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Pediatric Annular Pancreas : A 12-Year Experience

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Abstract

Annular pancreas is a rare congenital anomaly that consists of normal pancreatic tissue arranging in a continuous ring around the descending duodenum. This paper reported an experience of 52 pediatric patients with annular pancreas and duodenal obstruction who were treated at the Surgical Department of the Children's Hospital during 1985-1996. Thirty-five were male and seventeen were female. Age at onset of symptom ranged from 1 day to 20 months. Vomiting was the most common presentation in these patients. Jaundice and maternal polyhydramnios were noted in 50 and 21 per cent respectively. All underwent surgical treatment and duodenoduodenostomy was the most common procedure performed. Complete duodenal obstruction with annular pancreas was noted in 35 of 52 cases and their survival rate was 74.3 per cent. Incomplete duodenal obstruction with annular pancreas was found in 17 of 52 patients and the survival rate was 88.2 per cent. The overall survival rate was 78.9 per cent. Congenital heart disease, pulmonary complication, prematurity and septicemia were among the major causes of death.

Our experience from this study suggested that the high incidence of associated serious congenital anomalies resulted in the high mortality in these patients. Duodenal bypass by duodenoduodenostomy was the most popular and effective procedure for correction of duodenal obstruction with annular pancreas.

Annular pancreas is an uncommon congenital anomaly in which pancreatic tissue encircles in a partial or complete ring around the second or third part of duodenum. This condition may manifest symptoms of duodenal obstruction in infancy, childhood or adult life. Most of pediatric patients develop the clinical presentation in neonatal period. Tiedeman was credited for the first description of this congenital anomaly in 1819 and "annular pancreas" was named by Ecker in 1862.[1] The first successful operation for correction of duodenal obstruction with the annular pancreas by a gastrojejunostomy was performed by Vidal in 1905.[1-3]

Since then, many authors reported successful treatment of this anomaly in the world's literature.[4-7] Our recent experience of duodenal obstruction with annular pancreas is reviewed and presented herein.

Materials and Methods

A review of the operations that were performed for correction of duodenal obstruction in the Surgical Department of Children's Hospital during January 1985 to December 1996 was carried out. Fifty-two cases were found to have duodenal obstruction with annular pancreas. The clinical data were collected for evaluation of the clinical presentation, operative procedures and eventual outcome.

Results

General incidence

During a 12-year period, 212 pediatric patients underwent laparotomy for treatment of congenital duodenal obstruction. Fifty-two cases (24.5%) were noted to have annular pancreas associated with duodenal obstruction (Table 1).

Of the 52 patients, 35 were male and 17 were female. The male to female ratio was 2:1. Thirty four (65.5%) were premature babies. Age at onset of symptom varied from one day to 20 months. Forty-seven infants (90.4%) developed symptoms of high gut obstruction within one week after birth. Only 7 cases were diagnosed beyond the age of one month.

Symptomatology

More than one half of the patients developed vomiting in the first oral feeding. Bilious vomiting occurred in 44 cases (84.6%) while non-bilious episodes were found in 4 cases (7.7%). There were 4 babies who had no history of vomiting because of the association with esophageal atresia in 3 and imperforate anus in 1. Jaundice, epigastric distension and maternal polyhydramnios were presented in 50, 42.3 and 21.2 per cent respectively (Table 2). Five neonates had drowsiness with convulsion due to electrolyte imbalance and sepsis.

Associated congenital anomalies

Other congenital anomalies were noted in 28 of the 52 patients. Down syndrome was the most common anomaly in this study (Table 3). Congenital heart disease was found in 8 patients and ventricular septal defect (VSD) was present in all of cases. Esophageal atresia, vertebral anomalies and Meckel's diverticulum were noted in 3 cases each.

Radiological manifestation

Abdominal x-rays revealed the typical "double-bubble" sign in 35 cases (67.3%). The remaining 17 cases had dilatation of the stomach and the first part of duodenum with minimal gas in distal small intestine. Contrast study of upper gastrointestinal tract was performed in all of the 17 patients and it demonstrated incomplete obstruction of the second portion of duodenum in 14 and the third portion in the other 3 (Figure 1). Two of the 17 patients had abnormal location of the duodenojejunal (DJ) junction and the proximal jejunum was on the right side of abdomen. This finding indicated malrotation of the intestine.

Treatment

All of 52 patients were operated upon. A transverse incision was performed in the right upper quadrant of abdomen in all except 2 cases, who had been preoperatively diagnosed as malrotation. A midline incision was used in these 2 cases. A diamond-shaped

Table 1 Correlation of duodenal obstruction and annular pancreas in 52 patients.

Type of duodenal obstruction	No. of patients	Death (cases)
Complete obstruction		
1. Duodenal atresia with annular pancreas	34	8
2. Duodenal atresia with annular pancreas and malrotation	1	1
Incomplete obstruction		
1. Duodenal stenosis with annular pancreas	15	2
2. Duodenal stenosis with annular pancreas and malrotation	2	0

Table 2 Clinical presentation in 52 patients

Symptomatology	No. of patients	Per cent
Vomiting	48	92.3
Jaundice	26	5.0
Epigastric distension	22	24.3
Maternal polyhydramnios	11	21.2
Dehydration	10	19.2
Neurological disorders	5	9.6

Table 3 Associated congenital anomalies in 28 patients

Associated anomalies	No. of patients
Down syndrome	11
Congenital heart disease	8
Esophageal atresia	3
Vertebral anomalies	3
Meckel's diverticulum	3
Hypospadias	2
Undescended testis	2
Limb anomalies	2
Imperforate anus	1
Subglottic stenosis	1

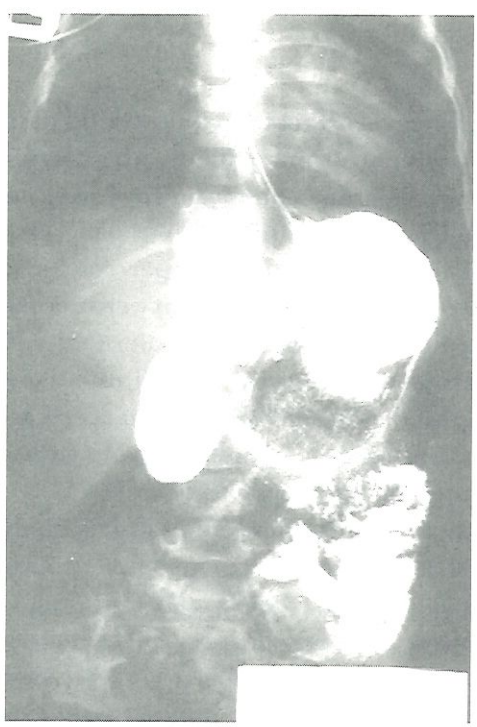

Fig. 1 Contrast study showed incomplete obstruction at the second part of duodenum.

duodenoduodenostomy by Kimura's technique[8] was performed in all of them (Figure 2). Three cases with malrotation had both duodenoduodenostomy and Ladd's procedure.

Twenty six of the 35 patients (74.3%) who had association of duodenal atresia and annular pancreas survived, while 15 of the 17 patients (88.2%) who had duodenal stenosis with annular pancreas did so. The overall survival rate was 78.9 per cent.

Common complications were pneumonia, septicemia and heart failure. Three infants had anastomotic leakage and they were treated conservatively by NPO, placement of a nasogastric tube with continuous suction, antibiotics and peripheral parenteral nutrition. Two of these 3 cases died because of sepsis, Two infants developed hemorrhagic gastritis and one had chylous ascites postoperatively. All of these 3 patients recovered well after conservative management for a peroid of time.

Discussion

Many theories had been suggested to explain the embryogenesis of annular pancreas but Lecco's theory was probably the most acceptable.[1,9] In the embryonic period, pancreas developes as dorsal and ventral outpouchings from duodenum about the fourth week of gestation. In the normal course of development, the ventral pancreatic anlage with the duct of Wirsung is rotated dorsally around duodenum and fuses with dorsal pancreatic anlage, which is drained by the duct of Santorini. The two duct systems anastomose and the ventral duct of ventral anlage persists together with the distal portion of the dorsal anlage duct to form the main pancreatic duct. According to Irving,[9] Lecco postulated that fixation of the tip of the ventral anlage to the duodenal wall results in the anlage being drawn around the right side of the duodenum and it is completely encircled by the annular pancreas (Figure 3).

Some authors[6,9-11] believed that the annular pancreas was not the cause of duodenal obstruction. In some instances, there was no duodenal obstruction but the annular pancreas was incidentally found at laparotomy of other disease. Mc Naught in 1933 and Stofer in 1944 reported this anomaly in autopsy cases of incidental findings in adults.[3] It becomes clinically significant when associates with duodenal obstruction.

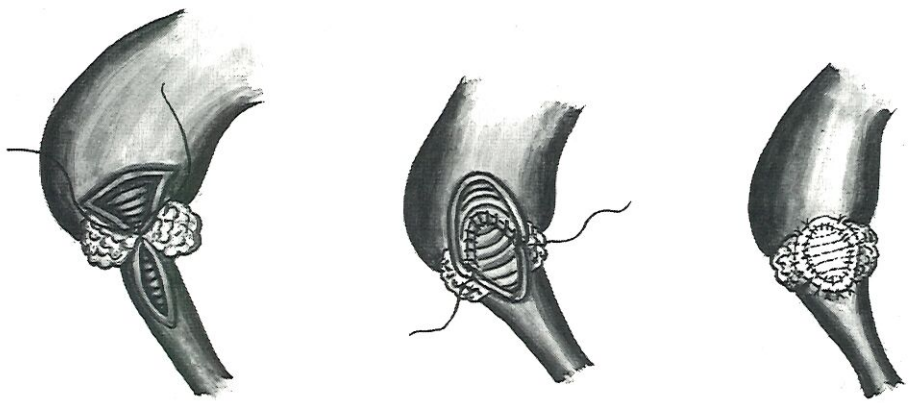

Fig. 2 Kimura's duodenoduodenostomy (diamond-shaped anastomosis)

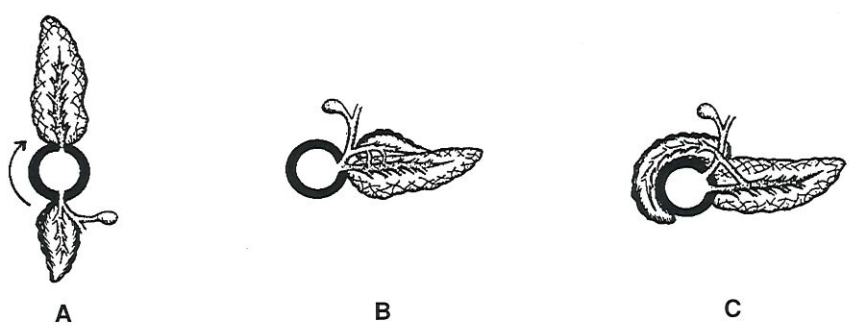

Fig. 3 Diagrammatic representation of normal migration of ventral pancreatic anlage (A and B) and failure of rotation as Lecco's theory (C).

There is a constricting ring of the aberrant glands encircling the second or third part of duodenum. It was often classified as a cause of extrinsic obstruction of duodenum[12-15] but many pediatric surgeons classified annular pancreas as an intrinsic obstruction because pancreatic tissue was microscopically demonstrated within the duodenal wall in many cases.[3,11,16,17] This was also found in about 10-20 per cent of all congenital duodenal obstruction.[13,18] In our study, we found in about 25 per cent of all duodenal obstruction and the annular pancreas was found to be associated with atresia more often than stenosis.

Clinical presentation is determined by degree of duodenal obstruction. Most of the patients in this study developed symptomatology in the neonatal period and many of them had a bilious vomiting before or during the first feeding. Few cases had incomplete duodenal obstruction. They had intermittent vomiting and they were commonly diagnosed as having gastroesophageal reflux for a long period of time. In only 10 per cent of our patients, the diagnosis was delayed until later than one year of age. A review from the Western country found annular pancreas in 51.5 per cent in children and 48.5 per cent in adults.[1] Eighty five per cent of the children were neonates and male was more common than female. Older children were not found in our study. Several authors reported annular pancreas in adult patients that presented with duodenal obstruction, abdominal pain, gastric ulcer, pancreatitis and pancreatic tumor.[19-22]

Plain x-ray films show either classic double-bubble sign of duodenal atresia or incomplete obstruction of duodenal stenosis. Upper gastrointestinal studies are useful in the cases of incomplete duodenal obstruction. Mast et al[23] described the classic roentgenographic signs of obstructive annular pancreas which include presence of a smooth annular filling defect in the second portion of duodenum, symmetrical lumi-

nal dilatation and reverse intestinal peristalsis proximal to the obstruction. Ultrasonography is used to demonstrate the annular pancreas in neonate but there have been only few cases reported.[24,25]

The operation for correction of duodenal obstruction with annular pancreas should not be done by dissection of the obstructing ring because that would risk complications of pancreatitis, pancreatic fistula and incomplete relief of obstruction.[1,7] A bypass procedure is recommended. Duodenoduodenostomy is preferred to gastrojejunostomy because of better anastomotic function and avoidance of the risk of stomal ulceration.

The mortality rate of duodenal obstruction with annular pancreas in the pediatric patients is 25-30 per cent.[1-3] In our study, the overall mortality rate was 23 per cent and the patients with duodenal atresia had higher mortality rate than the cases of duodenal stenosis ($p<.05$). The high mortality is resulted from associated severe congenital anomalies and serious complications. Prematurity, congenital heart diseases, pulmonary complication and septicemia are the main causes of death. Parenteral nutritional support, highly effective antibiotics and meticulous postoperative care may improve the survival of the seriously ill patients.

Summary

Two hundred and twelve pediatric patients were treated for congenital duodenal obstruction during 1985-1996. Fifty two of them (24.5%) had associated annular pancreas. About 90 per cent of the 52 patients developed bilious vomiting within one week of life. The roentgenographic findings revealed the classical "double-bubble" sign in about 67 per cent. Contrast study gave diagnostic assistance in the patients with incomplete duodenal obstruction.

Available informations from this study suggested that a high incidence of congenital anomalies and prematurity are responsible for the high mortality rate in these patients. In the surgical management of duodenal obstruction with annular pancreas, direct dissection on the obstructing annulus is discourage because of high risks of complication and probably incomplete relief of obstruction. Duodenal bypass by duodeno-duodenostomy was employed with excellent results.

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