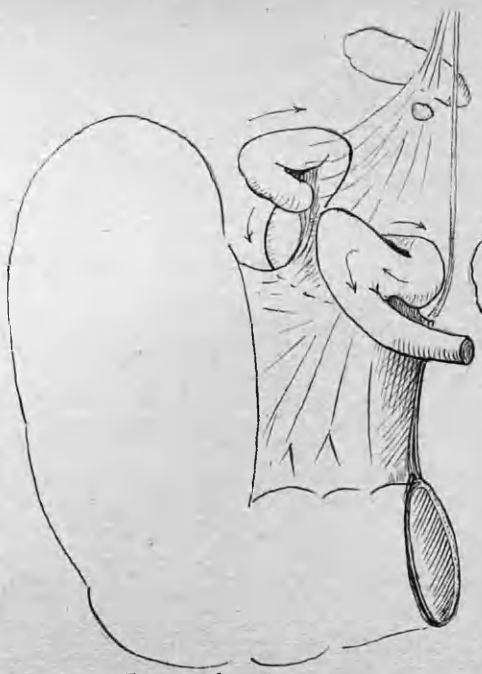


Fig. 12. (p. 37.)

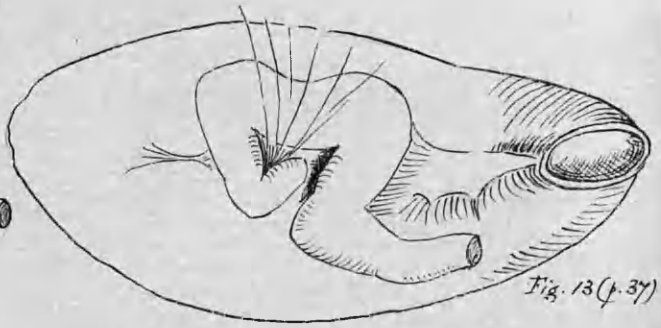


Fig. 13 (p. 37)

In fig. 12, parts viewed from right side. The mesentery (omitted from fig 11) is shown passing to blind end of non-distended limb. Arrows indicate direction of faecal current.

In fig 13, collapsed bowel viewed from below, ie. from surface opposite attachment of mesentery.

Fig 14 (p. 37)

Shows ridged mucous membrane giving place to granular surface at apex of blind end. At point indicated by x is "pit".

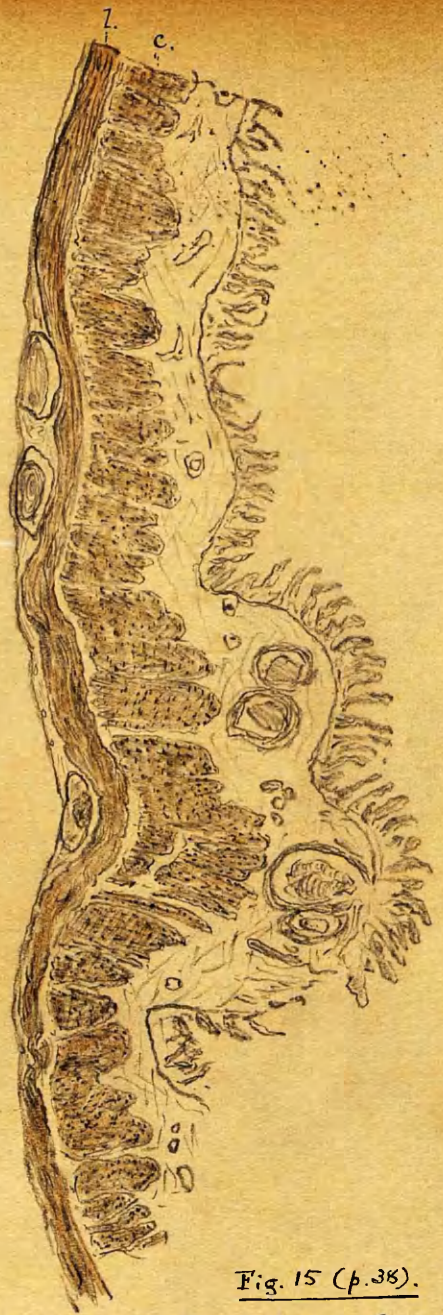


Fig. 15 (p. 38).

Wall of distended gut, some distance from blind End. Muscularis well marked. l. Longitudinal; C. Circular.

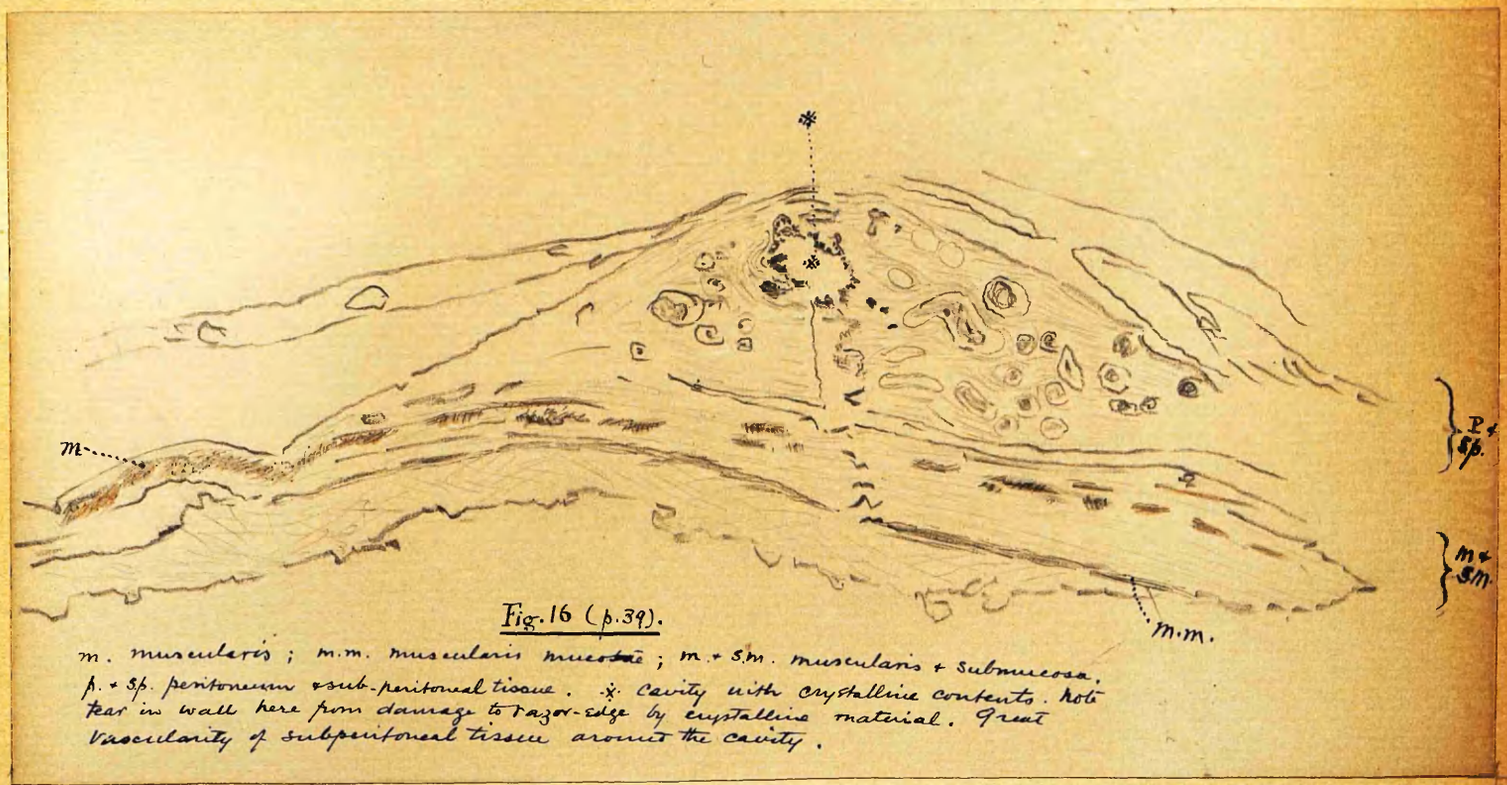


Fig. 16 (p. 39).

m. muscularis ; m.m. muscularis mucosae ; m. + S.m. muscularis + submucosa. P. + Sp. peritoneum + sub-peritoneal tissue. ✻ Cavity with crystalline contents. Note tear in wall here from damage to razor-edge by crystalline material. Great vascularity of subperitoneal tissue around the cavity.

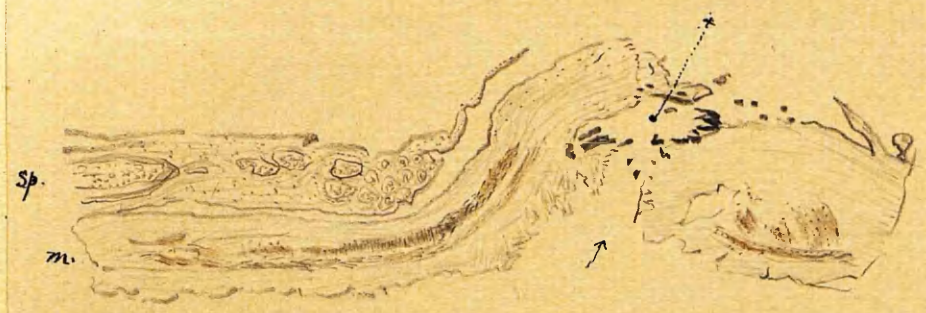


Fig 17 (p. 39)

Shows communication between "pit" (↑) + lumen of bowel. m. muscularis; s.p. subperitoneal coat (very vascular). * indicates cavity containing crystalline material.

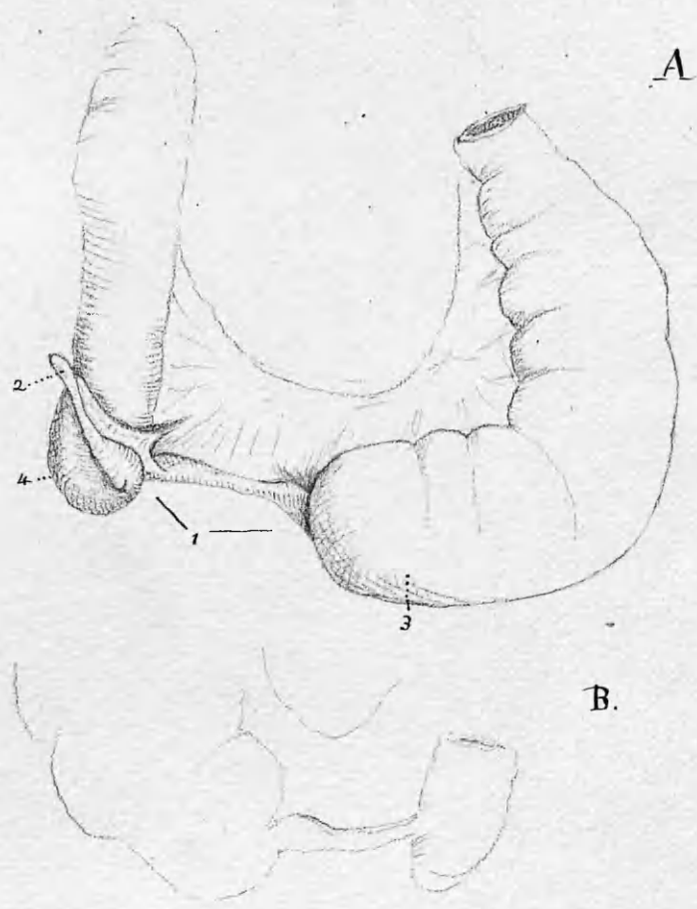


Fig. 18 (p. 41).

1. Cord-like communication between blind ends of ileum + Caecum
2. Appendix + meso-appendix
3. Ileum.
4. Caecum.

B. Appearance of parts from behind.

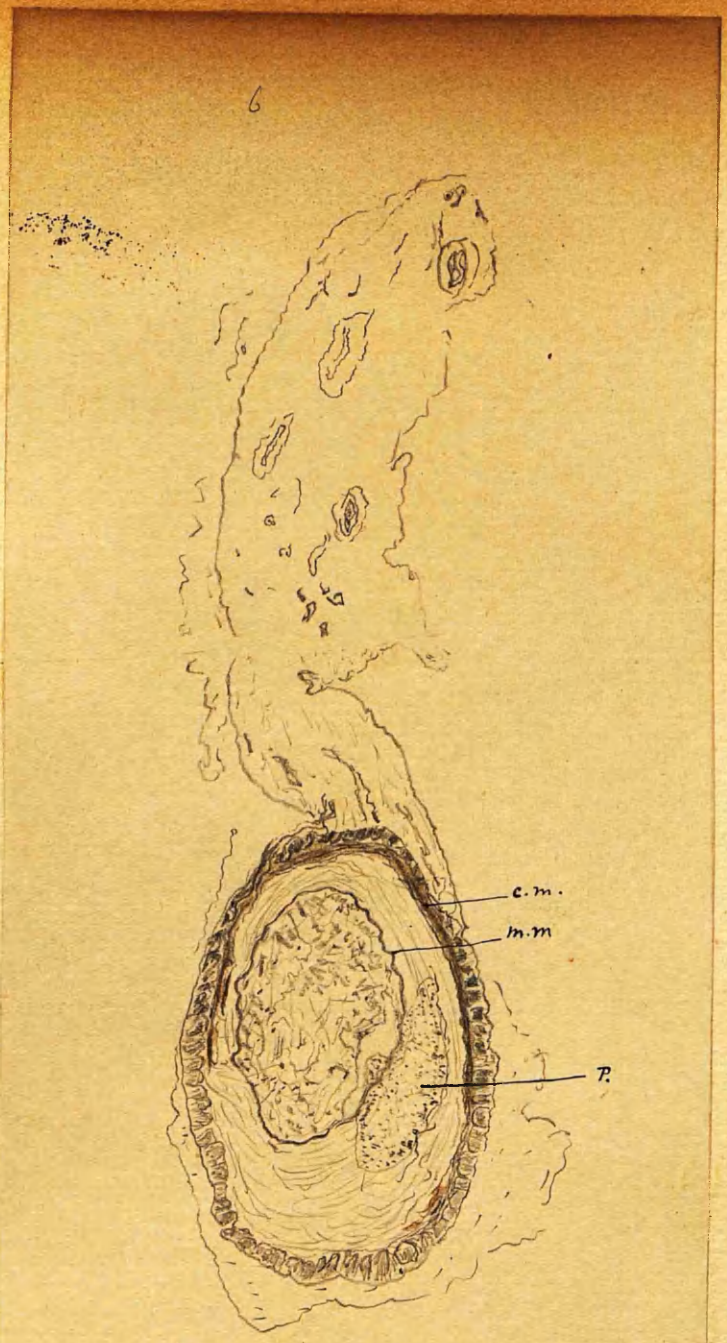


Fig. 19 (p. 42).

Section of "Cord" & mesentery, nearer cæcum than part showing mass of Epithelial débris. Lumen obliterated. P. - Peyer's patch. Circular muscularis is seen to be incomplete in distal portion of circumference.
c.m. Circular muscularis; m.m. muscularis mucosæ.

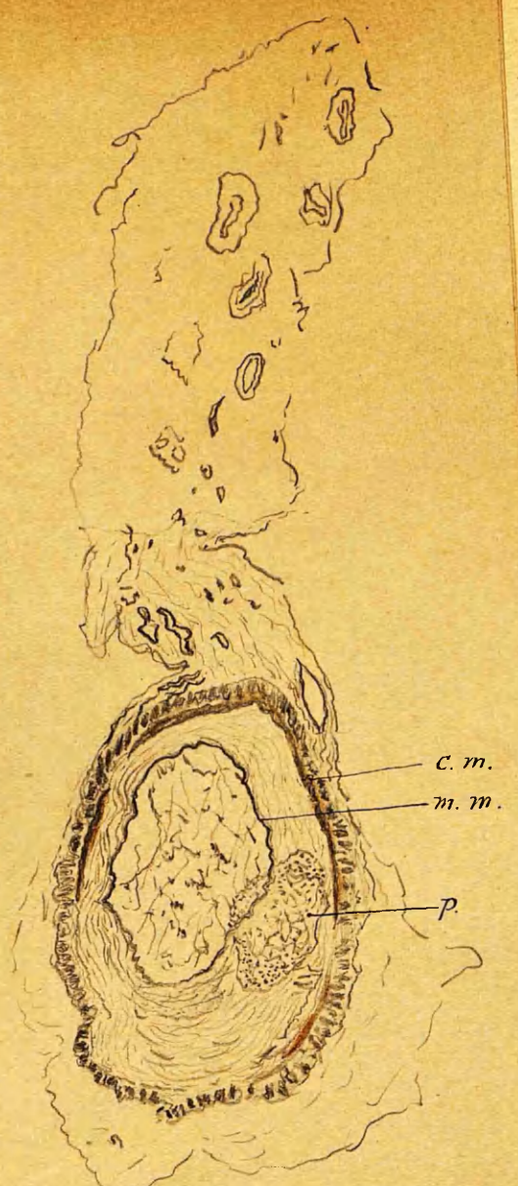


Fig. 19a.

Section a little nearer to cæcum than fig. 19. As in fig. 19, the Peyer's patch is partly within zone of muscularis mucosæ.