Original Article

Congenital Duodenal Obstruction
Sherif N Kaddah, Khaled HK Bahaa-Aldin, Hisham Fayad Aly, Hosam Samir Hassan
Departments of Pediatric Surgery, Cairo University & Tanta University, Egypt

Background/Purpose: Congenital duodenal obstruction is a frequent cause of intestinal obstruction in the newborn. This study aimed to analyze various factors affecting the outcome of these cases at our institution.

Materials & Methods: Seventy one cases of congenital duodenal obstruction were included in this retrospective review. Each case was studied as regard to: age at presentation, gestational age, clinical data, other associated congenital anomalies, cause of obstruction, management, and outcome. Patients with abdominal wall defects (omphalocoele, gastroschisis) and diaphragmatic hernias were excluded from the study.

Results: The causes of duodenal obstruction were: duodenal atresia (n= 37), duodenal diaphragm (n= 12), malrotation (n= 14), and annular pancreas (n= 8). Age ranged from 2 days to 24 months. Bilious vomiting was the main presenting symptom. Plain radiography was the most valuable diagnostic tool in all cases except malrotation and partial obstruction. Gastrointestinal (GIT) contrast study was very valuable in that later group. Overall mortality was 15 cases (21.1 %). The causes of deaths were: prolonged gastric stasis and neonatal sepsis (n= 7), other associated cardiac anomalies (n=5), and extensive bowel gangrene due to neglected volvulus neonatorum (n= 3).

Conclusion: This study showed that (1) the diagnosis of congenital duodenal obstruction is still delayed in many patients referred to our institutions. (2) early postoperative survival is still far from ideal; (3) the mortality is related to delayed presentation, associated cardiac defects, and prolonged gastric stasis; and (4) late complications are more common than previously expected.

Index Word: duodenal obstruction, duodenal atresia, duodenal stenosis, malrotation, volvulus neonatorum, annular pancreas

INTRODUCTION

Although neonatal duodenal obstruction is quite rare, the incidence has been variously estimated as 1 in 10 000 to 1 in 40 000 births. In large series the ratio of atresia : stenosis is 3:2 or 2:2. Duodenal atresia and stenosis is considered frequent cause of intestinal obstruction in the newborn.

Various types of duodenal obstruction exist; they may be either partial or complete, extrinsic or intrinsic or even both. Atresia and stenosis are classed as ‘intrinsic’ obstruction, while annular pancreas and obstructions from errors of midgut rotation produce ‘extrinsic’ obstructions. Annular pancreas may be combined with a complete atresia. Multiple atresias occur in about 15% of all cases. Although all of these forms cause duodenal obstruction there are significant differences between them.
Duodenal atresia can be classified into three major types [Gray and Skandalakis 17]: A type I defect represents a mucosal diaphragmatic membrane (atresia), and is the most common duodenal anomaly observed. The muscle wall in these cases is intact. The portion of the duodenum proximal to the atresia is dilated, and that portion distal to the atretic area is narrowed. The septum may be stretched distally because of peristalsis and high intraluminal pressure ('windsock anomaly').2,9,10 Type 2 duodenal defects have a short fibrous cord connecting the two ends of the atretic duodenum; it is less commonly observed. The two blind ends of the duodenum are separated by a gap, the mesentery showing a V-shaped defect in type 3. Most of the unusual biliary duct anomalies that coexist in these cases are seen in babies with a type 3 defect.8

The first reported congenital obstruction of the duodenum by pyloric membrane was reported by Calder in 17332,10 and by duodenal atresia by Crosby-Leonard in 1856 in London.11 Treatment does not seem to have been attempted until the end of the 19th century.

The only surgical interventions were a gastrostomy and a duodenostomy both of which carried a 100% mortality. The first survivor was not recorded until 1916 by Ernst in Denmark.12

During the past 70 years gradual improvement in the survival of babies with this condition has been achieved.

The aim of this study is to report 71 infants and children with congenital duodenal obstruction treated at 3 pediatric surgical units to find out the different causes and to analyze various factors that have an impact on the outcome of this group of patients.

MATERIALS AND METHODS

Seventy one cases of congenital duodenal obstruction were retrospectively reviewed. These patients were treated by the authors at Departments of Pediatric Surgery at Abuleesh Children’s Hospital, Benha Children’s Hospital, and Tanta University Hospital from June 2000 to June 2005.

The causes of duodenal obstruction were: duodenal atresia (n=37), duodenal diaphragm (n=12), malrotation (n=14), and annular pancreas (n=8) (Fig 1-3). Age ranged from 2 days to 24 months. Patients who had abdominal wall defects (omphalocoele, gastrochisis) and diaphragmatic hernias were excluded from the study.

Charts were reviewed for age at operation, sex, gestational age, other medical problems or anomalies, preoperative clinical data, type and results of imaging studies, operative finding, details of surgical technique, duration of postoperative gastric decompression, time required for resumption of oral intake, hospital stay, and postoperative complications. Follow up period ranged from 5 to 33 months.

RESULTS

The age of patients at time of presentation was variable according to the cause of duodenal obstruction. Patients with complete duodenal obstruction presented during the first few days of life in contrast to those with chronic intermittent or with incomplete obstruction (Table 1)

Preoperative plain radiographs were done in 68 cases, and was sufficient to diagnose duodenal obstruction in all patients except those with malrotation and/or partial obstruction. Upper GIT contrast study was done in 21 cases with incomplete duodenal obstruction.

Duodeno-duodenostomy was the operation of choice in cases of duodenal obstruction, duodenal atresia, duodeal diaphragm and annular pancreas. Division of Ladd bands and proper alignment of intestinal mesentery and ilio-cecal valve was the treatment of choice in malrotation. Three patients with malrotation presented with volvulus neonatorum had gangrenous intestinal loop that requires resection and re-anastomosis

The overall mortality was 15 cases (21.1 %). The causes of deaths were: prolonged gastric stasis and neonatal sepsis (n=7), other associated cardiac anomalies (n=5), and extensive bowel gangrene due to neglected volvulus neonatorum(n=3). The postoperative findings are summarized in Table 2.
Fig 1A. Three-day old baby presented with neonatal intestinal obstruction. Exploration revealed duodenal atresia type I.

Fig 1B. Construction of diamond anastomosis.

Fig 2A. Twelve-month old baby presented with intermittent vomiting and loss of weight. Operative photo showing the site of duodenal obstruction.

Fig 2B. A duodenal diaphragm is bulging from inside.

Fig 3A. Upper GIT series showing duodenal obstruction.

Fig 3B. Abdominal exploration showing annular pancreas.
Table 1. cases of duodenal obstruction

<table>
<thead>
<tr>
<th></th>
<th>DA (n=37)</th>
<th>DD (n=12)</th>
<th>MR(n=14)</th>
<th>AP(n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age at presentation</td>
<td>3.3 days</td>
<td>117.6 days</td>
<td>48.4 days</td>
<td>3.7 days</td>
</tr>
<tr>
<td>No of premature cases</td>
<td>10 (27.2%)</td>
<td>2 (16.7%)</td>
<td>3 (21.4%)</td>
<td>0</td>
</tr>
<tr>
<td>Other anomalies</td>
<td>12 (32.4%)</td>
<td>2 (16.7%)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Acute symptoms</td>
<td>37 (100%)</td>
<td>5 (41.7%)</td>
<td>11 (78.6%)</td>
<td>7 (87.5%)</td>
</tr>
<tr>
<td>Chronic symptoms</td>
<td>0</td>
<td>7 (58.3%)</td>
<td>3 (21.4%)</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>Upper GIT study</td>
<td>2 (5.4%)</td>
<td>10 (83.3%)</td>
<td>6 (42.9%)</td>
<td>3 (37.5%)</td>
</tr>
</tbody>
</table>

DA: Duodenal atresia, DD: Duodenal diaphragm, MR: Malrotation, AP: Annular pancreas

Table 2. Post operative course

<table>
<thead>
<tr>
<th></th>
<th>DA (n=37)</th>
<th>DD (n=12)</th>
<th>MR(n=14)</th>
<th>AP(n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean time to full feeding (days)</td>
<td>7.7</td>
<td>4.7</td>
<td>3</td>
<td>5.6</td>
</tr>
<tr>
<td>The mean Postoperative hospital stay (days)</td>
<td>17.4</td>
<td>7.5</td>
<td>6.3</td>
<td>6</td>
</tr>
</tbody>
</table>

Complications:

<table>
<thead>
<tr>
<th></th>
<th>DA (n=37)</th>
<th>DD (n=12)</th>
<th>MR(n=14)</th>
<th>AP(n=8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wound infection</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Gastric stasis</td>
<td>16</td>
<td>4</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Neonatal septicemia</td>
<td>8</td>
<td>1</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Mortality</td>
<td>10</td>
<td>1</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Anastomotic leak</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

DA: Duodenal atresia, DD: Duodenal diaphragm, MR: Malrotation, AP: Annular pancreas, NG: Nasogastric

DISCUSSION

Approximately 40% of the atresias are found in the duodenum, 35% in the ileum and 25% in the jejunum. Likewise, almost 75% of all intestinal stenosis are supposed to be in the duodenum, 20% in the ileum and 5% in the jejunum. The present series does not represent the total number of patients with congenital duodenal obstruction treated at the three mentioned institutions during the period of the study; it is rather an experience of the contributing authors.

Congenital duodenal obstruction may be associated with other GI and biliary tract abnormalities, or some syndromes as (VACTERL) (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb) association.

Usually the atresias as well as the stenoses are limited to the first and second part of the duodenum. They are relatively uncommon proximal to the ampulla of Vater, the most common site being just at the ampulla. Multiple atresias are rare.

Partial obstruction may be caused by a diaphragm with a central or eccentric opening, or a mucosal web. Both anomalies may cause neonatal obstruction, but they may be diagnosed only later in childhood, depending on the size of the opening. Duodenal diaphragm represented about 17% (12/ 71) of patients in this series. More than half of this subset group of patients (58.3%) presented as incomplete obstruction. The mean age at presentation was 117.6 days

Partial obstruction may also be associated with an extrinsic narrowing of the duodenal lumen by mesenteric bands in association with malrotation, or by an annular pancreas or by aberrant pancreatic tissue in the duodenal wall.
The most common form of extrinsic obstruction consists of pancreatic tissue completely or partially surrounding the second part of the duodenum. All degrees of obstruction are possible, but in addition quite often an atresia of the duodenum at the same level is apparent. Symptomless annular pancreas may be noted by chance during laparotomy or autopsy.

Annular pancreas is a recognized cause of duodenal obstruction in all age groups. Patients most commonly present in infancy or early childhood. Jimenez and his colleagues conducted a thorough review of the English-language literature, and found only 5 series of pediatric patients with annular pancreas since 1950. The total number of patients in these 5 series was 50 patients. Only one of these series included more than 10 patients and only one of these series was published within the last 20 years. Other large series of pediatric duodenal obstruction included patients with annular pancreas but did not analyze them separately.

Whatever the nature of the obstruction the results are very similar: the stomach and proximal duodenum dilate, their walls become hypertrophied, and the distal duodenum is small and remains thin-walled. The stomach and duodenum may perforate in utero or postnatally. Annular pancreas presented 11.3% of total patients in this series (8/71). Associated duodenal atresia was noted in 6 of them.

Bile-stained vomitus in neonates aged 24 hours or younger is the typical presentation of atresia or severe stenosis. Minimal duodenal obstruction in mild stenosis or membrane may have few symptoms. In a few cases, the atresia is proximal to the ampulla of Vater and the vomitus is free of bile. Down's syndrome occurs in approximately 30% of patients, polyhydramnios in 33% to 50%, and 45% are premature. Preoperative plain radiographs were done in 68 cases and was sufficient to diagnose duodenal obstruction in all patients except those with malrotation and/or partial obstruction. Plain radiographs that demonstrate the double-bubble appearance with no distal gas are characteristic of duodenal atresia. The double-bubble sign represents dilatation of the stomach and duodenum. This configuration most commonly occurs with duodenal atresia and an annular pancreas. An annular pancreas is almost always associated with duodenal atresia.

Upper GIT contrast study was done in 21 cases in series with incomplete duodenal obstruction. The majority of these cases had incomplete or intermittent duodenal obstruction. We believe that no oral contrast materials are necessary in the evaluation of complete duodenal obstruction. However, a small amount of contrast material can be instilled through a feeding tube into the distal stomach and duodenum to differentiate between the diaphragm and a long stenosis.

Occasionally, barium enema examination is suggested as an adjunct study in the evaluation of duodenal atresia. To demonstrate a malpositioned cecum but this is not always diagnostic of malrotation and volvulus.

In our series diamond shaped duodeno-duodenostomy was the standard treatment for patients with intrinsic and annular pancreas. We preferred this technique even in duodenal diaphragm cases as it prevents any injury to duodenal ampulla or pancreatic duct, further more there were no reports of re-stenosis compared to excision of the diaphragm. In cases of annular pancreas duodenoplasty is preferred by most authors to prevent re-stenosis, and pancreatic fistulas, on attempt of annulus excision.

Weber and his colleagues compared different surgical techniques in treatment of duodenal atresia and suggested better results of diamond shaped duodeno-duodenostomy than other techniques. Effective treatment of annular pancreas was accomplished only in the last half of the 20th century. In 1944, Gross and Chisholm reported first successful operation of duodenal by-pass instead of releasing the annulus around the pancreas in a 3 days old girl with annular pancreas Morbidity and mortality using bypass technique was clearly understood in later reports explaining that annular pancreas may not only cause extrinsic obstruction, but may lead, or be associated with intrinsic narrowing as well.

The overall mortality rate in is very high in our series (n=15, 21.1%) compared to other reports. Esscher and his colleagues reported 3% early mortality. Our highest mortality rate was in cases of duodenal atresia (n=10, 27%). This very high mortality is attributed to prolonged stay in NICU, gastro-duodenal dysfunction from mega duodenum and gastric dilatation leading to neonatal sepsis. In our series gastric stasis manifested by mega...
duodenum with blind loop syndrome, gastro-esophageal reflux, and delayed transit that occurred in 22 (31%) patients of intrinsic duodenal obstruction, 9 of them were complicated by neonatal sepsis. Compared to our series, Spigland and Yazbeck, reported gastric stasis symptoms in 77% of their patients of congenital intrinsic duodenal obstruction, with mortality 6%. Weber and his colleagues reported no mortalities in their series. Other causes of early mortality were the presence of multiple congenital anomalies. Two of our patients with duodenal atresia had associated congenital heart and TEF, the other patient was premature (<30 weeks). In the current series, Mean hospital stay in cases of duodenal atresia was 17.4 days and the Mean time till onset of feeding was 7.7 days. These figures were higher compared to Weber et al who reported 16.2, 4.1 days respectively, and Alexander et al who reported 9 and 6 respectively. Mean total hospital stay in cases of malrotation was 7 and 3 days, mean time till onset of feeding was 3 days, compared to John et al which was 7 and 3 respectively, in our series there were 3 cases of volvulus neonatorum. The incidence of post-operative wound infection in our series was (7.6%).

CONCLUSION
The morbidity and mortality rate of patients with congenital duodenal obstruction in this series are still far from ideal. Effective measures are urgently needed to improve the outcome. Early diagnosis and intervention, proper preoperative preparation, nutritional support are among these measures.

REFERENCES