Congenital Duodenal Stenosis: Early and Late Presentation

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Abstract

Background/Purpose: Congenital duodenal obstruction is a frequent cause of intestinal obstruction in the newborn. The incomplete nature of the obstruction in duodenal stenosis results in a variable and often delayed presentation. It usually results in recurrent episodes of vomiting, aspiration, or failure to thrive. Some patients present in adulthood with gastroesophageal reflux, peptic ulceration, or obstruction of the duodenum proximal to the stenosis by a bezoar.

The Aim of this Study: Is to report children with congenital duodenal stenosis treated at our pediatric surgery unit, to find out the factors affecting the delayed presentation and its effect upon the prognosis and to describe the management and outcome of those patients.

Material and Methods: Over a 5 years period, 13 patients with congenital duodenal stenosis were treated at pediatric surgery unit, Assiut university children hospital. There were 6 males and 7 females. 4 patients presented the 1st month after birth, 9 cases presented the following months.

Results: 13 patients were operated upon. 5 patients had associated anomalies, 2 patients had malrotation, one patient had Down’s syndrome, patent foramen ovale and PDA. One patient had inguinal hernia and one patient had polydactyl. We used duodenoplasty, joining the bowel just proximal and distal to the obstruction with excision of the duodenal membrane. The mean operative time was 55 minutes. Most patients passed stool within 24 hours and started oral feeding within 2-3 days and discharged within one week.

Conclusion:
• Duodenal stenosis can present in neonates, infants, children and even adults.
• A neonate with repeated bilious vomiting should be investigated as a duodenal obstruction till proved otherwise.
• Early presenting cases of duodenal stenosis resembles duodenal atresia.
• Late presentation occurs due to incomplete nature of the obstruction, negligence of parents or unexpectancy of the diagnosis.
• In patients with malrotation, duodenal stenosis, as an association, should be excluded.

Key Words: Duodenal stenosis — Duodenal diaphragm — Duodenal atresia.

Introduction

DUODENAL atresia and stenosis are rare causes of intestinal obstruction in the newborn; the prevalence of intrinsic duodenal obstruction (atresia, web, or severe stenosis) is 1:6,000 [1].

Failure of recanalization of the duodenal lumen during the eighth to tenth week of gestation, results in duodenal atresia. Incomplete recanalization can lead to duodenal stenosis or the presence of a duodenal web [2].

Duodenal stenosis is relatively rare in comparison with duodenal atresia [2]. Obstruction may be caused by a diaphragm with a central or eccentric opening, or a mucosal web. Both anomalies may cause neonatal obstruction, but Duodenal stenosis may be diagnosed only later in childhood depending on the size of the opening [3,4].

Apart from the routine open surgical options for the treatment of duodenal stenosis, newer options have become available in recent years like laparoscopic duodeno-duodenostomy for duodenal stenosis in neonates and image-guided balloon dilatation for membranous duodenal stenosis in children [5,6].

Fiberoptic diagnosis together with endoscopic membranectomy using a high-frequency wave cutter could become an alternative option to other more standard diagnostic and therapeutic procedures in managing duodenal intrinsic stenosis [7].
The aim of this study is to report children with congenital duodenal stenosis treated at our pediatric surgery unit, to find out the factors affecting the delayed presentation and its effect upon the prognosis and to describe the management and outcome of those patients.

**Patients and Methods**

Over a 5 years period, 13 patients with congenital duodenal stenosis were treated at pediatric surgery unit, Assiut university children hospital From Jan. 2012 — June 2013. There was 6 males and 7 females. The patients were selected from patients presented by neonatal bilious vomiting but without the typical picture of duodenal atresia or who was diagnosed as duodenal atresia with operative finding of duodenal stenosis and patients presented beyond the neonatal period were considered primarily as duodenal stenosis.

4 patients presented the 1st month after birth, 2 of them referred from our gynaecological hospital and 2 cases referred from other centers.

9 cases presented the following months referred from other centers or come primarily to our unit without referral.

**Table (1): Age distribution.**

<table>
<thead>
<tr>
<th>Day of presentation</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Up to 1 month</td>
<td>4</td>
</tr>
<tr>
<td>1 month - 1 year</td>
<td>4</td>
</tr>
<tr>
<td>1-2 years</td>
<td>3</td>
</tr>
<tr>
<td>More than 2 year</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>13</strong></td>
</tr>
</tbody>
</table>

The main presenting symptom of all patients was repeated attacks of bilious vomiting since birth in 10 patients, and started after that in 3 patients who were tolerating only milk and fluids till the age of 3 months. All patients passed meconium within 48 hours after birth, with chronic constipation in most cases. Early presented cases had a fair general condition. Cases presented later had a bad general condition with anaemia, hypoproteinaemia, hypokalaemia, hyponatremia, marasmus and low weight for age.

**Table (2): Associated medical complications.**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaemia</td>
<td>9 cases</td>
</tr>
<tr>
<td>Hypoproteinaemia</td>
<td>5 cases</td>
</tr>
<tr>
<td>Hypokalaemia</td>
<td>8 cases</td>
</tr>
<tr>
<td>Hyponatremia</td>
<td>8 cases</td>
</tr>
<tr>
<td>Marasmus</td>
<td>5 cases</td>
</tr>
<tr>
<td>Monilial infection</td>
<td>4 cases</td>
</tr>
<tr>
<td>Chest infection</td>
<td>3 cases</td>
</tr>
</tbody>
</table>

All patients were subjected to full physical examination, CBC, kidney and liver functions, serum proteins, sodium and potassium. Abdominal sonography, plain radiograph of the abdomen and an upper gastrointestinal contrast study. Gastroduodenoscopy was done for 3 patients older than 2 years. It was inconclusive in 2 patients and diagnostic in the patient aged 9 years.

Diagnosis was done on clinical suspicion and through investigations by plain X-ray abdomen and an upper gastrointestinal contrast study. No cases were diagnosed prenataally.

All patients were prepared for surgery after correction of anaemia hypoproteinaemia and electrolyte imbalance and stabilization of the general condition usually within a few days. Prophylactic antibiotics was given as a routine.

**Operative technique:**

All patients subjected to open surgical treatment through a right transverse supraumbilical incision, not reaching the midline. The intestine explored to search for other GIT anomalies, malrotation, etc. The level of duodenal obstruction identified (proximal dilatation and distal collapse). To be sure that the obstruction is incomplete, milking of the contents in the proximal dilated stomach and duodenum or normal saline, given from the nasogastric catheter pass to the distal end but with great difficulty.

A 2cm longitudinal incision was made at the antimesenteric border centered at the constriction ring. The obstructing diaphragm was identified as a double layer mucous diaphragm opposite to the constriction ring with a variably located and sized tiny opening. After localizing the site of ampula of Vater, the mucosal diaphragm was excised circumferentially. The resection line is oversewn with continuous suture vicryl 5-0 and the enterotomy was closed transversely. A tube drain inserted and
the abdomen closed in layers. The patient kept on IV fluids for 2-3 days and usually passed stool after 24 hours, broad spectrum combination antibiotics and metronidazol were given for 7 days and the patient discharged within one week.

Results

This study included 13 patients with duodenal stenosis. 9 patients diagnosed preoperatively to have duodenal stenosis, 2 patient diagnosed as malrotation and 2 patients diagnosed to have duodenal atresia and intraoperatively proved to be duodenal stenosis.

Diagnosis was done on clinical suspicion, plain radiograph of the abdomen showed the classic double bubble with small bowel gas is observed distal to the double bubble in most cases. An upper gastrointestinal contrast study pre-operatively showed a dilated stomach and duodenum with arrest of the dye in the 2nd or 3rd part of the duodenum. Abdominal sonography revealed only dilated stomach and duodenum.

13 patients were operated upon. 6 males and 7 females. 5 patients had associated anomalies, 2 patients had malrotation, one patient had Down's syndrome, patent foramen ovale and PDA. One patient had inguinal hernia and one patient had polydactyl. There was no associated anomalies that could contraindicate surgery or anaesthesia. The operation was completed successfully in all patients.

Table (4): Associated anomalies.

<table>
<thead>
<tr>
<th>Associated anomaly</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malrotation</td>
<td>2 cases</td>
</tr>
<tr>
<td>Down's syndrome</td>
<td>One case</td>
</tr>
<tr>
<td>Patent foramen ovale</td>
<td>One case</td>
</tr>
<tr>
<td>PDA</td>
<td>One case</td>
</tr>
<tr>
<td>Polydactyl</td>
<td>One case</td>
</tr>
<tr>
<td>Inguinal hernia</td>
<td>One case</td>
</tr>
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</table>

At laparotomy the duodenum was dilated and the wall thickness was increased, with a level of obstruction in the duodenum, in the 2nd part in 4 cases and in the 3rd part in 9 cases.

2 neonate patients were diagnosed as duodenal atresia, at laparotomy duodenal stenosis was diagnosed and the operation completed as the other cases of duodenal stenosis.

The 2 patients with malrotation, one was operated upon for malrotation with persistent post operative symptoms, re-exploration revealed duodenal stenosis in the 3rd part of the duodenum, and in the other patient who was diagnosed preoperatively as malrotation, we noted that the site of stenosis was the same level of obstructing Ladd's band in the 3rd part of the duodenum and differs from usual level of malrotation obstruction in that in duodenal stenosis is thickened on palpation with visible constiction on the duodenal wall. Ladd's procedure and duodenoplasty was performed.

There was no intraoperative complications except a minor bleeding occurred from the duodenal wall in one patient controlled by compression.

The mean operative time was 55 minutes. Most patients passed stool within 24 hours and started oral feeding within 2-3 days and discharged within one week.

1st wound dressing after 3 days and every 3 days, the drain removed after 7 days and the wound left exposed. Nice wound healing occurred in all patients. Minor complications have occurred in 3 patients from those presented after the 1st month and included One patient had superficial wound infection with minimal purulent discharge whom responded well to conservative treatment.

There was no deaths and all patients are doing well long term follow-up from 2-4 years.
Congenital Duodenal Stenosis

Fig. (1D): Gastrographin meal of the same patient, lateral view.

Fig. (1E): Postoperative plain X-ray of the same patient.

Fig. (2A): 3rd part duodenal stenosis.

Fig. (2B): 3rd part duodenal stenosis, constriction ring.

Fig. (2C): Duodenal diaphragm.

Fig. (2D): Excised diaphragm, malrotation (Caecum on the left side).
Fig. (2E): Excised diaphragm.

Fig. (2F): Excised diaphragm.

Fig. (2G): Duodenoplasty.

Fig. (2H): Duodenoplasty, with malrotation (Caecum on the left side).

Fig. (3A): 2nd part duodenal stenosis.

Fig. (3B): 2nd part duodenal diaphragm.
Fig. (3C): 2nd part duodenal diaphragm.

Fig. (3D): Excised 2nd part duodenal diaphragm.

Fig. (3E): Excised 2nd part duodenal diaphragm.

Fig. (4A): Gastrografin of a 9 years child showing dilated stomach, first and second parts of the duodenum.

Fig. (4B): Endoscopic view of the same patient showing narrowing of the duodenal lumen.

Fig. (4C): Operative view of the same patient showing dilated stomach, first and second part of stomach with collapsed third part of duodenum.
Discussion

Congenital duodenal obstruction may be due to annular pancreas, atresia or duodenal diaphragm and the condition may be associated with Down's syndrome and cardiac defects [8,9].

The incomplete nature of the obstruction in duodenal stenosis results in a variable and often delayed presentation [10].

In this situation symptoms first occur when advancing the infant from formula to solid food, or it may be unmasked much later in infancy, childhood or, in rare instances, adulthood, when a progressive decrease in motility or impaction of food or a foreign body causes more pronounced symptomatology [11,12].

The delayed presentation of these anomalies in the adult is difficult to explain, but the presence of a dilated stomach and a proximal duodenal bulb with a patulous pylorus suggests a progressive loss of compensatory peristaltic action to overcome a small duodenal aperture or narrowing of the descending duodenum. Most studies of these lesions are single case reports or small series, which do not allow a single surgeon to accumulate extensive experience; therefore, reliance on the combined experience of others in recognition and appropriate management has been the norm [9,13,14]. It usually results in recurrent episodes of vomiting, aspiration, or failure to thrive. Some patients present in adulthood with gastroesophageal reflux, peptic ulceration, or obstruction of the duodenum proximal to the atresia by a bezoar [10]. Chandler reported one case aged 19 years. Dorken reported one patient aged 78 years with congenital duodenal stenosis of the second part of the duodenum with symptoms dating since childhood. Ward reported one patient aged 55 years with congenital duodenal stenosis of the first part of the duodenum [15-17]. We had only one case aged 9 years presented by repeated attacks of bilious vomiting and failure to thrive.

In our series, early presented cases resembles duodenal stenosis to a great extent, 2 of them diagnosed as duodenal atresia. Late presentation occurred due to the incomplete nature of the obstruction, negligence of parents or unexpectency of the diagnosis.

Inspite duodenal atresia is diagnosed antenatally in up to 50% of cases, with polyhydramnios, dilatation of the stomach and proximal duodenum visible on the third trimester ultrasound [18], none of our cases were diagnosed prenatally. The radiographic sign of duodenal atresia is the "double bubble" with gaseous distension of the stomach and proximal duodenum and total absence of intestinal gas distally [18]. If small bowel gas is observed distal to the double bubble, the differential diagnosis includes duodenal stenosis, duodenal web and intestinal malrotation with midgut volvulus [19]. We think that plain radiograph of the abdomen is a reliable method for diagnosis of duodenal stenosis as it usually shows gas distal to the classic "double bubble" of duodenal atresia as proved in our series. Inspite the lack of more distal intestinal gas is diagnostic of a complete obstruction [20], whereas the presence of gas indicates a partial obstruction, we had 2 neoates diagnosed to have complete duodenal obstruction proved intraoperatively to be duodenal stenosis. This could be explained by the fact that the opening in the duodenal diaphragm was very narrow or the plain radiograph were taken before passage of gases distal to the stenosis.

In recent years, infants have been evaluated by fiberoptic flexible upper gastrointestinal endoscopy. This allows the direct observation of anomalies in the duodenum such as duodenal stenosis, atresia and membrance or extrinsic compression [18]. In our series we did not depend on endoscopy as a diagnostic tool as the diagnosis was obvious in most of our cases. It was done for 3 patients older than 2 years. It was inconclusive in 2 patients and diagnostic in the patient aged 9 years.

Associated congenital anomalies have been reported in more than 50% of patients with duodenal obstruction and can include congenital heart disease, pancreatic anomalies, intestinal malrotation, esophageal atresia, variants of imperforate anus, renal anomalies or other combinations of anomalies such as VATER/VACTERALS. The association between Down syndrome and gastrointestinal abnormalities is well known. Down syndrome is recognized as a possible predisposing condition for gut abnormalities and congenital heart disease as part of the VATER syndrome [21]. We had one case with Down's syndrome presented by duodenal stenosis, PDA and patent foramen ovale, although presented clinically few days after birth, diagnosed at the age of 13 months. The association between duodenal stenosis and malrotation to our knowledge have not been reported previously. We reported 2 cases.

Duodenal stenosis could be treated surgical or endoscopic. The choice of surgical procedure is largely based on the preference of the surgeon. Duodenoduodenostomy, duodenotomy with incision or excision of the diaphragm, duodenojjun-
ostomy, or duodenoplasty can all be considered as
different modes of operative management [21]. We
use duodenoplasty, joining the bowel just proximal
and distal to the obstruction with excision of the
duodenal membrane closed transversely in Heineke-
Mikulicz fashion as we think that it is the best
operative procedure as it is the most direct, phys-
iological repair and, of the available options, has the
least potential for later complications.

In recent years, apart from the above-mentioned
open procedures, laparoscopic duodeno-duoden-
ostomy and image-guided balloon dilatation have
been tried. Laparoscopic duodeno-duodenostomy
can be done in neonates with no intraoperative and
postoperative complications [22,23].

Patients undergoing laparoscopic repair of CDO
had a shorter length of hospitalization and more
rapid advancement to full feeding compared to
babies undergoing the open approach [24].

Image-guided balloon dilatation has also been
tried in children with membranous duodenal steno-
sis [23].

Duodenal stenosis differs from duodenal atresia
in that the patient usually passes stool but with
constipation, during operation the proximal part
is dilated and the distal part is narrow but usually
full and not collapsed and squeeze of the proximal
part passes its contents, with difficulty, into the
distal part and differs from malrotation in that the
obstruction about to be complete and the bilious
vomiting is almost always continous. We tried to
find the cause of delayed presentation in most
cases and we noticed that unexpecation of the
condition especially by pediatricians or by =spe-
ialized surgeons and negligence or ignorance of
the parents are probably the main cause as most
patients were passing stool.

Gray and Skandalakis have classified congenital
duodenal obstructions into 3 types-I, II, and III ill.
The type I or membranous duodenal obstruction
(mucosal diaphragm, or web) has been quoted to
account for 0.8-92% of all cases [27].

We think that duodenal stenosis is an incomplete
type 1 duodenal atresia.

Although prognosis of intestinal atresia in gen-
eral is good, an overall mortality of 7% for duodenal
obstruction is shown in large series. Associated
congenital anomalies are identified as an independ-
ent risk factor for an impaired clinical course.
Low birth weight and the problems of prematurity
further increase mortality risk [28].

In the absence of other serious anomalies or
prematurity, the overall survival for duodenal
stenosis or atresia is nearly 100% [29].

We have no deaths in our series and the overall
prognosis is good with low morbidity.

Conclusion and Recommendations:
- Duodenal stenosis can present in neonares, infants,
children and even adults.
- A neonate with repeated bilious vomiting should
be investigated as a duodenal obstruction till
proved otherwise.
- Early presenting cases of duodenal stenosis re-
sembles duodenal atresia.
- Late presentation occurs due to incomplete nature
of the obstruction, negligence of parents or =ex-
pectancy of the diagnosis.
- In patients with malrotation, duodenal stenosis,
as an association, should be excluded.
- Duodenal stenosis could be treated surgically by
duodenoplasty either open or laparoscopic or
endoscopic by dilatation or diaphragm excision.
- Duodenoplasty for duodenal stenosis is easier
than duodenoduodenostomy for duodenal atresia.

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