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Paediatrics

CONGENITAL FENESTRATED DIAPHRAGMS OF GUT.

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ABSTRACT

Background: Congenital gastrointestinal (GI) diaphragm are uncommon causes of intestinal obstruction. We review our experience and discuss the mode of presentation, diagnostic difficulties, associated anomalies and intraoperative surgical maneuvers to diagnose and treat these rare anomalies.

Methods and Methods: This is a retrospective review of 21 patients with congenital GI diaphragms at a tertiary care teaching hospital. The demography and the clinical spectrum was analyzed.

Results: There were 13 boys and 8 girls, presenting from birth to 4 years. The major symptom was vomiting 17(81%), 6(28%) children had failure to thrive, 2 children had visible peristalsis, 1 abdominal distension, 1 constipation, and 1 absent anus. Ultrasonography and Radiological Investigations was done in all cases. All children underwent surgery. There were 11 duodenal, 5 gastric diaphragms, 1 jejunal, 2 ileal and 2 rectal congenital diaphragms. Mean duration of surgery was 105 min (60-180 min), commencement of feeding 4 (3-7days) and average hospital stay was 9 days(7-21days)There is no mortality in this series.

Conclusion: The clinical presentation of gastrointestinal diaphragms is variable depending on the site and the size fenestration of the diaphragm. They may be diagnosed antenatally on ultrasonography or may present with recurrent vomiting and/or failure to thrive. We propose to investigate all cases of unexplained recurrent vomiting by contrast study and institute appropriate treatment.

KEYWORDS : Duodenal diaphragm, Pyloric diaphragm, Ileal diaphragm, Jejunal diaphragm, Rectal diaphragm.

Introduction

Congenital gastrointestinal diaphragms are rare causes of chronic intestinal obstruction in children. They may be associated with other congenital anomalies like, Mongolism, Malrotation of Gut, Annular Pancreas and pre duodenal portal vein. They are a type of intestinal atresia and may occur in any part of GI tract however it is most commonly found in stomach and duodenum. The reported incidence of gastric outlet obstruction, excluding infantile hypertrophic pyloric stenosis, was one in 100,000 live births [1]. In a reported series of 131 intestinal atresias there were 14 cases of jejunoileal diaphragms [2]. However the true incidence of this entity is unknown.

The symptoms are often vague such as abdominal pain, recurrent non-bilious vomiting, and failure to thrive after a variable period of normal growth and food intake[3].

We have evaluated the clinical spectrum and the surgical procedures of the patients diagnosed with the congenital GI diaphragms.

Materials and Methods

We reviewed the case files of all the children who were diagnosed with congenital gastrointestinal (GI) diaphragms from 2002-12. The demographic data, duration of symptoms, clinical presentation, associated anomalies, antenatal events, investigations, surgical procedures and outcomes were collected and analyzed.

Demography and Clinical Presentation

Twenty one patients were diagnosed and treated for congenital GI fenestrated diaphragms. There were 13 boys (62%) and 8 girls [Table 1]. The major symptom was vomiting 17 (81%), it was bilious in nature in 12 cases and non-bilious in 5 cases, 6 (28%) children had failure to thrive and visible peristalsis was seen in 2 (9%) cases.

Table 1: Age at presentation

Age group	Male	Female
1-30 days	3	3
1-12 months	7	4
1-5 years	3	1
Total	13	8

Eleven (53%) children had duodenal diaphragm, 5 (24%) had pyloric diaphragm, 1 (5%) jejunal, 2 (9%) ileal and 2 (9%) had rectal diaphragms. Two patients had maternal polyhydraminos, and one of the baby's antenatal scan suggested duodenal diaphragm. There was one (5%) diaphragm in the first part of the duodenum, 8 (38%) in the second part and 2(10%) in the third part. All the gastric diaphragms were present in the prepyloric region, a jejunal diaphragm was present about 30 centimeters from the duodenojejunal flexure and 2 diaphragms were seen on the ileal region about 30 cm from the ileo-cecal junction. There were two rectal diaphragms, one associated with anorectal malformation and the other presented with intractable constipation. [Chart 1]

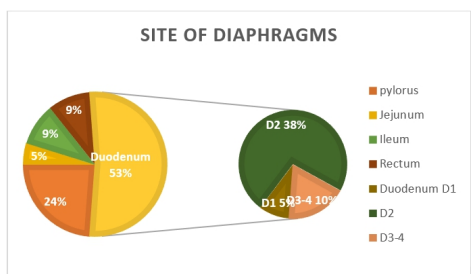


Chart 1: Distribution of the diaphragms in the GI Tract.

The most frequent associated congenital anomaly was Gastrointestinal (43%) followed by Cardiac (19%), Renal (9%), Central Nervous system 1 (5 %) and Heterotaxy 1(5%). Two (9%) children were diagnosed to have Downs Syndrome. Malrotation of the gut was present in 7 (63%) of the duodenal diaphragms and one case of gastric diaphragm, annular pancreas in two cases and one child had anorectal malformation. Atrial septal defect was the commonest cardiac anomaly, one case had horse shoe kidney and one child had grade 3 vesicoureteric reflux. One baby had congenital aqueductal stenosis. One child with duodenal diaphragm had heterotaxy with complete siatus inversus and pre duodenal portal vein. [Chart 2]

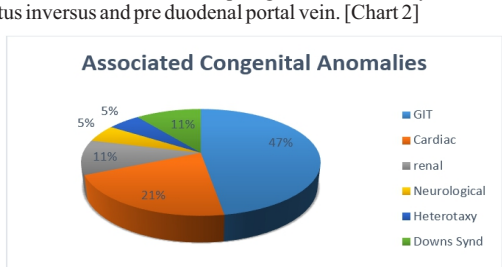


Chart 2: Pie diagram depicting the associated systemic anomalies.

Investigations

Plain abdominal radiographs and ultrasonography were obtained in all cases. Accurate pre-operative diagnosis was made only in 9 cases and all cases were further subjected to Barium contrast study with delayed films were taken to document intraluminal obstruction. All the children with diaphragms in the duodenum showed an enlarged stomach, a distended duodenal cap and varying amount of gas and fluid in the rest of the abdomen. Five children had gastric dilatation and were diagnosed to have pyloric antral diaphragms, one month old boy had 3 fluid levels apart from a distended stomach, had jejunal diaphragm on laparotomy.

One child was antenatally diagnosed as duodenal atresia, postnatal abdominal radiograph and contrast study suggested intra-luminal fenestrated duodenal diaphragm. A fenestrated duodenal diaphragm was excised from the second part of the duodenum.

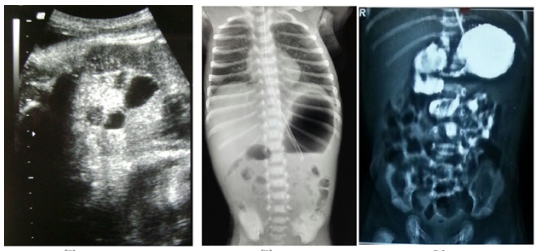


Figure 1-3: Antenatal double bubble on ultrasonography with post natal radio radiography showing duodenal cap and contrast in distal bowel.

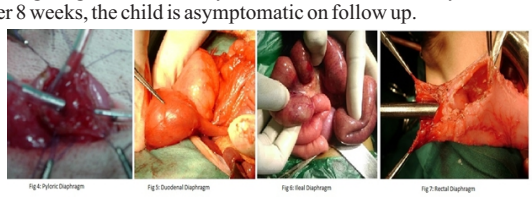
Surgery and Outcome

All children underwent surgery. The children electrolytes, acid-base imbalance was optimized prior to surgery. Nasogastric decompression with gastric lavage was done in select cases. Mean duration of surgery was 105 min (60-180 min), none of the children required blood transfusion. Commencement of feeding was 4 days (3-7days) and average hospital stay was 9 days (7-21days). There is no mortality in this series. Malrotation was present in 7 cases of duodenal diaphragms and in one case of gastric diaphragm. Malrotation was corrected in all cases. Per operative there was a sudden change in caliber of the duodenum and, frequently there was indentation at the probable site of diaphragm. [Fig 5] A large caliber nasogastric tube was manipulated into proximal distended part and air was insufflated, the distension and the indentation of the diaphragm would become more prominent. This technique of was helpful in 6 cases of duodenal diaphragms and 4 cases of gastric diaphragms.

A two days old of girl with double bubble sign and few specks of gas distally was explored for malrotation of gut, per operative she had two diaphragms first in the second part of and the second diaphragm close to fourth part of the duodenum. The distended proximal duodenum was plicated in three cases of duodenal diaphragm. A Foley's catheter was used to diagnose jejunal diaphragm, 22 Fr Foley's catheter was passed across the distended jejunum and into the collapsed bowel. The balloon of the Foley was inflated with saline solution and pulled proximally till the balloon hitched to the diaphragm. The diaphragm and a significant part of the distended bowel was excised.

Two cases of ileal diaphragm were found during emergency laparotomy. A 2 months old boy's ileum was perforated proximal to the diaphragm, while a 12 months old girl's ileal diaphragm accumulated melon seeds leading to acute intestinal obstruction [Fig 6].

A nine months old boy with anorectal malformation was found to have diaphragm during surgery.[Fig 7] A contrast enema done for constipation showed rectal stenosis in a two years old boy and during surgery a fenestrated rectal diaphragm was found. Excision of diaphragm and primary anastomosis was done through posterior saggital approach. Child developed leak from the anastomosis on third day, high sigmoid colostomy was done. The colostomy was closed after 8 weeks, the child is asymptomatic on follow up.



Figures 4-7: Intraoperative pictures of the diaphragm. There is marked difference in the caliber of the bowel before and after the indentation due to diaphragm.

Discussion

Congenital fenestrated diaphragms causing obstruction of intestines are rare. They are commonly found in duodenum. The estimated incidence of congenital duodenal fenestrated diaphragm varies from one in 9000 live births to one in 40,000 live births [4]. The incidence of jejunal, ileal and rectal diaphragms are even lesser.

The complete diaphragms are usually diagnosed at birth the present in neonates. The fenestrated diaphragms may often escape the diagnosis and the child may present at a later date with recurrent forceful vomiting, failure to thrive, upper abdominal dyspepsia, recurrent pneumonia and GERD. Rarely the diaphragm may present as intestinal obstruction in adult hood. Nazir et al reported late-onset primary gastric outlet obstruction in children with failure to thrive in a wide age group ranging from 3 months to 17 years[5].

Membranous obstruction, accounts for only 0.8% to 25% of all intestinal obstructions and 2% of all duodenal obstructions [6].The origin this membrane is a developmental anomaly. Various theories have been put forth from time to time. Bland -Sutton (1889) proposed that all intestinal stenosis or atresia occur where tissues fuse or where outgrowths occur. They suggested duodenal diaphragms as a "developmental" accident that occurred in the region of ampulla of Vater, from where the diverticula for the formation of liver and pancreas normally originate [7]. Tandler J (1900), while studying the development of the duodenum by dissection of intestinal tract of embryos from 30 to 60 days of gestational age suggested that during this stage of development the entire lumen of the intestine become occluded by proliferating epithelium and a solid core is formed, subsequently a stage of vacuolization occurs. Multiple large cystic spaces so created merge to form one single continuous lumen of intestine. Incomplete vacuolization of the solid core in any part of intestine develop into a diaphragm. The diaphragm may be complete or partial[8]. However this theory fails to explain the muscular component of the diaphragm. Boyden et al in 1967 studied embryos at various stages of development and observed that the ampullary region of duodenum is normally the narrowest part. And as the vacuoles coalesce, openings from the pancreatic and the biliary ducts join the future epithelial lining at the ampullary level. Any delay in vacuolization here will lead to stenosis in al already narrowed segment [9]. Saunders J.B and Lindner H.H proposed that the duodenum develops in four stages. Problems in the third stage would lead to Artesia, stenosis, or diaphragm formation, depending on the degree of abnormal development [10]. Louw J H and Barnard C N conducted elegant experiments in rat embryos and proposed that vascular accidents occurring in utero, particularly those which compromise the mesenteric blood supply result in narrowing or stenosis or atresia of intestine[11].

The diaphragms are usually single. But there can be multiple as is in one of our case where a second diaphragm was located 2.5cm distal to the first diaphragm in the 4th part of duodenum[12]. The fenestration may be skewed or in the center. The site may vary from the pyloric antrum to the rectum. The size of opening influences the course of the disease. Narrow opening frequently get obstructed in early neonatal period and presently early. Moderate size opening restricts the growth of the child, often these symptoms are attributed to gastroesophageal reflux and constipation. A great majority of these children are asymptomatic. A significant number of these cases are diagnosed in adult hood[13].

The clinical symptoms parallel with the diameter of the fenestration. Narrow opening in the diaphragm leads intractable vomiting in neonates necessitating in early intervention. In children with slightly larger opening the clinical features may range from frank projectile vomiting to nonspecific abdominal symptoms. Abdominal discomfort is usually located in the epigastrium. It is aggravated on eating solid food and relieved on vomiting, belching or assuming certain posture. Sometimes failure to thrive is the only visible symptom. If the diaphragm's fenestration is large then the presentation may be delayed by years or may be noticed only during autopsy. Kreg stated onset of symptoms in 30% of his series is after 24 years of age[14].

Plain abdominal radiography and ultrasonography only had good predictive values for duodenal diaphragms. Contrast-enhanced radiographic studies have better predictive value, about 89% in gastric and 100% in duodenal and jejunal diaphragms[15].The most diagnostic feature to diagnose the duodenal obstruction is retention of the barium

in the duodenum even after 6 hours[4]. One of our patient retained barium for over 24 hours. A congenital membranous obstruction may an incidental finding in a barium meal follow through[6].

Gastrointestinal anomalies like malrotation of gut, annular pancreas, and preduodenal portal vein are well documented. Seven cases of duodenal diaphragm were associated with malrotation of Gut. Cardiovascular and renal anomalies are also reported with lesser frequency. Two children in our series had Down's syndrome.

The older children may also develop atypical bleeding, duodenal ulcers; distal diaphragms may obstruct gall stones; marbles, seeds, peels may get stuck in the diaphragm [16] and cause acute intestinal obstruction and in duodenum it may cause cholangitis [4]. Recurrent Pancreatitis has also be reported in the individuals with duodenal diaphragm[17]. The histological examination of the excised membrane revels thickened the mucosa, submucosa and normal muscular layers [18]. The opening in the diaphragm may vary from 2mm to 10mm[6].

In 1922 Morton, suggested complete excision of duodenal diaphragm through duodenotomy, after studying a postmortem case [19]. He was successful in performing one such surgery in 1923. Subsequently many different surgeries have been described which include radial incision on the diaphragm, gastro- jejunostomy, side to side duodeno-duodenostomy, and duodenotomy with excision of the diaphragm and closing the longitudinal incision transversely. Although all the surgeries, yield good results by far the simplest and the most rational procedure is duodenotomy with excision of the diaphragm and closure of the longitudinal incision transversely. Plication of the distended duodenum helps in reducing the postoperative paralytic ileus and hastens intestinal functional recovery. All our patients with gastric and duodenal diaphragm underwent excision of the diaphragm and transverse closure of the incision. Three children whose proximal duodenum was markedly distended underwent plication. Ruangtrakool et al, studied different surgical techniques and reported no difference in postoperative feeding between duodeno-duodenostomy and diaphragm excision with duodenoplasty [20]. Escobar et al, also reported that duodenal atresia was rarely associated with early complications however 12% to 15% cases may have late complications, and about 6% of them may die later in life.[21] There was no mortality in this series, and no adhesion ileus was found in follow-up. Endoscopic excision of the diaphragm using laser has been reported[22].

The diaphragms of rectum are exceeding rare, their etiopathogenisis is unclear. Primary excision of the rectal diaphragm should be attempted with caution, we had performed high sigmoid colostomy in view of postoperative anastomotic leak. They may be associated with Anorectal malformation.

Conclusions

The diagnosis of the fenestrated diaphragms pose a clinical challenge, we propose that intraluminal obstruction should be considered in all cases whenever there is a sudden change in the caliber of intestine. The stomach or duodenum should be insufflated with air after passing the large bore catheter proximal to the suspected site of obstruction and confirm the free passage of air into distal duodenum / intestine. If any suspicion persists, a Foley's balloon catheter should be passed through an enterotomy across the suspected site of obstruction, and confirm the presence of obstruction.

Primary excision of the diaphragm and restoring the continuity of the bowel is the procedure of choice. Plication or excision of massively distended proximal bowel helps in early return of peristalsis. Rectal diaphragm should be excised and a temporary diverting colostomy should be considered.

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