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Etiology of Congenital Intrinsic Duodenal Obstruction
—With Special Reference to the T-shaped Bile Duct Anomaly—

by

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Much remains unknown regarding the clinical pathology and etiology of congenital intr insic duodenal obstruction (atresia, stenosis). As this condition is, in general, associated frequently with other abnormalities, the view is predominant that it is an embryological developmental anomaly. In contrast, it is widely recognized, both clinically and experimentally, that jejuno-ileal atresia develops secondarily to accidental injury in early fetal period. Lately, we experienced 2 interesting cases of duodenal atresia possessing abnormal entrance of the bile duct and pancreatic duct into the separated duodenum. As this was demonstrable by radiography, the cases are herein reported, together with a discussion on the etiology.

Case report

Case I : A male infant weighing 2.6kg with Down's syndrome was admitted immediately after birth due to massive bile stained vomiting. Plain roentgenogram of the abdomen revealed a typical double bubble sign. On the 2nd day he was transported to our hospital in a state of shock following massive hematemesis of fresh blood and marked abdominal distension.

The double bubble sign could no longer be seen by radiography, and the pneumoperitoneum was demonstrable. Blood transfusion, fluid replacement with sodium bicarbonate and peritoneal tap for decompression of the abdominal cavity were immediately conducted, but as the bleeding continued, emergency surgery was performed. The operative diagnosis was gastric rupture in association with duodenal atresia. As the general condition was extremely poor, the atretic part of the duodenum were not investigated in detail, a side to side duodenojejunostomy (shunt anastomosis) and a two layered suture of the gastric rupture were performed. The postoperative condition deteriorated further and the patient died 12 hrs later.

Necropsy findings ; An annular pancreas was seen at the 2nd part of the duodenum which was markedly distended above the panceatic ring. When the duodenum with annular pancreas was incised longitudinally, the lumen was separated completely beneath

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the pancreas.

A papilla was recognized in the upper pouch of the blind end, and by introducing a thin probe into it, the existence of a fine duct communicating with the distal pouch was confirmed. The common bile duct was found to open at approximately the central part of this duct to present a T-shaped figure. The pancreatic duct was not distended and opened into the central portion of the T-shaped duct. (Fig. 1).

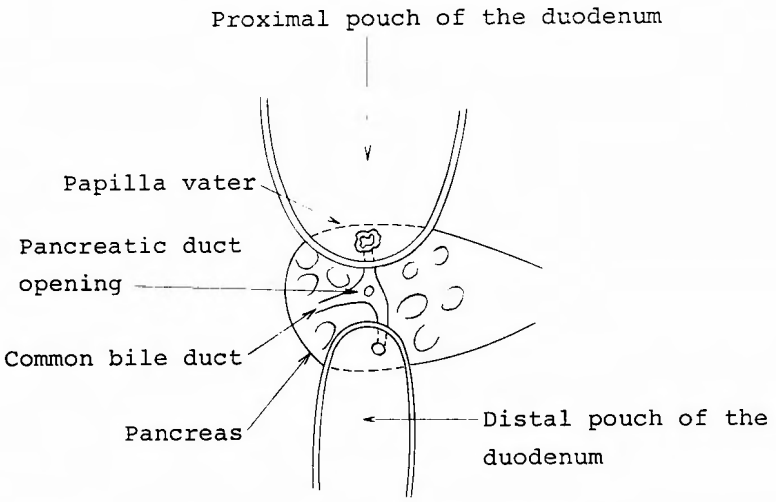

Fig. 1. Schema of necropsy finding (case 1)

Microscopic findings; In order to clarify further these complicated anatomical constitution, gross serial sections were prepared and examined histologically. The duct which opened into the proximal pouch was found to be composed of the epithelium of the bile duct and formed the papilla (Fig. 2). In the distal pouch there was seen no clear histological structure of a papilla, but an opening of a duct composed similarly of the bile duct epi-

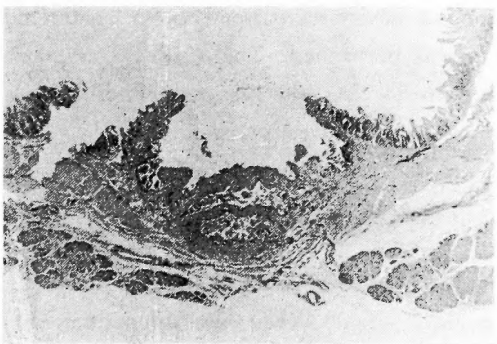

Fig. 2. The microscopic finding of the proximal pouch. The papilla was recognized at the central portion.

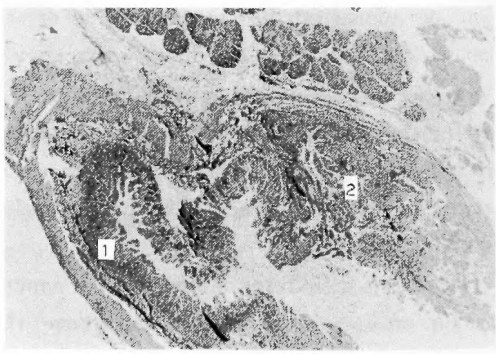

Fig. 3. The microscopic finding of the distal pouch (1). An opening of a duct composed of the bile duct epithelium (2) was recognized.

thelium was recognized (Fig. 3). In these lesions cell infiltration, fibrosis and calcification, indicative of necrotic or inflammatory changes could not be recognized. In other words, pathological findings arising from ischemic processes noted in jejunoileal atresia could not be seen.

Case II : This infant was adminitted immediately after birth with respiratory distress syndrome. He was premature infant weighing 1400g at birth who was diagnosed congenital duodenal obstruction with Down's syndrome. Due to this anomalies, the consent of the parents could not be obtained and the patient died shortly after without surgery.

Necropsy findings revealed, as in the Case I, a separated type of atresia at the 2nd part of the duodenum, but it was not associated with annular pancreas. By opening the distal and proximal pouch of the atretic segment, there was recognized macroscopically the presence of distinct papilla in both, and to flow out the bile through the both papillas by oppressing the gall bladder.

Then radiographic study was performed with injection of contrast medium into the gall bladder, and it was proved a very interesting cholangiogram, as shown in the Fig. 4. that the bile duct opened in a T-shaped pattern into the both blind pouches of the duodenum. The opening of the pancreatic duct was similar to that of the Case I, and the pancreas showed no abnormal findings. The liver and the gall bladder were normal in both Case I and II.

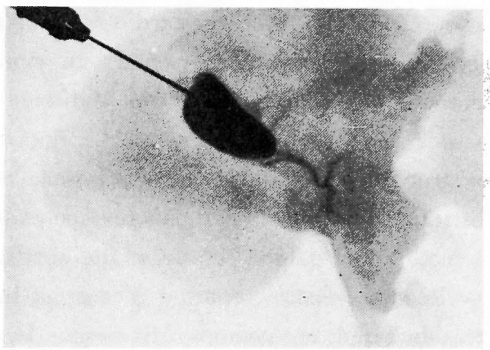

Fig. 4. The cholangiogram of the Case II. The bile duct opened in a T-shaped pattern into the both blind pouches of the duodenum.

Discussion

During the past 10 years, 16 cases of congenital intrinsic duodenal obstruction (atresia and stenosis) were experienced at the 1st Department of Surgery, Nagoya City University Medical School. Of these, 8 cases developed at the duodenojejunal flexure, 7 at the 2nd part of the duodenum and 1 at the 3rd part. As shown in table (1), those developing in the duodenojejunal flexure rarely had associated anomalies. In contrast the 8 cases developing in the 2nd and 3rd parts of the duodenum, were characterized, with the exception of 1 case, by the presence of 1 or more than 2 associated anomalies. For example, the 2 cases reported here were accompanied by biliary and pancreatic duct anomaly and Down's syndrome, and there were 2 cases with biliary atresia (of which 1 was a gap type atresia with malrotation), 3 with associated annular pancreas and other 3 with associated cardiac anomaly.

From these findings, it can be stated that duodenal atresia of the 2nd and 3rd parts is characterized by associated developmental abnormalities of the neighbouring organs. NIXON[2], WILKINSON[3] and MOORE[4] consider the association of annular pancreas, malrotation

Table. 1. Type of intrinsic duodenal obstruction and associated abnormalities in our 16 cases

Site of obstruction	No. of cases	Type of obstruction	Associated abnormalities	Prognosis
2nd part of duodenum	7	Gap atresia 4 Membranous 3	Down's syndrome 2 Bile & pancreatic duct anomaly 2 Biliary atresia 2 Annular pancreas 2 Cardiac anomaly 3	Died 4 No surgery 1
3rd part of duodenum	1	Membranous 1	Annular pancreas 1	
Duodeno jejunal flexure	8	Membranous 8	Cardiac anomaly 1 Down's syndrome 1	Died 2

and biliary atresia as most important, but in general the association of major anomalies such as Down's syndrome, cardio-vascular anomaly, esophageal atresia and anorectal malformation have been often reported. In contrast, the presence of abnomalities in the bile duct and the pancreas has not sufficiently been considered. For this, the following 3 reasons may be pointed out : (1) the duodenum are very complicated anatomically in its structure and position. (2) existence of many cases where both extrincic and intrinsic obstrution due to annular pancreas co-exist, (3) due to the performance of by-pass surgery in most cases, detailed study of the obstructive leasion is seldom undertaken.

Recently, REID[5] reported 9 cases of biliary tract abnormalities associated with duodenal atresia, based on necropsy findings. It would seem that the frequency of associated abnormalities of the pancreas and biliary tract system will be much greater than that so-far reported. Among the cases of REID[5] and GOURVITCH[6] were those duodenal atresia with T-shaped bile duct reported here. Further, there were also cases with associated biliary tract abnormalities reported by BOYDON[1], Karpa and Katz.

Regarding the etiology of congenital intestinal obstruction, there is the well-known theory of disturbance of recanalization (developmental anomaly) presented by TANDLER[7] in 1900.

However, since LOUW and BARNARD[8] published the vascular accident theory in 1955, it is now widely recognized that this secondary ischemic necrosis is an important factor in the cause of intestinal obstruction. However, as etiology of the membranous atresia developing frequently in the duodenum and duodenojejunal flexure Tandler's theory seems more appropriate.

Further, BOYDEN[1] et al. considered the abnormalities during the complex developmental stage of the duodenum in early embryonic period as etiology of this condition. They stated that at the stage of vacuolization, the duodenum became divided into two channels and the bile and pancreatic duct opened into both lumen. If some abnormality occurred at such a stage, the occurance of duodenal atresia associated with the T-shaped bile duct could be expected. Also, at this stage there will be rotation of the duodenum, ventral

pancreas and bile duct, in cases of obstruction of the 2nd part of the duodenum, there will be expected the association of developmental abnormalities of neighbouring organs.

In Case I reported here, there appeared an associated annular pancreas, but necropsy findings revealed a separated type of duodenal atresia. Cases of annular pancreas characterized by vomiting in the newborn seem to be all those with associated intrinsic obstruction.

Further, in a case recently experienced of duodenal stenosis due to annular pancreas occuring in a adult, there was recognized the existence of a web in the lumen. Namely, during the operation (duodeno-duodenostomy) a study of the proximal pouch revealed the existence of a web beneath the annular pancreas that was the main cause of the stenosis.

However, it cannot be denied that there are some cases of duodenal atresia believed to be due to accidental injury. One of our cases of duodenal atresia with congenital biliary atresia showed complete separation at the 2nd part of the duodenum and distal segment presented a so-called Christmas tree pattern. There was no formation of duodenojejunal flexure. This case is believed to be a gap atresia caused by accidental injury resulting from abnormal intestinal rotation. In the report of FONKULSRUD (1969)[9] on 350 cases there are 6 cases of duodenal atresia due to vascular accident.

Sammary

Among cases of congenital duodenal atresia of the separated type, there were experienced 2 cases with the common bile duct and opening in a T-shaped manner into the upper and lower pouches. This was confirmed by postmortem cholangiography. There are interesting cases indicating the etiology of duodenal atresia to be due to developmental anomaly occurring in early embryonic period, and endorse the etiological theory of BOYDEN and BILL[1].

The contents of this paper were reported at the 5th Annual Meeting of the Pacific Association of Pediatric Surgeons, 1972, Tokyo[10].

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和文抄録

先天性内因性十二指腸閉塞症の成因

——特にT字型胆管開口異常例について——

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由良二郎，高橋英城，柴田清人

教室で経験した16例の内因性閉塞症について検討した結果，十二指腸第2部の閉鎖例では，胆管の異常，輪状膵を合併する頻度が著しく高く，総胆管が離断した上下の盲端にT字型に開口する特殊な症例を2例経験した．これを死後の胆管造影により描写し得たので報告した．本症は胎生早期における十二指腸の発生が，胆管，膵管の開口をともなう複雑な形成過程をとるために生ずる発生異常と解され，Boyden, Bill の成因論に賛意を表するものである．