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Congenital intrinsic duodenal obstruction [webs]- 13 case series and review of literature

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Abstract:
Aim: *to study the clinical presentation ,associated anomalies, management and outcome of children with duodenal webs over a period of 14 years in our department and review of literature*
Materials and methods*: It is a retrospective study of 13 children with duodenal webs were managed between 2000 to 2014 in our department of ped.surgery. The hospital records were retrieved and analysed.*
Results: *the age of presentation between 2 days to 3 years median age 15 days,weight between 0.75 kg to 7 kg mean 3.7 ,M:F=10:3, antenatal diagnosis was made in only 2 cases[15%]. commonest presentation was bilious vomiting.There was delay in presentation 3 cases. Associated anomalies were present in 9/13 patients.Lateral duodenotomy and excision of the obstructive membrane was done in 11 children and diamond shaped duodenoduodenostomy as described by Kimura in 2 ,in those who presented with annular pancreas. The mean hospital stay for the survivors was 11 days. Feeding neither throughTrans anastamotic stent[TAT]] norTotal parental nutrition (TPN) was given. The mortality rate was 3/13 (23%).*
Conclusions: *Congenital duodenal webs are rare and separate entity. Diagnosis can be missed especially in case of perforated webs. Survival has been increased to 45 to 95% due to improved diagnosis ,surgical, anesthesia and neonatal intensive care. Mortality depends mainly upon the presence of sepsis, prematurity, very low birth weight and associated anomalies.*
Key words: *congenital, duodenal atresia, Duodenal web,windsock deformity*

I. Introduction

Duodenal atresia and stenosis are intrinsic duodenal obstructions occurring 1in 6000 to 1 in 10000 live births[1]].Gray and Skandalakis have grouped duodenal atresia into 3 types .Type 1;-There is an obstructing septum[web]formed from mucosa and submucosa with no defect in the muscularis,the mesentry is intact.A variant of type1,a 'wind sock deformity' can occur if the membrane is thin and elongated. Type2;-A short fibrous cord connects the 2 blind ends of duodenum the mesentry is intact.Type 3;- There is no connection between the 2 blind ends of duodenum and there is a v shaped mesentric defect.[1]. A distinct pattern of clinical behavior was noted in type1, that highlighted the need to deal with this group of patients separately from the other subtypes.

II. Materials & Methods

It is a retrospective study a retrospective study was performed by retrieving the medical records of children who underwent surgery for duodenal obstruction.A total of 26 children with congenital duodenal obstruction were managed over a period of 14 years (2000 to 2014). Of these, 13 (50%) had Type I duodenal obstruction. Type 2 and 3 duodenal atresia children were excluded from this study. onlyType I duodenal obstructions[webs] were studied in detail with respect to the antenatal diagnosis, clinical presentation, associated anomalies, delay in diagnosis, surgical procedure and outcome.

III. Results

Age and sex:

The age distribution is shown in Table 1. The median age at presentation was15 days (range 2days to 3 years). There were 8 neonates. Four patients were born prematurely (32-34 weeks). There M: F ratio being 10:3.

Table1
Age of presentation

Age	Number of patients
Birth to 1 month	9
1month to 1 year	2
1year to 2 years	1
2year to 3yrs	1

Antenatal diagnosis

An antenatal diagnosis of duodenal obstruction on ultrasonography by charactaristic double bubble sign was made in only two patients (15%).Two mothers were diagnosed to have associated polyhydramnios(15%).

Clinical Presentation:

The commonest presentation was bilious vomiting in majority of the children. Three patients had vomited on and off became continuous for which medical help was sought. Upper abdominal fullness in 4,severe dehydration in 4 neonates.One patient presented with imperforate anus. One patient with associated esophageal atresia and tracheoesophageal fistula (EA with TEF) presented with frothing from the mouth.

Associated Anomalies:

The associated anomalies are malrotation of gut 3,High anorectal malformation was seen in one patient, Down's syndrome was seen in two patients,annular pancrease in two, EA with TEF in one.

Delay in Diagnosis:

Three patients who presented beyond one year of age group had fenestrated duodenal membranes causing partial obstruction were presented late (23%).

Investigations:

plain abdominal radiographs were taken,all shown characteristic double-bubble appearance (Fig. 1); in 3 patients who presented late,in addition to double-bubble appearance ,distal paucity of gas shadows were seen indicates partial obstruction. Upper gastrointestinal (GI) contrast study was performed for them confirmed partial obstruction. (Fig. 2).and for 3 year old child in addition gastroduodenoscopy performed shown diaphragm [web] with central perforation.

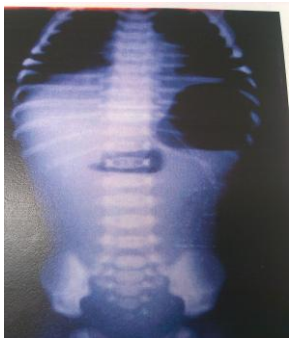

Fig. 1 Plain Radiograph showing double-bubble sign

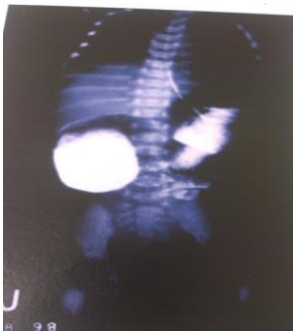

Fig. 2 GI contrast showing partial obstruction

Surgery:

After an initial hemodynamic stabilization and nasogastric decompression the patients underwent laparotomy through a right upper quadrant transverse muscle cutting incision. A lateral duodenotomy with excision of the obstructive membrane without injuring ampulla was done in 11patients (Fig. 3). The duodenotomy was closed transversely using 5'0 vicryl interrupted [delayed absorbable] sutures in a single layer. The location of the web was distal to ampulla ,The presence of the 'windsock' deformity was seen in 2 patients. Diamond shaped duodenoduodenostomy as described by Kimura for 2 who presented with annular pancreas for the fear of injury to pancreas, for 3 patients Ladd's procedure was done for associated malrotation,pelvic loop

colostomy for 1 who presented with high ARM and right postero-lateral throcotomy for EA+TEF . A trans-anastomotic tube (TAT)/TPN was not used in any children . Post operative ventilatory support and supportive intensive care were required in 4 children all were neonates.

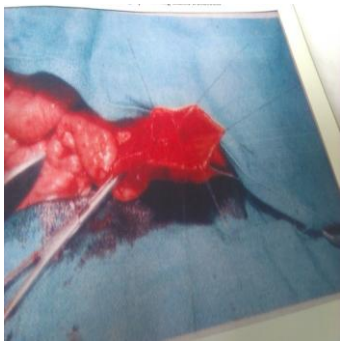

Fig.3 Operative photograph showing duodenal web

Outcome: There were 3 deaths in the postoperative period giving a mortality rate of 23%. The causes of death were low birth wt,sepsis with shock , leak in one did duodenoduodenostomy necessitating re- exploration ultimately died of sepsis and neonate with EA+TEF. The mean hospital stay for the survivors was 11 days (range 7-24 days). The mean time taken to achieve oral feeds in these 10 patients was 7 days (range 5 to 10] days). Two patients survived sepsis. For lack of numbers, any test of statistical significance could not be performed.

IV. Discussion

Calder reported first case duodenal atresia in 1733. Failure of recanalisation of duodenal lumen produces duodenal atresia or stenosis,total failure leads to atresia where as partial failure leads to a stenotic perforated membrane.[1] . Reports of different authors vary quoting the incidence of duodenal membranes from 0.8% to 92% [2,4,5]. Few studies have reported duodenal membranes as a separate entity with only occasional case reports from India [5-9].In a series of congenital intrinsic duodenal obstructions by Fronkalsurd et al, atresia was reported in 49%, membranes in 41% and stenosis in 10% cases [3].In a population based study of small intestinal atresia and stenosis over a 15 year period, Forrester et al have described the incidence of congenital duodenal obstructions as 1.3/ 10,000 live births [10].Antenatal diagnosis of congenital duodenal obstruction on ultrasonography is made by the presence of double-bubble appearance. Waever et al have reported that prenatal ultrasonography picked up duodenal atresia in 90% of cases (n=40) [5]. Although this a congenital anomaly, detected antenatally does not warrant medical termination of pregnancy or Caesarian section. As seen in three of our patients, those with fenestrated duodenal membranes may present as late as infancy or childhood or occasionally even in adulthood [1,12]. Although some studies have reported non-bilious vomiting as the most common presenting feature, majority of our patients had bilious vomiting indicating that the site of obstruction was post-ampullary [6,15]. A plain X-ray abdomen with a characteristic 'double-bubble' sign was diagnostic in most neonates. The most common site of location is between the first and second parts (85%) [1,15]. In a series of 10 patients, Rowe et al have described the location of a windsock anomaly to be preampullary in 40% of cases [8].Inability to pass a 10F Foley's catheter into the duodenum should raise the suspicion of a duodenal web. Only 32 cases of double duodenal webs have been reported in literature till date to the best of our knowledge out of which 2 were reported in adults [16-23]. Reid in his study of 140 patients of intrinsic duodenal obstructions, found only four double duodenal intrinsic obstructions, of which two were due to webs [16]. Stinger et al have reported four patients with double duodenal obstructions of which two were due to webs [17].The use of TAT for enteral feeding is controversial with no advantage being noted by some authors [4,25]. With advancement in pediatric intensive care and anesthesia and surgery the survival rates for duodenal obstructions have improved to 45- 95% in the developed world ,the major causes of mortality being associated congenital anomalies [1].

V. Conclusion

Congenital duodenal webs are rare and separate entity. By antenatal ultrasonography these anomalies can be picked up by characteristic double-bubble sign , Diagnosis can be missed especially in case of perforated webs. plain radiographs are sufficient to diagnose in many children. Survival has been increased to 45 to 95% due to improved diagnosis, surgical, anesthesia and neonatal intensive care .Mortality depends mainly upon the presence of sepsis, prematurity, very low birth weight and associated anomalies in developing countries. Mortality in our study is 23%.

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