Congenital occlusion of the duodenum by E.M. Cowell. Reprinted from the Quarterly Journal of Medicine

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CONGENITAL OCCLUSION OF THE DUODENUM [1]

By E. MARSHALL COWELL

With Plate 19

Definition. By Congenital Occlusion of the Duodenum is meant a partial or complete interruption of the lumen of the bowel, usually situated in the vicinity of the opening of the bile duct. The occlusion may result from developmental errors or from antenatal disease.

Historical. The condition is somewhat rare. The earliest writing on the subject is by Aubéry in 1805. In 1812 Meckel quotes three fresh cases together with one case of his own. In 1837 Billiard, in his great work on children, describes a fresh case with a case described by Schäfer in 1824. In this year Rokitansky also fully describes a case of his own. Theremin, in 1877, writing on atresias of the alimentary canal, gives seven cases. He emphasizes the rarity of the condition: in 111,401 births there were two examples of the abnormality. In the lying-in hospitals of St. Petersburg 9 cases had occurred, but in Moscow and Prague the condition had never been found. Gaertner, in 1883, collected 16 cases and gives most of the literature up to date. Champneys, in 1897, showed a good specimen before the Pathological Society of London, but in his paper no extensive references are given.

The first recent paper which is at all comprehensive is by Cordes in 1901. He tabulates and analyses 57 cases with minute care. Kuliga, in 1903, collected 185 cases of stricture of the alimentary canal; in 59 of these the duodenum was affected. Operative treatment was mentioned by Ucke of St. Petersburg in 1907, but none of the latest papers have echoed his suggestion, and the operation of gastro-enterostomy has not yet been attempted. The papers mentioned are the chief ones on the subject; the most important are those by Cordes (57 cases) and Kuliga. At other times many observers have described isolated cases, so that now I have been able to collect 92 cases for analysis.

Clinical picture. This is best described by reference to the notes of a case recently observed.

The patient, a male child of 5¾ lb., was born on March 16, 1911, four weeks before full term. It seemed as well nourished as usual for its size. The parents were healthy; the mother had previously given birth to ten healthy children.

Within two hours of its birth the child was vomiting vigorously. The

[1] My best thanks are due to Dr. J. Poynton for permission to publish the case, and to Dr. Charles Bolton for many valuable suggestions.

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vomit was dark in colour from the first and consisted of 'coffee grounds' material. There was never mucus ejected in any quantity. A few hours later the bowels were well open and a large meconium stool passed.

Haematemesis continued for fifty hours at gradually increasing intervals. The infant became weak and finally died 52 hours from birth. The motions were normal—contained no blood—normal urine was also passed, and beyond small feeds, which were immediately vomited, no treatment was adopted. On physical examination, the abdomen was distended in the upper part; no mass was felt. In this case peristalsis was not visible.

No definite diagnosis was arrived at. Purpura neonatorum was negatived on account of these facts: (1) Vomiting began two hours after birth. Thirty to forty hours is the common time for haematemesis neonatorum to commence. (2) Absence of bleeding from bowel and skin or mucous membranes.

Post-mortem. The abdomen was somewhat distended. When opened the greatly distended stomach immediately attracted attention. The stomach and first part of the duodenum were greatly dilated. The pylorus was easily distinguished by a groove. The stomach preserved its usual outline, while the dilated duodenum formed a globular sac which pressed upwards on to the gall-bladder. Just above the opening of the common bile duct the spherical distension ended abruptly in a narrow cord. Beyond this the rest of the duodenum continued in a perfectly normal fashion. The pancreas, liver, and spleen were normal. The remainder of the bowel was quite normal, containing meconium. The meconium presented the usual microscopical appearance, except that no hairs were seen.

The following measurements were taken after hardening. A list of measurements in a thirty-six weeks child of the same weight is given for comparison.

Stomach.

Length from cardia to pylorus (great curvature)	16 cm.	Usual length	9 cm.
Maximum circumference	11·5 cm.	„ „	7 cm.
Circumference of pylorus	4·5 cm.	„ „	3·2 cm.

Duodenum.

Length from pylorus to occlusion (lower curvature)	10·2 cm.
Maximum circumference	11 cm.
Circumference of occluded portion	9 cm.
Circumference of second portion	2 cm.

On section the stomach and duodenum were distended with gas, but also contained about half a drachm of 'coffee grounds' material mixed with watery secretion. No bleeding-point was seen, but proper steps to find an ulcer were not taken. (The stomach was not opened and pinned out before hardening.) The first part of the duodenum ended in a little blind dimple. On dissection the common bile duct was seen opening coincidently with the pancreatic duct a short distance below the stricture. Two longitudinal folds of mucous membrane ran up the posterior wall of the duodenum. They increased in size and gradually diverged as they reached the neighbourhood of the bile papilla. At this point each fold became very marked and turned sharply outwards to meet the lateral walls of the duodenum. The folds presented free edges; arching outwards they practically met beneath the anterior wall, but left a small tunnel on the posterior wall. This led into a space which rapidly narrowed until the bowel was no longer pervious. A small fold in this space contained the opening of the bile duct. Section of this portion showed the whole diameter much diminished

and muscular walls increasing in thickness as the lumen diminished till the whole formed a small solid rod.

Length of strictured portion	1·1 cm.
Length of solid cord	0·5 cm.
Circumference	0·9 cm.

Analysis of Recorded Cases.

Cordes has made a careful analysis of the collected cases of his series. The results of the additional 35 cases are given below.

Sex. My figures show 8 male and 8 female, the sex is not mentioned in 19. In the whole series there are 21 males, 22 females, and 49 doubtful cases.

Frequency and nature of vomit.

Cordes's cases.		*Fresh series collected.*	*Total.*
Vomiting present	41	13	54
Not recorded	16	22	38
Blood, old or recent	27	7	34
Bile	2	2	4
Food	2	—	2
? Nature	10	4	14

Position of occlusion.

	Cordes.	*Fresh series.*	*Total.*
Above opening of common duct	20	11	31
Below opening	13	7	20
Level with	6	3	9
Not mentioned	18	14	32

Presence of other abnormalities in the alimentary canal.

None beside duodenal occlusion	54 cases.
Stenosis of other parts of bowel	8 cases.
Foetal peritonitis	3 cases.
Enlarged liver and spleen	1 case.

Pathology.

Position of occlusion. In the table prepared from the study of all the recorded cases the situation is accurately mentioned in 60. The common bile duct opens above in 31 cases, below in 20, and on a level with the occlusion in 20 cases. The Vaterian segment is the commonest portion of the duodenum affected. One would have expected this result, since this is the situation of so many important embryological events. Here the liver and pancreatic buds grow out, the lumen is at one time occluded, and the morphological fore-gut becomes the mid-gut.

Date at which occlusion occurs. In discussing the source of the stricture we shall see that certain changes occur before the second month of intra-uterine life which greatly help in the explanation of the formation of this condition. In a large number of cases in which the septum is below the common bile duct normal meconium is present in the bowel. This shows that the stricture was formed after the third month, when bile begins to be formed. In the present case the occluded portion measured 0·9 cm. in circumference, while the rest of the duodenum was 2 cm. There was no evidence of any gross disease having caused the difference in size. Supposing at this point this portion of duodenum ceased to grow, then by comparison with other foetuses we should be able to time the occlusion. Unfortunately I have not been able to obtain such data at present.

Source of the occlusion. There are two possible methods by which such an occlusion may arise: (1) By an error of development; (2) As the result of definite antenatal pathological processes. In the majority of cases no actual cause can be discovered, and an explanation is sought by means of developmental theories. Tandler has very carefully studied the development of the Vaterian segment of the duodenum in embryos of from thirty to sixty days. In his series the lumen of the gut is clear in the youngest and oldest embryos, but in the intermediate stages becomes blocked by a cellular proliferation. It is easy to see how a complete or partial failure of this plug to absorb can account for any grade of occlusion. The difficulty, if we accept this view of the pathogenesis of the condition, is to reconcile the dates of this cellular proliferation and the excretion of bile. In a small proportion of the cases a definite cause for the condition can be assigned.

Foetal volvulus. At one time this was the favourite explanation of the occlusion. Kutner (1857), in considering his case, dismisses volvulus and says there is no evidence for this explanation. Rokitansky, Schottelius, and Gaertner all quote cases, but the evidence is poor. Kuliga (1903) says that in one or two of the undoubted volvulus cases, the volvulus is the result and not the cause of the occlusion. Gross (1905) describes a very interesting case of a newborn infant with a mesoduodenum. The rotation of this portion of the bowel had not occurred, and the duodenum hung as a movable loop, so that no attempt at volvulus, however, was present. Claremont (1905) found in an adult a duodenum completely twisted on itself, but no obstruction was present.

Foetal peritonitis. Cases where the occlusion has undoubtedly been due to adhesions in foetal peritonitis are quoted by Theremin, Hirschsprung, v. Dohrn, Fielder, and Gaertner. Silbermann, in his study of foetal peritonitis, however, finds that it is very rare to see an associated stenosis of the bowel. Volvulus is sometimes associated with this condition; cases are described by Fielder, Wiederhofer, Schottelius, and Mauclaire.

Rarer causes of occlusion of the duodenum. These causes are so rare that they become merely pathological curiosities. Wyss and Hammer have described hypertrophy of valvulae conniventes causing obstruction. Baillie, in 1827, wrote of a case where he found in an adult a very large valvula connivens almost occluding the bowel. Hess met with a case apparently due to compression by the mesocolon. Nobiling found the bowel strangulated by a loop of the omphalo-mesenteric artery. Schott and Wünsche respectively found a cyst in the iliac fossa and an inguinal hernia pulling on the mesentery. Hirschsprung and Sever found cases where the head of the pancreas seemed press on the bowel. Wiederhofer described an antenatal carcinoma of the liver, and Kristella a large liver pressing on the bowel.

Why does the stomach dilate? The dilatation of parts above the stricture is generally a marked feature of the case. The pylorus becomes marked as a groove. Some of the earlier authors failed to recognize this fact, and spoke of a bilocular stomach. In cases where the condition is found in stillborn infants the stomach is full of mucus or watery fluid, while in the cases born alive large quantities of fluid are usually vomited from the very first. The presence of this fluid is probably the cause of the dilatation. The mucosa-lined muscular sac secretes fluid under pressure and dilates in just the same way as does a gall-bladder with obstructed duct. The dilated stomach may in fact be looked upon as a mucocele.

Source of the bleeding. Haematemesis occurs in about 65 per cent. of the collected cases. The exact cause of the bleeding, however, is not clear. In my case the presence of an ulcer cannot be absolutely excluded, since the stomach was not opened and pinned out at once after death. In the case of Schütz a tiny erosion was actually discovered and the opening into the vessel was found plugged with a recent clot. The entire portal system appears to be congested in a good many of the cases. In the photograph shown the vessels stand out clearly. No bleeding occurred from the mucosa below the stricture, however, while the dilated portion above was full of blood, so that simple portal congestion will not account for the bleeding. In the majority of cases there is haematemesis from the first. Occasionally, however, mucus is first vomited, and only later as the vomiting continues does blood appear. In the case of Funck-Brentano, the child vomited mucus, then material streaked with blood, and finally large quantities of blood. The conclusion is, therefore, that in most of these cases the gastrorrhagia is mechanical in origin, resulting from the strain of vomiting on an already over-distended stomach.

Diagnosis.

The points on which a diagnosis can be made are quite definite. *Vomiting.* In all the cases where vomiting is mentioned at all it is recorded as being present, so that in fifty-four cases vomiting was present; the point is not mentioned in thirty-eight. The vomiting usually begins at once after birth; the material is forcibly ejected and is got rid of in large quantities. *Haematemesis* occurs in about 65 per cent. of the cases. The bleeding differs from that of purpura neonatorum in that it usually comes on almost at once after birth, and is not

accompanied by melaena or other evidence of purpura. Although the vomiting is persistent and severe, yet usually the bowels are well open and normal meconium is passed. In occlusions of the bowel lower down constipation is usually absolute. *Of physical signs.* The upper abdomen is distended; sometimes peristalsis may be seen. Cyanosis is usually present, since the diaphragmatic action is so impeded. Bismuth given for X-ray diagnosis has not yet been tried, but should prove a useful aid in these cases.

Treatment.

The condition has been so little recognized that but scanty opportunity for treatment has arisen. In the few cases where a diagnosis has been made, lavage has been tried. Five cases have been subjected to laparotomy, those of Wyss, Simmonds, Hess, Kirmisson, and Schütz. Enterostomy has been attempted in each of these cases, but in none has the infant survived more than a few hours. Ucke of St. Petersburg, in 1907, suggested gastro-enterostomy, but this has not yet been carried out. As far back as 1899 Abel performed a successful gastro-enterostomy on an eight-weeks-old infant for hypertrophic pyloric stenosis (the time of operation was forty minutes). So that with the present-day improvements in technique this operation should be at least considered justifiable to attempt. In more than half the cases available for analysis the opening of the common bile duct was below the stricture, so that in a gastro-enterostomy there is not a great risk of trouble from this source. In about ten per cent. of the cases some other deformity exists, as multiple stenosis of small intestine, absence of anus, and so on. In these cases the clinical picture characteristic of occlusion of the duodenum, vomiting normal action of the bowel, does not obtain. The infants live a variable time with complete occlusion. Death usually takes place on the fourth or fifth day. The longest life is nine months (quoted by Keith). With partial stenosis one case lived as long as eighteen months (Buchannan). From the consideration of these facts it would seem that the cases should be subjected to laparotomy, and if possible gastro-enterostomy should then be performed.

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DESCRIPTION OF FIGURES.

FIG. 1. View from the front, showing dilated stomach and duodenum.

FIG. 2. Tracing from photograph of anterior view of viscera. The size of a normal stomach and duodenum is represented by the dotted line.

FIG. 3. View from behind entrance of bile and pancreatic ducts just below occlusion. D^{I}. First portion of duodenum. D^{II}. Second portion of duodenum. Between these two parts is seen the bile duct B opening just beyond the stricture.

FIG. 4. Diagram of site of atresia magnified about three times. (*a*) First portion of duodenum in blind pit. (*b*) Cord-like portion above opening of common bile duct. (*c*) Normal second portion, with longitudinal folds. (*d*) Common opening of bile and pancreatic ducts.

FIG. 5. Diagrammatic sections taken from below upwards (×3). I. The two folds. II. Turning outwards before reaching the bile duct. III. The bile duct. IV. Just above bile duct.

FIG. 3

FIG. 2

FIG. 1

FIG. 5

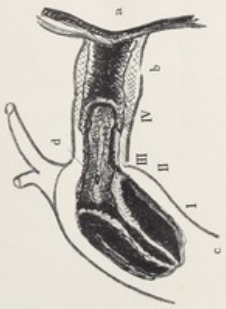

FIG. 4