Congenital duodenal web: successful management with endoscopic dilatation

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Background and study aims: Congenital duodenal web (CDW) is an uncommon cause of duodenal obstruction and endoscopic balloon dilatation has been reported in just eight pediatric cases to date. Here we are reporting three cases of CDW managed successfully with balloon dilatation.

Cases and methods: In 2014 we diagnosed three cases of CDW on the basis of typical radiological and endoscopic findings. Endoscopic balloon dilatation was done under conscious sedation with a through-the-scope controlled radial expansion (CRE) balloon.

Results: All three children presented late (median age 8 [range 2–9] years) with bilious vomiting, upper abdominal distension, and failure to thrive. One of them had associated Down syndrome and another had horseshoe kidney. In all cases, CDW was observed in the second part of the duodenum beyond the ampulla, causing partial duodenal obstruction. After repeated endoscopic dilatation (2–4 sessions), all three patients became asymptomatic. None of the patients experienced complications after balloon dilatation.

Conclusions: Duodenal diaphragm should be suspected in patients with abdominal distension with bilious vomiting, even in relatively older children. Endoscopic balloon dilatation is a simple and effective method of treating this condition.
ery 3 weeks until a patient became asymptomatic. Patients were observed for complications for 6 hours post-dilatation. The initial dilatation session was performed on an inpatient basis and subsequent dilatations were done as outpatient procedures.

Case Reports

All three children presented late (median age 8 years [range, 2–9]) with bilious vomiting, upper abdominal distension, and failure to thrive. One of them had associated Down syndrome and another had a horseshoe kidney. The clinical details about the patients are listed in Table 1. The outcome of their endoscopic dilatation procedures is listed in Table 2. The details of individual cases are as follows.

Patient 1

A 9-year-old boy presented with vomiting, abdominal distension, and failure to thrive that began at age of 3 months. He had a history of large-volume, projectile, bilious vomiting that occurred three to four times a week. On examination, he was emaciated and had phenotypic features of Down syndrome. His abdomen was grossly distended with visible peristalsis and borborygmi. A bilateral ultrasound showed a 10-mm CRE balloon under fluoroscopic guidance (Fig. 1 c). Following the dilatation, the endoscope could be negotiated easily beyond the narrowing. The distal duodenum was normal and narrowing was appreciated as a membrane (suggestive of duodenal web). Two more sessions of dilatation were done (Table 2). At the same time, the child was started on a proton pump inhibitor for reflux esophagitis and peptic stricture. On follow-up, the child’s vomiting and distension had subsided, he began gaining weight and his esophageal ulcer healed.

Patient 2

An 8-year-old girl presented with complaints of vomiting and failure to thrive since birth. She was having bilious vomiting two to three times per week with increased frequency of vomiting over the last 3 years. She also complained of pain and upper abdominal distension, which used to decrease after vomiting. On examination she had significant failure to thrive (Table 1) and her abdominal examination revealed epigastric distension. Investigations revealed normal CBC, serum creatinine, and thyroid profile. Abdominal ultrasonography showed a horseshoe kidney and no evidence of annular pancreas. Her abdominal x-ray (erect) showed a double bubble sign and barium meal follow-through (Fig. 2a) revealed narrowing in the second part of the duodenum with dilated proximal duodenum and stomach. Abdominal ultrasonography did not show any evidence of annular pancreas. UGI endoscopy showed a passage narrowing at the lower end of the esophagus with circumferential ulcers (LA-D GERD) and hiatal hernia. There was a narrowing in the second part of the duodenum just distal to the ampulla, with a tiny hole at the center with proximal duodenal dilatation (Fig. 1b). Endoscopic dilatation of duodenal web was done with a 10-mm CRE balloon under fluoroscopic guidance (Fig. 1c). Following the dilatation, the endoscope could be negotiated easily beyond the narrowing. The distal duodenum was normal and narrowing was appreciated as a membrane (suggestive of duodenal web). Two more sessions of dilatation were done (Table 2). At the same time, the child was started on a proton pump inhibitor for reflux esophagitis and peptic stricture. On follow-up, the child’s vomiting and distension had subsided, he began gaining weight and his esophageal ulcer healed.

Patient 3

A 2-year-old female child presented with vomiting and failure to thrive. She was having bilious vomiting and distending abdomen which used to decrease after vomiting. On examination she had symptoms of upper abdominal distension with visible peristalsis and borborygmi. An abdominal (erect) x-ray showed a double bubble sign and barium meal follow-through (Fig. 1a) revealed narrowing in the second part of the duodenum with dilated proximal duodenum and stomach. Abdominal ultrasonography did not show any evidence of annular pancreas. UGI endoscopy showed a passage narrowing at the lower end of the esophagus with circumferential ulcers (LA-D GERD) and hiatal hernia. There was a narrowing in the second part of the duodenum just distal to the ampulla, with a tiny hole at the center with proximal duodenal dilatation (Fig. 1b). Endoscopic dilatation of duodenal web was done with a 10-mm CRE balloon under fluoroscopic guidance (Fig. 1c). Following the dilatation, the endoscope could be negotiated easily beyond the narrowing. The distal duodenum was normal and narrowing was appreciated as a membrane (suggestive of duodenal web). Two more sessions of dilatation were done (Table 2). At the same time, the child was started on a proton pump inhibitor for reflux esophagitis and peptic stricture. On follow-up, the child’s vomiting and distension had subsided, she began gaining weight.

Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)/sex</th>
<th>Age at onset of symptoms</th>
<th>Location of membrane</th>
<th>Symptoms</th>
<th>Weight and height z-score</th>
<th>Associated condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9/M</td>
<td>3 months</td>
<td>Second part duodenum distal to ampulla of Vater</td>
<td>– Stale food and bilious vomiting – Upper abdominal distension with visible peristalsis and borborygmi – Failure to thrive</td>
<td>–4.25 and –6.73</td>
<td>Down syndrome, hiatal hernia, GERD, peptic esophageal stricture</td>
</tr>
<tr>
<td>2</td>
<td>8/F</td>
<td>Birth</td>
<td>Second part duodenum distal to ampulla of Vater</td>
<td>– Bilious-vomiting – Upper abdominal distension, borborygmi and pain abdomen – Failure-to-thrive</td>
<td>–3.98 and –2.72</td>
<td>Horseshoe kidney</td>
</tr>
<tr>
<td>3</td>
<td>2 yr, 3 mo/f</td>
<td>6 months</td>
<td>Second part duodenum distal to ampulla of Vater</td>
<td>– Upper abdominal distension (postprandial with visible peristalsis) – Bilious-vomiting – Constipation – Failure-to-thrive</td>
<td>–2.25 and –3.88</td>
<td>–</td>
</tr>
</tbody>
</table>

Table 2

<table>
<thead>
<tr>
<th>Size of balloon</th>
<th>No. of sessions</th>
<th>Interval between sessions</th>
<th>Complications</th>
<th>Follow up after last session</th>
<th>Clinical status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>12, 15 and 18 mm</td>
<td>3</td>
<td>3 weekly</td>
<td>Nil</td>
<td>7 months</td>
</tr>
<tr>
<td>Case 2</td>
<td>10 and 12 mm</td>
<td>2</td>
<td>3 weeks</td>
<td>Nil</td>
<td>9 months</td>
</tr>
<tr>
<td>Case 3</td>
<td>8, 12, 15, and 18 mm</td>
<td>4</td>
<td>3 weekly</td>
<td>Nil</td>
<td>6 months</td>
</tr>
</tbody>
</table>

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A 2-year-old girl presented with a history of vomiting, failure to thrive, and upper abdominal distension for 6 months. She was having bilious vomiting once or twice a week. On examination the patient had wasting, stunting (● Table 1), and her abdominal examination revealed epigastric distension. Investigations revealed normal CBC and thyroid profile. Barium meal follow-through (● Fig. 3 a) showed narrowing in the second part of the duodenum with a dilated proximal duodenum. Abdominal ultrasonography did not show any evidence of annular pancreas. UGI endoscopy showed a membrane-like obstruction with a tiny opening in the second part just distal to the papilla (the membrane was bellowing into the proximal dilated duodenum [wind-sock] due to reverse peristalsis, a characteristic picture of duodenal diaphragm) (● Fig. 3 b). Endoscopic dilatation of duodenal web was started with an 8-mm CRE balloon (● Fig. 3 c).

Three more dilatation were performed at 3-week intervals (● Table 2), using progressively larger balloons, after which the child’s vomiting subsided, her abdominal distension decreased, she started gaining weight.

Discussion

Congenital duodenal web is a rare condition that usually presents early in life. Patients with complete-type duodenal web present early in the neonatal period with duodenal obstruction whereas the fenestrated type may present late. Reports also exist of congenital duodenal web diagnosed in adulthood [5, 9]. In our case series, all three patients presented late as they had the fenestrated type of web with recurrent bilious vomiting, abdominal distension, and failure to thrive. All three children had a web in the second part of the duodenum (distal to the ampulla), which is the most common site of CDW formation [4]. Multiple associated disorders like Down syndrome, cardiac anomalies, malrotation of the gut, vertebral defect, renal anomalies etc. have been described with CDW [10]. In our series one child had associated Down syndrome and another one had a horseshoe kidney.

Surgery, in the form of excision and duodenoplasty or bypass procedures like duodeno-duodenostomy and duodeno-jejunostomy used to be the conventional modality for treatment of CDW [2, 3]. However, with the advent of endoscopic procedures for various gastrointestinal conditions, treatment with procedures such as endoscopic membranotomy with laser [11], sphincterotomy [12], high-frequency-wave snare/cutter [12], hot biopsy forceps [4], insulated-tip diathermic knife [13] and needle knife [14] have been reported. Endoscopic therapy has become popular because it produces no abdominal incision (scar), is not asso-
associated with complications such as adhesion development, involves shorter hospital stay, and the procedures sometimes can be performed without general anaesthesia. More invasive endoscopic procedures such as cutting the web with an electrocautery device or laser carry risks of perforation, excessive bleeding and trauma to the ampulla of Vater. Endoscopic dilatation, in contrast, is a relatively safe procedure with a reduced risk of the previously mentioned complications. Endoscopic balloon dilatation produces tearing in the membrane of the diaphragm, which contains only mucosa and submucosa and lacks a muscular layer. The first endoscopic balloon dilatation of CDW, done with a 12-mm balloon catheter was reported by Asabe et al [6] in 2008 in a 9-day-old boy. The dilatation was repeated at 30 and 51 days of life with 13-mm and 13.5-mm balloon catheters. Mochizuki et al [7] reported another case of 7-month-old girl and, like us, they used a through-the-scope CRE balloons measuring 10 mm, 12 mm, and 15 mm for proximal jejunal web. Recently, Huang et al [8] reported their experience with endoscopic balloon dilatation in a series of six cases involving children aged 2 to 37 months with CDW. They used 10-mm to 16-mm balloon catheters and with the exception of one child, all were treated only with balloon dilatation. One child required endoscopic electrocauterization of an obstructing membrane following balloon dilatation. In our series, we managed to treat all three patients with endoscopic balloon dilatation only. All of our patients became asymptomatic after two to four sessions of endoscopic dilatation and none developed any complications.

In conclusion, duodenal diaphragm should be suspected in cases of abdominal distension with bilious vomiting in children, even those who are relatively older. Endoscopy usually confirms the diagnosis. Endoscopic balloon dilatation is a simple and effective method of treating this condition.

**Competing interests:** None

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**References**