Original Article

Delayed Presentation of Duodenal Atresia

Charu Tiwari, Nitinkumar Bhajandas Borkar, Sunita Singh, Sonali Mane, Chandrasen Sinha[1]

Department of Paediatric Surgery AIIMS, Raipur, Chhattisgarh, India, [1]Department of Paediatric Surgery, St. George's University Hospital, London, England

Abstract

Background: The most common type of duodenal atresia (DA) (Type I), also known as duodenal web or membrane can present later in infancy or early childhood if the membrane or web is fenestrated. We describe six patients with delayed presentation of DA. **Materials and Methods:** Retrospective review of hospital records of six patients with delayed presentation of DA due to fenestrated web managed in Paediatric Surgery Department at a tertiary care institute over a period of 2 years (January 2019 to December 2020) was done. The data of these patients were analysed on the basis of age at presentation, clinical presentation, associated anomalies, radiological findings, intra-operative findings, management and postoperative course. **Results:** The median age at presentation was 6.5 months (range: 1 month to 10 years). There were four males and two females. The most common presentation was emesis seen in all six patients. Two patients had Down syndrome. Associated congenital anomalies were cardiac in one patient, anterior ectopic anus in one patient and malrotation of midgut in one patient. Upper gastrointestinal contrast suggested incomplete duodenal obstruction in all patients. At laparotomy, fenestrated duodenal membrane was observed in all patients – preampullary in three patients and postampullary in three patients. Lateral duodenotomy, web excision and transverse closure was done in all six patients. The postoperative period was uneventful in all patients and mean duration of hospital stay was 9 days. **Conclusion:** Fenestrated duodenal webs present a diagnostic challenge to the paediatric surgeons because of delayed and variable clinical presentation. A modification of the present classification of DA has been proposed in this study which would help in better reporting of epidemiology and aid in early diagnosis of this congenital anomaly.

Keywords: Artesia, congenital, delayed presentation, duodenal

Introduction

Congenital duodenal obstruction is amongst the common causes of neonatal intestinal obstruction; the cause of which may be intrinsic or extrinsic.[1] Its incidence is approximately one in 2500–10,000 live births and it accounts for up to 50% of neonatal intestinal obstruction.[2-4] Usually, intrinsic causes like duodenal atresia (DA) cause complete obstruction have early clinical presentation; whereas extrinsic causes like Ladd's band and annular pancreas may have variable presentation due to incomplete duodenal obstruction.[1]

The present classification of congenital (intrinsic) DA as proposed by Gray and Skandalakis which is being followed divides it into three types; the most common being Type I, also known as duodenal web or membrane.[5,6] Some such patients with web can also present later in infancy or early childhood if the membrane or web is fenestrated. There are various reports of duodenal membrane or web with fenestration in literature where the child presented later with failure to thrive and malnutrition.[6-8] Even paediatric surgery textbook has also mentioned about this variety; however, it has never been incorporated into the classification.[5]

With this background, we describe six patients with delayed presentation of DA caused by fenestrated duodenal web and present an extensive review of literature highlighting the difficulties in diagnosing such cases. We have also described a brief history of attempts at classification of DA done previously by various authors before the current classification by Gray and Skandalakis was accepted. This is followed by our suggestion of incorporating the fenestrated duodenal web into the present classification.

Received: 17-04-2021 **Revised:** 08-04-2022 **Accepted:** 09-06-2022 **Available Online:** 04-01-2023

Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.ajps_66_21

Address for correspondence: Dr. Nitinkumar Bhajandas Borkar, Department of Paediatric Surgery, AIIMS, Raipur - 492 099, Chhattisgarh, India.
E-mail: drnitinborkar25@gmail.com

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How to cite this article: Tiwari C, Borkar NB, Singh S, Mane S, Sinha C. Delayed presentation of duodenal atresia. Afr J Paediatr Surg 2023;20:97-101.

Materials and Methods

The hospital records of six patients with delayed presentation of DA due to fenestrated web managed in Paediatric Surgery Department at a tertiary care institute for 2 years (January 2019 to December 2020) were retrospectively reviewed. Descriptive analysis was performed on the basis of age at presentation, clinical presentation, associated anomalies, radiological findings, intra-operative findings, management and post-operative course. The data were assessed and those with a normal distribution were reported as mean with standard deviation and those with a skewed distribution were reported as median with range.

Results

Six patients with delayed presentation of DA (because of incomplete intrinsic obstruction caused by fenestrated duodenal membrane) were managed for 2 years from January 2019 to December 2020. The median age at presentation was 6.5 months (range: 1 month to 10 years). There were four males and two females. The most common presentation was emesis seen in all six patients. It was bilious in three patients and nonbilious in three other patients. Other presenting symptoms were failure to thrive (n = 4), upper abdominal distension (n = 2) and acute upper gastrointestinal obstruction (n = 1). Two patients had Down syndrome. One patient had cardiac anomalies, one patient had anterior ectopic anus and malrotation of the midgut was identified intraoperatively in one patient.

Upper gastrointestinal contrast series were done in all patients and suggested incomplete duodenal obstruction [Figure 1]. One patient underwent contrast-enhanced computed tomography of abdomen which suggested dilated stomach and proximal duodenum with the passage of contrast in the small bowel with few filling defects [Figure 2]. At laparotomy, fenestrated duodenal membrane was observed in all patients – preampullary in three patients and postampullary in three patients [Figures 3 and 4]. One patient had associated malrotation of midgut for which Ladd's Procedure was done. Lateral duodenotomy, web excision and transverse closure were done in all six patients. One patient who was a known case of Down's syndrome (diagnosis confirmed previously by karyotyping) presented at 10 years of age with acute obstruction caused by impaction of berry seed at the fenestrated duodenal membrane [Figure 5]. The post-operative period was uneventful in all patients and the mean duration of hospital stay was 9 days. Table 1 summarises the demography, clinical presentation and management of all these six patients.

Discussion

A neonate with congenital DA usually presents with vomiting soon after birth which may be bilious or nonbilious depending upon the site of atresia with respect to the opening of the ampulla. Usually, the common site of DA is between the first and second parts of duodenum (85%).[5] There is minimal or no epigastric fullness. The erect abdominal X-ray shows the classic double bubble sign. The diagnosis of such neonates is easy and management is surgical. Recently, with improvement in antenatal care and regular use of antenatal scans, there has been an increase in the incidence of antenatal diagnosis of DA. Duodenal obstruction is being detected as double bubble sign with polyhydramnios on foetal ultrasonography.[9]

Embryologically, intrinsic obstruction of the duodenum is speculated to be related to a failure of recanalisation of the duodenal lumen (Tandler's Theory).[5] The duodenum is derived from the distal proportion of the foregut and has a nearly closed lumen during the embryonic period. Between weeks 8 and 10 of gestation, vacuolisation begins ultimately, leading to a patent lumen.[10] Failure of this process leads to various anomalies that have been classified by Gray and Skandalakis as stated in the introduction section.[5,6]

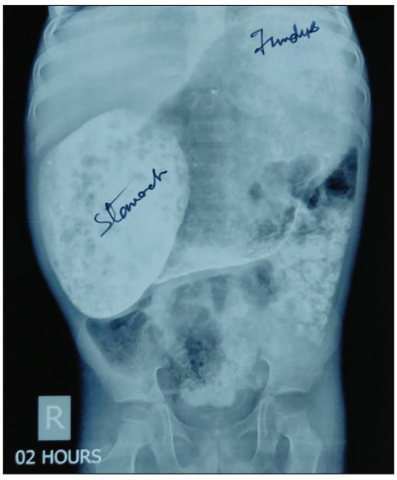


Figure 1: Upper gastrointestinal contrast series (2-h Image) showing dilated stomach and proximal duodenum with passage of contrast in distal small bowel

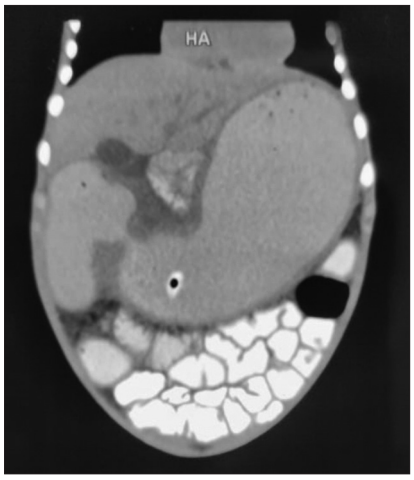


Figure 2: CT scan of the Downs syndrome patient showing dilated stomach and proximal duodenum with passage of contrast in distal small bowel and filling defect. CT: Computed tomography

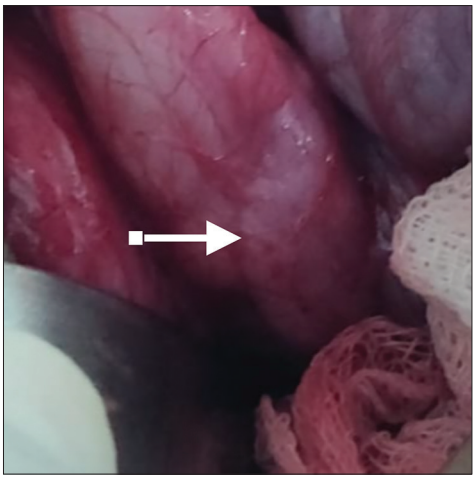


Figure 3: Intra-operative image showing the site of duodenal obstruction (white arrow)

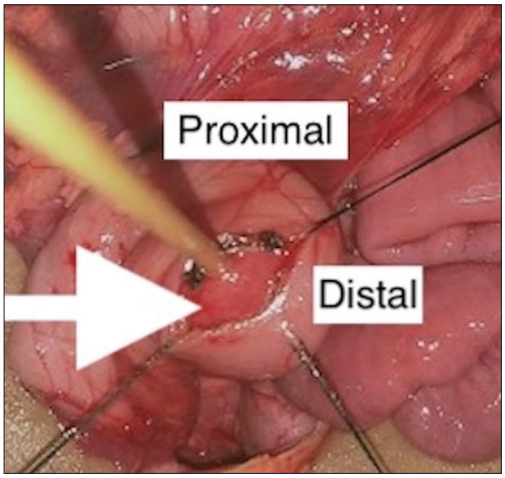


Figure 4: Intra-operative image showing the Foleys catheter in the fenestrated duodenal web (white arrow)

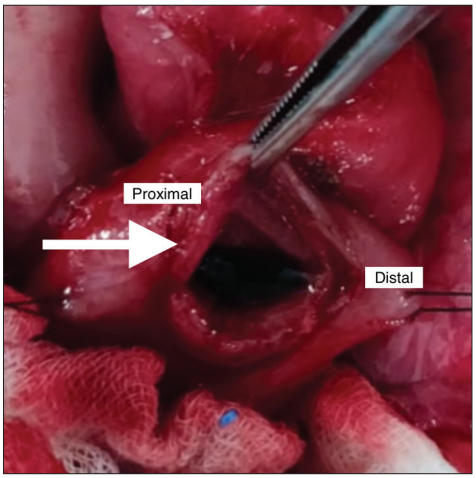


Figure 5: Intra-operative image showing the obstruction of fenestrated duodenal web (white arrow) by Berry seeds

However, in a subset of patients with congenital DA, there is a fenestrated duodenal web, which causes partial obstruction.

Table 1: Details of the six patients with fenestrated web having delayed clinical presentation

Age at presentation	Sex	Clinical presentation	Associated anomaly
3 months	Female	Bilious emesis, FTT	Low ARM (anterior ectopic anus)
10 years	Male	Chronic on acute obstruction with abdominal pain and distention	Down's syndrome
35 days	Male	Nonbilious emesis, FTT	Nil
2.5 years	Male	Nonbilious emesis, malnutrition, abdominal distention	Nil
10 months	Female	Mixed emesis, FTT	K/C/O downs syndrome with cardiac anomaly (ASD)
35 days	Male	Bilious emesis, FTT	Malrotation of gut

FTT: Failure to Thrive, ARM: Ano-Rectal Malformation, ASD: Atrial Septal Defect

The clinical presentation in such cases is variable and delayed. Usually, the child does not present during the neonatal period as the obstruction is partial and does not hamper the passage of milk. As the consistency of feeds changes after weaning there can be obstruction of the fenestrated web by food particles. It dilates the duodenal segment proximal to the site of the web. It can lead to atony and ineffective peristalsis.[6] In the present series, one patient presented with acute symptoms due to obstruction of the fenestrated web by berry seeds.

Common clinical presentations of patients with fenestrated duodenal web are vomiting, failure to thrive, malnutrition, with or without upper abdominal distention. Erect abdominal X-ray usually shows dilated stomach with normal gas shadow beyond the site of obstruction. Upper gastro-intestinal contrast series usually shows proximal dilatation of duodenum and distal normal calibre small bowel.

The incidence of associated anomalies in patients with DA varies from 38% to 55%; Trisomy 21, intestinal malrotation, situs inversus and congenital cardiac diseases being the most common.[5] Prematurity and low-birthweight may also be associated.[5] Two patients in our series had Down's syndrome and one had associated anterior ectopic anus and one had malrotation of the midgut. One had no associated anomaly.

The available literature on delayed presentation of the fenestrated duodenal web is limited to a few studies and individual case reports which were reviewed.[6,8,11-13] There are studies describing variable incidences of fenestrated duodenal webs ranging from 23.07% to 75% amongst the total duodenal webs reported by them in the respective studies.[6,8,9,14-17] Table 2 briefly summarises various studies reporting delayed presentation of the fenestrated web.[6,8,9,14-17]

Fenestrated duodenal webs present a diagnostic challenge because of variable clinical presentations, as observed in our study – age at presentation can be delayed up to 10 years and

Table 2: Review of literature of various studies with duodenal atresia, duodenal webs and delayed presentation

Study by authors	Year published	Total number of patients with duodenal atresia (*n*)	Total number of patients with fenestrated duodenal webs (*n*)	Total number of patients with delayed presentation (*n*)
Sarin *et al*.[6]	2012	18	3	3
Krieg *et al*.[8]	1937	18	11	8
S. Kumar *et al*.[9]	2015	13	3	3
Nawaz *et al*.[14]	2004	8	6	2
Kaddah *et al*.[15]	2006	12	7	7
Lin *et al*.[16]	2012	13	7	7
Mousavi *et al*.[17]	2016	19	8	7

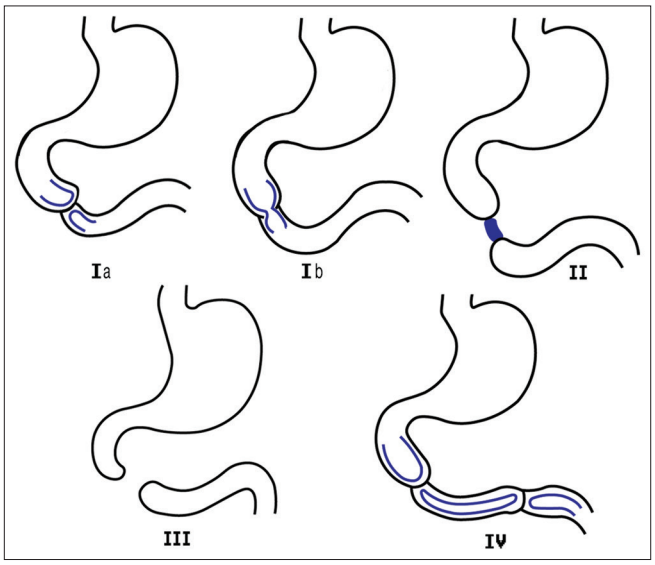


Figure 6: The proposed modification of congenital duodenal atresia

clinically may present with vomiting, abdominal distention and failure to thrive. However, the other types of DA (a complete web or Type II and Type III atresia) presents at the neonatal period itself because of complete obstruction. This adds to the challenge as such children are less often thought to have a congenital lesion. Hence, we emphasise that this type of DA, i.e., the fenestrated web should be incorporated in the present classification of DA so that this congenital lesion is kept as one of the differential diagnosis in children with similar presentation.

The attempts at classifying congenital DA date back to the early 20th century. Buchanan in 1929 classified DA as intrinsic and extrinsic.[18] Krieg in 1937 suggested unified classification system for better management of such patients.[8] In his article, he suggested Forssner's classification to be used in all cases to aid investigations. Forssner's classified duodenal malformations into three types – Diaphragm (complete and incomplete), Stenosis (complete and incomplete) and Atresia (blind ends connected by a band and blind ends unconnected).[8,19] In 1941, Kelly published a case report of the successful management of DA and proposed classifying intrinsic DA into three types– A (diaphragm across the lumen), B (blind ends connected by cord) and C (blind ends with no connection).[20] However, the present and widely used classification of DA was established by Gray and Skandalakis in 1972 which did not include incomplete/fenestrated web and multiple atresia.[5]

Table 3: The proposed new classification of congenital duodenal atresia

Congenital duodenal atresia classification	
Type of Duodenal Atresia	**Description**
Type I	Duodenal web/membrane
	Type Ia: Complete duodenal web/membrane
	Type Ib: Fenestrated duodenal web/membrane
Type II	Two blind ends of duodenum with intact mesentery and separated by a fibrous cord
Type III	Two blind ends of duodenum with a V-shaped defect in the mesentery
Type IV	Multiple duodenal atresias/webs

Multiple DA are rare but have been reported in many individual case reports.[9,21-24] We thus, try to propose a new classification of DA which would include both fenestrated web and multiple atresia, as summarised in Table 3 and Figure 6. The fenestrated web has been classified as Type I b and multiple DA has been included as Type IV in our proposed new classification. The new proposed classification would help in better reporting of epidemiology and aid in early diagnosis of such cases. It would also help to avoid misdiagnosis or missed diagnosis.

Conclusion

Fenestrated duodenal webs present a diagnostic challenge to the paediatric surgeons because of delayed and variable clinical presentation; high index of suspicion is required keeping this congenital anomaly as a cause of partial duodenal obstruction while managing such patients. A modification of the present classification of DA has been proposed in this study which would help in better reporting of epidemiology and aid in early diagnosis of this congenital anomaly.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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