ORIGINAL ARTICLE

Delayed Presentation of Perforated Duodenal Diaphragm in Infants and Children: Our Institutional Experience in a Tertiary Care Hospital

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Abstract:
Background: This was our institutional experience about infant and children of perforated duodenal diaphragm presenting with recurrent vomiting, delayed due to late visit to the paediatric surgeon. *Aim and Objective:* The aim of this study was to observe the clinical presentation, diagnostic challenges, reasons for delay in the diagnosis and management of infant and children with perforated duodenal diaphragm. *Material and Methods:* This is a clinical observational study conducted from September 2018 to December 2020. There were eight patients (2 male and 6 female) in our study. *Results:* Out of eight patients, five patients presented with on and off non-bilious vomiting, six (75%) had < 3rd percentile of weight for age at the time of admission. Associated anomalies were malrotation (50%), ventricular septal defect (25%), and Downs's syndrome in one patient (12.5%). *Conclusion:* Delayed presentation of perforated duodenal diaphragm is a rare surgical condition. Early diagnosis is a challenge for the treating physician. Detailed history of recurrent vomiting, proper clinical examination, relevant investigation and referral to the paediatric surgeon with high index of suspicion can aid in early diagnosis.

Keywords: Double bubble sign, Duodenal obstruction, Duodenal web, Malrotation, Perforated duodenal diaphragm, Recurrent vomiting.

Introduction:

Congenital duodenal obstruction (atresia or diaphragm) is one of the common surgical causes of neonatal intestinal obstruction with a reported incidence of 1 in 5000 to 10000 live births. Atresia and complete duodenal diaphragm can be easily diagnosed by establishing bilious vomiting with the classical "double bubble" sign in X-ray abdomen generated by the proximal left-sided air- and fluid-filled stomach tapering at the pylorus and the distal dilated proximal duodenum to the right of the midline in the neonatal period [1]. The perforated or incomplete duodenal diaphragm masks the features of obstruction and leads to delay in diagnosis due to passage of milk through it. This will present with symptoms when the passage (hole) gets blocked. This can occur intermittently so diagnosis becomes delayed. The diagnosis can be missed both preoperatively and intraoperative, in the patients with perforated diaphragm without any external duodenal wall deformity or if there is wind sock into distal duodenum[2].

This study was aimed to observe the clinical presentation, diagnostic challenges, reasons for delay in the diagnosis and management of infant and children with perforated duodenal diaphragm.

Material and Methods:

This was a prospective clinical observational study comprising of eight consecutive cases of intrinsic duodenal obstruction due to perforated duodenal diaphragm. Patients for the study were selected from the department of paediatric surgery at our institution during September 2018 to December 2020.

Patients who presented with recurrent vomiting either bilious or non-bilious for a long duration with age beyond the neonatal period (1month) were included in the study. All eight patients referred to us; were treated by paediatric physicians either in the peripheral centres or paediatric hospitals for recurrent vomiting. All admitted patients had X-ray and base line blood investigations done. The patients were re-evaluated clinically with detailed history and physical

examination, and radiologically with plain X-ray of abdomen, ultrasonography abdomen and upper gastrointestinal (UGI) series by administering gastrografin (1:3 dilution) through a nasogastric tube in situ and details were recorded. The patients were investigated for complete blood count, renal functions, liver functions and viral markers. The UGI endoscopy or CT abdomen was reserved for patients with diagnostic dilemma. Karyotyping was performed on patients who could afford this procedure to rule out Down's syndrome and findings were noted. Exploratory laparotomy was performed on patients after correction of dehydration, electrolyte imbalance, renal function and anaemia. Pre anaesthetic check-up was done in all patients before laparotomy. The findings were noted and surgically dealt with accordingly. Post operatively the patients were observed for complications. After discharge, routine follow up was done and parameters like weight gain, milestone development etc were recorded. We included all patients beyond the age of neonatal period with duodenal obstruction due to perforated duodenal diaphragm. Patients with perforated duodenal web presented within neonatal period (one month) and patients with duodenal obstruction due to extrinsic lesions like annular pancreas, isolated malrotation ± volvulus, pre duodenal portal vein and duodenal duplication or neoplasm were excluded from the study.

Results:

All the patients of perforated duodenal diaphragm which were admitted at our institution had a history of taking medications for on and off vomiting. They were diagnosed with gastroenteritis but finally diagnosed as perforated duodenal diaphragm. There was no previous history of loose motions or fever. Six patients had a history of hospitalizations for vomiting or dehydration at paediatric hospitals. All the patients had record/prescription of anti-emetic medication. After clinical suspicion of gastrointestinal surgical cause, patients were admitted, optimised and investigated at our institute. The mean age at presentation was 7 months. The clinical and demographic details along with weight for age are depicted in Table 1[3]. Five patients had normal appearing plain X-ray of the abdomen (n=8). X-ray of the abdomen of two patients had double bubbles with gas in distal bowel [Figure 1 (A)]. X-ray of the abdomen of one patient showed atypical large gas shadow in left upper abdomen with gas in distal bowel on supine, single large air fluid level on erect and multiple air fluid like shadow on lateral supine film (figure 2). Three patients had features of malrotation without volvulus in their ultrasonography of the abdomen and five patients had dilated stomach and duodenum. The UGI gastrografin contrast study was performed in seven patients; five revealed typical finding of dilated stomach and duodenal bulb with delayed clearance of contrast distally [Figure 1(B and C)]. The remaining two had an atypical inconclusive pattern. One patient underwent UGI endoscopy, which revealed dilated stomach and duodenum along with duodenal diaphragm with wind soak deformity. There was a small foreign body (FB) over duodenal diaphragm and an eccentric small hole through which scope was not negotiable at the second part of duodenum (D2) [Figure 3 (B)]. Computed tomography was performed in two patients with diagnostic dilemma, revealed malrotation, but was unable to rule out or confirm duodenal diaphragm. Preoperative diagnosis of perforated duodenal diaphragm was possible in six patients, with diagnostic dilemma in remaining two with malrotation. For three affordable patients, karyotype was done; one had clinical as well as 21st trisomy suggestive of Down's syndrome. On 2D echocardiography, two patients had small ventricular septal defect. Exploratory laparotomy was performed in all the patients. Out of eight patients, 4 (50%) had pre-ampullary and 4(50%) had post-ampullary perforated duodenal diaphragm. All patients were looked for distal multiple webs by inserting 10F foley's catheter in duodenum distal to web up to duodeno-jejunal junction and the inflating balloon with 3-5ml normal saline and slowly retrieving back. All the patients were regularly followed up after discharge; every month for first three months and then quarterly, with the longest being two years. All patients were gaining weight appropriate for age and none has developed major complications till the date of writing this manuscript.

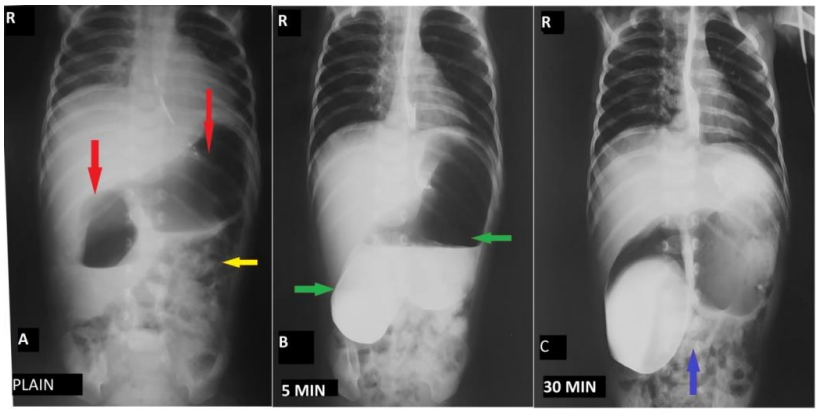

Figure 1: Classical findings of perforated duodenal diaphragm. A: Plain X ray of abdomen, double bubble (red arrow) with gas in distal bowel (yellow arrow); B: UGI contrast study showing dilated stomach and duodenal bulb (green arrow); C: Slow transit of contrast into distal bowel beyond perforated duodenal diaphragm at 30 minutes (blue arrow)

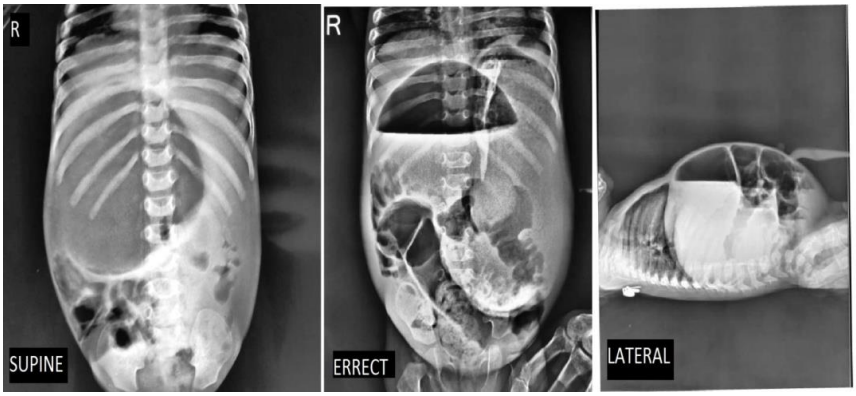

Figure 2: An atypical X ray of abdomen findings in spine, erect and supine lateral films showing single large bubble in left upper abdomen, large air-fluid level and multiple air fluid like shadows with gas in distal bowel respectively in a patient with incomplete duodenal diaphragm

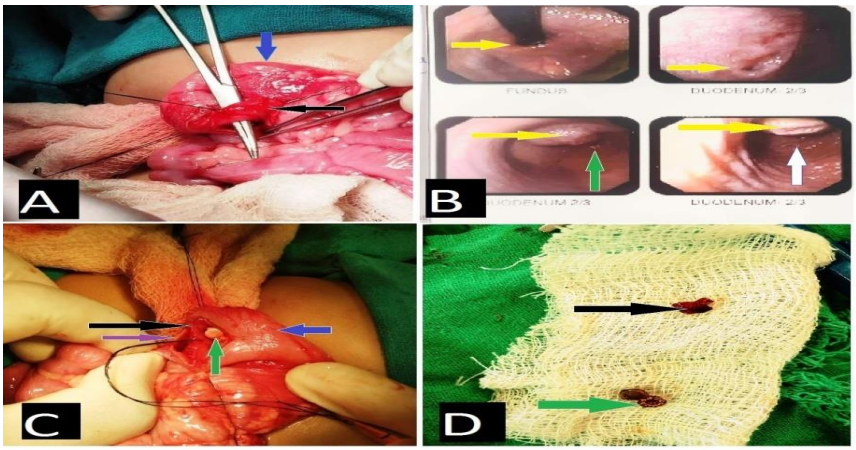

Figure 3: Intraoperative and endoscopic findings of incomplete duodenal diaphragm. A, C and D: dilated duodenum (blue arrow), duodenal diaphragm with hole (black arrow), distal narrow duodenum (purple arrow) and foreign body (green arrow); B: small eccentric hole in duodenal diaphragm (yellow arrow), wind soak (white arrow) and foreign body (green arrow)

Discussion:

Table No. 1: Clinical and demographic details

Case no.	Age/sex	BW	Presentation	WOP	WFA*	Associated morbidities
1	2 months / F	2.2 kg	On and off bilious vomiting for last 3weeks, FTT, severe dehydration	2.2 kg	< 3rd percentile	Anaemia
2	8 months / M	2.5 kg	Recurrent abdominal distension with on and off non-bilious vomiting for last 4months, FTT, dehydration	8.5 kg	50th percentile	Anaemia, malrotation
3	2 months / F	2.5kg	Respiratory distress with on and off non-bilious vomiting since birth, FTT, severe dehydration	1.7 kg	< 3rd percentile	Anaemia, Aspiration pneumonitis, electrolyte imbalance, VSD
4	8 months / F	2.7 kg	On and Off bilious vomiting for last 2 months, FTT, dehydration	5.5kg	< 3rd percentile	Malrotation, Meckels diverticulum
5	4 months / F	2.8 kg	On and Off bilious vomiting since birth, FTT, dehydration	3.2 kg	< 3rd percentile	Down's syndrome, Malrotation, VSD
6	8 months / F	2.5 kg	On and Off non-bilious vomiting for last 1month	7.5 kg	Between 15 to 50th percentile	Ladds bands, Impacted foreign body
7	1Y 9 months / M	2.7 kg	On and Off non-bilious vomiting for last 10 months, FTT, dehydration	5.5 kg	< 3rd percentile	Anaemia, Atypical malrotation, Impacted food particles
8	3 months / F	3.1 kg	On and Off non-bilious vomiting since birth, FTT, severe dehydration	1.8 kg	< 3rd percentile	Electrolyte imbalance

M: male, F: female, BW: birth weight, WOP: weight on presentation, WFA*: weight for age [3], FTT: failure to thrive, VSD: ventricular septal defect.

Table No. 2: Intraoperative findings, surgical procedure performed, post-operative complications and follow-up data

Case no.	Location of diaphragm	Type of diaphragm	Surgery	Post-operative Complications	Length of hospital stay	Follow-up after surgery	Weight in kg (WOP+gain)
1	Post ampullary, D2-D3	Diaphragm with a central hole	EW and HMP	Melena on day 3	14 days	2 years	2.2 +12
2	Post ampullary, D2-D3	Diaphragm with a central hole	EW and HMP +LP	None	15 days	2 years	8.5+8
3	Pre ampullary, D2	Diaphragm with a central hole	EW and HMP	Sepsis	37 days	15 months	1.7+9.5
4	Post ampullary, D3	Diaphragm with a central hole	EW and HMP +LP	None	15 days	13 months	5.5+7
5	Post ampullary, D2-D3	Diaphragm with a central hole	EW and HMP +LP	None	15 days	12 months	3.2+5
6	Pre ampullary, D2	Diaphragm with an eccentric hole	EW and HMP +ladds band release +FB retrieval	None	14 days	12months	7.5+6
7	Pre ampullary, D2	Diaphragm with a central hole	EW and HMP + LP	Sepsis, melena, NG tube bleeding	26 days	6 months	5.5+5
8	Pre ampullary, D2	Diaphragm with a central hole	Side to side Duodeno-Jejunostomy	None	11 days	2 months	1.8+1.4

D: duodenum, EW and HMP: excision of web and Heineke-Mikulicz duodenoplasty, LP: ladds procedure, FB: foreign body, NG tube: nasogastric tube and WOP: weight on presentation

Table No. 3: Post-operative course

Case no	TPN	Glycerine enema	Passage of meconium	NG tube removal	Start of feed	Discharge
1	D1	N	D5	D10	D11	D13
2	D2	N	D3	D5	D7	D10
3	D0	D3	D6	D10	D11	D14
4	D1	N	D3	D7	D8	D10
5	D1	N	D4	D8	D10	D12
6	D1	N	D3	D5	D7	D10
7	D1	N	D2	D12	D11	D17
8	D1	D2	D2	D6	D7	D9

TPN: total parental nutrition, D: day, N: not given, NG tube: nasogastric tube

Although congenital intrinsic duodenal obstruction is one of the common surgical causes of neonatal intestinal obstruction, duodenal diaphragm is relatively uncommon with incidence of 1 in 20000 to 40000 live births [4]. Embryonic insult causing failure of recanalization of duodenal lumen after temporary obliteration by epithelial proliferation during 4^{th} to 11^{th} week of gestation is thought to result in an intrinsic web, atresia or stenosis [1]. More than 50% of patients with duodenal atresia are associated with other congenital anomalies commonly down's syndrome, congenital heart disease, annular pancreas and malrotation [1,2,4].

In our series, 5 patients (62.5%) had associated congenital anomalies like malrotation, congenital cardiac anomaly (VSD), Down's syndrome, and Meckel's diverticulum. Malrotation was present in 4 patients (50%, n=8) in our series. There are few reported cases of delayed presentation of perforated duodenal web associated with malrotation. A case of duodenal web in the fourth part associated with malrotation was reported in a 14-month-old male child by author Singh AP et al. (2020) [5]. Similar sign and symptoms were present in our cases. Among these four patients one patient had VSD with Down's syndrome and one patient had broad base Meckel's diverticulum (Table 1).

The most common presenting symptom is bilious vomiting in early infancy, but pre-ampullary duodenal obstruction presents with non-bilious vomiting. In case of duodenal diaphragm, timing of presentation depends on whether it is complete or incomplete. Complete duodenal diaphragm usually presents in neonatal period, but presentation is late in incomplete duodenal diaphragm; timing and nature of clinical symptoms depend on the size and location of the hole. The delayed in diagnosis has been reported even up to adulthood [6,7]. The mean age of diagnosis in our series was 7 months. The Mousavi et al. and Ratani et al, reported the mean age of diagnosis as 26.7 months and 9.5 months respectively [4,8].

Some cases of perforated duodenal web present sudden bilious vomiting and upper abdomen distension. These patients become symptomatic due to blockage of duodenal web aperture by large food particle or foreign body ingestion. Young L et al. reported a case of 16-month-old female child that presented with recurrent vomiting and perforated duodenal web. Duodenal web was unmasked by ingestion of a foreign body, which caused acute complete duodenal obstruction [9]. In our series one 8-month-old female child was admitted with similar presentation. The foreign body was impacted in perforated duodenal web which was removed during laparotomy.

The most common reason which lead to diagnostic delay in the present series was on and off vomiting; moreover 50% had non-bilious vomiting with in between normal feeding and were treated at paediatric hospitals. The clinical presentation in the perforated duodenal diaphragm can be confusing due to presence of other associated congenital anomalies; they may present with nonspecific abdominal symptoms like abdominal discomfort, worsened after feed and relieved by an episode of vomiting [10].

The prenatal diagnosis may be possible by ultrasonography in as many as 50% of cases of congenital duodenal obstruction; more common in the presence of other congenital anomalies, and is therefore associated with higher overall mortality rates [1]. During postnatal period, complete obstruction can be easily diagnosed with plain abdominal X-ray by typical double bubble appearance one on each side of the vertebral column. But in case of incomplete duodenal diaphragm UGI contrast study is the gold standard for diagnosis. Most of the perforated duodenal diaphragm was present in second part of duodenum (85-90%). For older infants UGI endoscopy may help to diagnose and locate duodenal diaphragm. A stenotic lumen can also identify radiolucent foreign body/impacted food particles [11].

The presentation with unwell, emaciated, pale looking child with recurrent on and off vomiting, dehydration, failure to thrive and mild upper abdominal fullness with no visible peristalsis should warrant thorough evaluation to rule out the perforated duodenal web. The commonly practiced open surgical procedures for delayed presenting incomplete duodenal diaphragm are anterolateral duodenotomy with excision of web and transverse duodenoplasty i.e. Heineke-Mikulicz duodenoplasty or duodenoduodenostomy and rarely duodenojejunostomy [12].

These procedures now a days are increasingly being performed by either laparoscopically or by surgical robots, but require skilled expertise [13,14]. Recently, in many case reports endoscopic surgery has been reported to be safe and effective with balloon catheter dilatation (Hercules 3-Stage Esophageal Dilation Balloon) of stenotic lumen of incomplete diaphragm or incision/resection with an insulated-tip knife [11,15,16,17,18] Newer approaches need resources, skilful and trained experts. Limitations of this study are small sample size and resources and trained expertise for minimal access or endoscopic surgery.

Conclusion:

Although incomplete duodenal diaphragm is a rare surgical condition, its diagnosis is very difficult for primarily treating physician as it can present with mild abdominal discomfort to complete obstructive clinical picture. Detailed history of recurrent vomiting, proper clinical examination, relevant investigation and referral to the paediatric surgeon with high index of suspicion can aid in early diagnosis.

Conflict of Interest - Nil

Sources of Support - Nil

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