



Duodenal Atresia: Clinical Presentation and Management in Tertiary Care Centre

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ABSTRACT

This prospective and retrospective cohort study done in 1 year period from march 2021 to march 2022 in tertiary care hospital in Indian population (mainly west India). The main aims of our study are- "Duodenal atresia- clinical presentation and management in tertiary care centre". All patient of intestinal atresia admitted in our centre in neonatal unit in 1 year period from March 2021 to March 2022, are data recorded and analysed. Data of demographics, antenatal history, presentation, location and type of IA (duodenal, jejuno-ileal, colonic) and peri-operative complications were recorded. After making our presumptive diagnosis, clinical assessment, an upright X-ray abdomen was taken and decision of surgery was taken. Statistical package for social sciences (SPSS 15.0 version, SPSS Inc, Chicago Ill) was used for data entry and analysis. Results were expressed as means, ranges and percentages. Males are 64 and females are 48 patients, M/F ratio was 1.3:1. average weight of atresia is 2.14 kg (ranges from 1.1-3.3 kg). mean age of presentation is 6.54 days (ranges from one day to 32 days). Patients have age more than one week only 28 (25%) patients found. Cases is highest in December and lowest cases in April month. Muslims population mostly associated with duodenal atresia. intestinal atresia included are duodenal atresia: (n = 44), Jejuno-ileal: (n = 60), Colonic: (n = 4), Multiple atresia: (n = 4). In duodenal atresia DA: Type-1 (n = 14) in which annular pancreas (n = 6) and perforated duodenal web (n = 4), DA: type-2 (n = 2) and DA: type-3 (n = 28) cases. In duodenal atresia, TEF associated with (n = 3), ARM associated with (n = 3) and ARM with TEF both associated with (n = 2) cases. in jejunoileal atresia type-1 (n = 10), Type-2 (n = 5), Type-3a (n = 36), Type-3b (n = 1), Type-4 (n = 12) are recorded. in colonic atresia type-1 (n = 3), type-2 (n = 0) and type-3 (n = 1) cases are recorded. in DA vomiting is chief complain while in JIA and IA abdominal distension, bilious vomiting and failure to pass meconium is chief complains. All patients go to surgical repair. We surprising result found in jejunal atresia patient with tapering enteroplasty with trans-anastomotic tube have 100% survival rates. Most common complication is functional obstruction followed by SSI and anastomotic leak. Mostly reoperation required in JIA cases. returning of bowel function is 4.3 days in DA, 6.2 days in JIA and 8.4 days in colonic atresia observed. Mean hospital stay for DA is 11.4 days, for jejunal atresia 12.8 days and for CA 11.5 days for survivors. Overall survival rates of surgery of intestinal atresia after surgery is 51.8%. for DA 59.1%, jejuno-ileal atresia: 50% and for CA: 50%. one month follow up comes with mainly cough and cold n = 7 (6.25%), fever n = 5 (4.46%), functional obstruction n = 2 (1.78%) and with sepsis in n = 1 (0.9%) found. Short-term survival of neonates with intestinal atresias in our unit is still poor when compared with statistics from developed countries. We trying to improve surgical outcomes in and understand difficulties faced by surgeons by this study.

INTRODUCTION

Congenital duodenal obstruction (CDO) is a common surgical anomaly in newborns that can be diagnosed prenatally and requires careful planning for surgical repair after birth^[1,2]. This chapter focuses on the surgical management duodenal atresia, a common form of CDO. With the rising popularity of laparoscopic surgery, duodenal atresia, one of the forms of CDO, also can be managed laparoscopically. However, use of a laparoscopic approach in these patients requires advanced experience in minimally invasive surgery and special equipment to perform such a demanding procedure in a limited operative space^[3,4].

Congenital duodenal obstruction accounts for 50% of all intestinal atresia cases^[1,2]. The incidence of the disease varies from 1 in 5000 to 1 in 10,000 newborns^[5-8]. Limited information is available about hereditary forms of CDO. Unlike other types of congenital intestinal obstruction, duodenal obstruction has a high association with other anomalies; such concomitant anomalies are reported in 38% Atresia-Pathology and Therapeutic Approach 2 of patients with CDO^[9,10]. The most common associated disease reported is Down syndrome registered in 25-46% of cases^[6,7-11]. Other associated anomalies include intestinal malrotation (54%), congenital cardiac anomalies (32-48%), esophageal atresia (9%), renal anomalies (5%) and anorectal malformations (7%). These anomalies can be part of VACTERL syndrome or isolated^[11]. 12% of patients with duodenal atresia have an associated anomaly of the biliary tract, such as biliary atresia. Associated diseases tend to determine the postoperative course in patients with duodenal atresia. In patients with associated esophageal atresia or cardiac defects-most often a complete atrioventricular septal defect high mortality rates have been reported.

In most cases, the diagnosis of duodenal obstruction can be established prenatally. Duodenal obstruction develops approximately by 12-14 weeks of fetal development, so there is no possibility of earlier detection of this anomaly. Ultrasound is used to define the "double bubble sign." These are two fluid levels, one in the distended stomach and the other one in the duodenum. Polyhydramnios develops in pregnancies complicated by duodenal obstruction. Postnatally, the diagnosis of duodenal obstruction is confirmed in an abdominal X-ray, showing the "double bubble" sign described above. Abdominal ultrasound is necessary to detect not only duodenal atresia but also to find concomitant anomalies and rare forms of situs inversus. These findings can necessitate alternative port placement during laparoscopy^[12].

Currently, the standard method of recanalizing the duodenal lumen is a diamond-shaped duodenal anastomosis. The introduction of minimally invasive

laparoscopic instruments, optical systems with small diameters and high-resolution screens has expanded the potential of laparoscopy. These developments have increased the interest of paediatric surgeons in laparoscopy as a modality for reconstruction in patients with CDO. However, laparoscopic duodenal anastomosis is considered the most demanding surgical procedure in paediatric surgery. Therefore, this procedure is restricted to be performed in advanced centres specializing in minimally invasive surgery in neonates. Duodenal stenosis is most often located in the second (descending) portion of the duodenum. Historically, such patients were treated surgically with laparotomy or laparoscopy. Despite the results of such treatment being satisfactory, these techniques are fraught with risks associated with the operation itself and general anaesthesia and do not show good cosmetical results. In addition, the risk of adhesive intestinal obstruction in infants after laparotomy is approximately 6-14% and is absent if the peritoneum is left intact, as, for example, with transoral access.

Intestinal atresia is one of the most common and leading causes of neonatal intestinal obstruction (NIO) and second most common cause of NIO in many developing countries^[1-3]. Mostly these patients, DA and JIA occur separately but sometimes they occur together in one patient^[4]. In most developed countries surgical outcomes are improved due to various causes like availability of prenatal diagnosis, awareness in parents, early presentation of clinical features, availability of paediatric surgeons, availability of neonatal parenteral nutrition, neonatal anaesthesia, better postoperative care and neonatal surgical intensive care services^[5,6]. In many low and middle income countries (LMICs), outcome has remained poor due to improper and less availability of these facility^[1,7].

It is a congenital obstruction of the intestine, sometimes associated with a loss of tissue, resulting in a disruption of intestinal continuity. The incidence of intestinal atresia is approximately 1 in 4000 live births. Etiopathogenesis of intestinal atresia is failure of recanalization of the initial solid-core phase of intestinal development and in utero vascular accident is the cause of it hypothesized. This can occur anywhere in the intestinal tract from duodenum, jejunioileal region and colon. DA mostly associated with other congenital anomalies, most commonly down syndrome and associated with imperforated anus. Jejunioileal atresia's occurs from Treitz ligament to ileocecal valve anywhere and mainly associated with cystic fibrosis and malrotation. Colonic atresia's unusual in that they found in same anatomical region of colon [transverse colon] and with same degree of

severity [lumen and mesentery loss. This is present with various degree of severity like mucosal web to complete lumen or mesentery loss and cause multiple atresia throughout the bowel^[13].

In this study, we basically highlight the short-term outcome of surgical management of intestinal atresia in our paediatric surgery unit.

Aims and objectives: This prospective and retrospective cohort study done in one-year period from march 2021 to march 2022 in tertiary care hospital in Indian population [mainly west India]. The main aims of our study are "Duodenal atresia- clinical presentation and management in tertiary care centre".

MATERIALS AND METHODS

All patient of intestinal atresia admitted in our centre in neonatal unit in one-year period from March 2021 to March 2022, are data recorded and analysed. All intestinal atresia patient selected and their history, demographic data, medical detail, treatments, surgical outcome, hospital stay and complications are recorded. In detail we also record the data antenatal history, presentation, location and type of IA (duodenal, jejuno-ileal, colonic) and peri-operative complications were recorded.

Patients who are died before operation or making definitive diagnosis, with volvulus, complicated meconium ileus, gastroschisis and who leave/discharge against medical advice, were excluded.

After making our presumptive diagnosis, clinical assessment, an upright X-ray abdomen was taken and decide that intestinal atresia patients, vitally stabilised by intravenous fluid and antibiotics. Nasogastric (NG) tube was inserted in all patients. After making definitive diagnosis, plan for surgery. Postoperatively antibiotics continued and NG tube was removed after when peristalsis movement comes or when NG aspirate was gastric and $<15 \text{ mL day}^{-1}$ and thereafter gradual feeding was started. Follow up for one months was taken.

Statistical Package for Social Sciences (SPSS 15.0 version, SPSS Inc, Chicago Ill) was used for data entry and analysis. Results were expressed as means, ranges and percentages.

RESULTS

There are 136 patients are studied, in which 6 patients expired before operation, 7 patients taken LAMA, 5 patients come with previously operated, 5 patients come with atresia with meconium ileus and 1 patient comes with atresia with gastroschisis. These all patients are excluded from data, so we analysed only 112 patients only who goes to operation. We found these results from our study:

- Males are 64 and females are 48 patients, M/F ratio was 1.3:1
- Average weight of atresia is 2.14 kg [ranges from 1.1- 3.3kg]
- Mean age of presentation is 6.54 days [ranges from one day to 32 days]. Patients have age more than one week only 28 [25%] patients found
- Monthly distribution of cases is highest in December and lowest cases in April months (Fig. 1)
- Muslims population mostly associated with duodenal atresia while Hindu population mostly associated with all intestinal atresia
- We found that state wise distribution of cases from Rajasthan [n = 95], Haryana [n = 6], Madhya-Pradesh [n = 1] and from Uttar-Pradesh [n = 10] cases found
- Intestinal atresia included are duodenal atresia: [n = 44], Jejuno-ileal: [n = 60], Colonic: [n = 4], Multiple atresia: [n = 4].
- In duodenal atresia DA: type-1 [n = 14] in which annular pancreas [n = 6] and perforated duodenal web [n = 4], DA: type-2 [n = 2] and DA-type: 3 [n = 28] cases (Fig. 2)
- In duodenal atresia, trachea-oesophageal fistula associated with [n = 3], anorectal malformation associated with [n = 3] and anorectal malformation with trachea-oesophageal both associated with [n = 2] cases
- In jejunileal atresia type-1 [n = 10], Type-2 [n = 5], Type-3a [n = 36], Type-3b [n = 1], Type-4 [n = 12] are recorded
- In colonic atresia type-1 [n = 3], type-2 [n = 0] and type-3 [n = 1] cases are recorded.
- In DA vomiting is chief complain while in JIA and IA abdominal distension, bilious vomiting and failure to pass meconium is chief complains

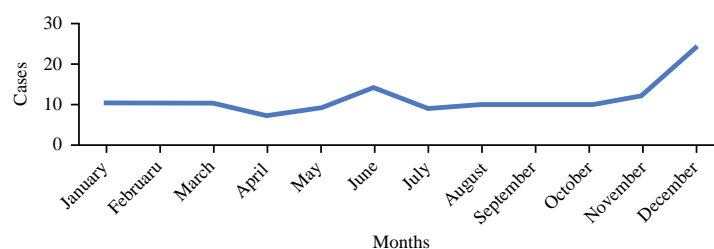


Fig. 1: Monthly distribution highest and lowest cases

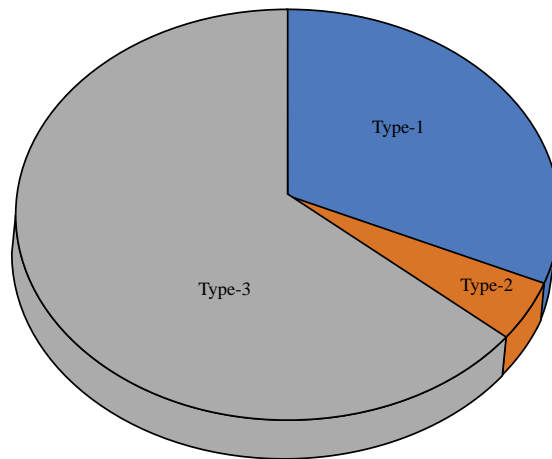


Fig. 2: Duodenal atresia

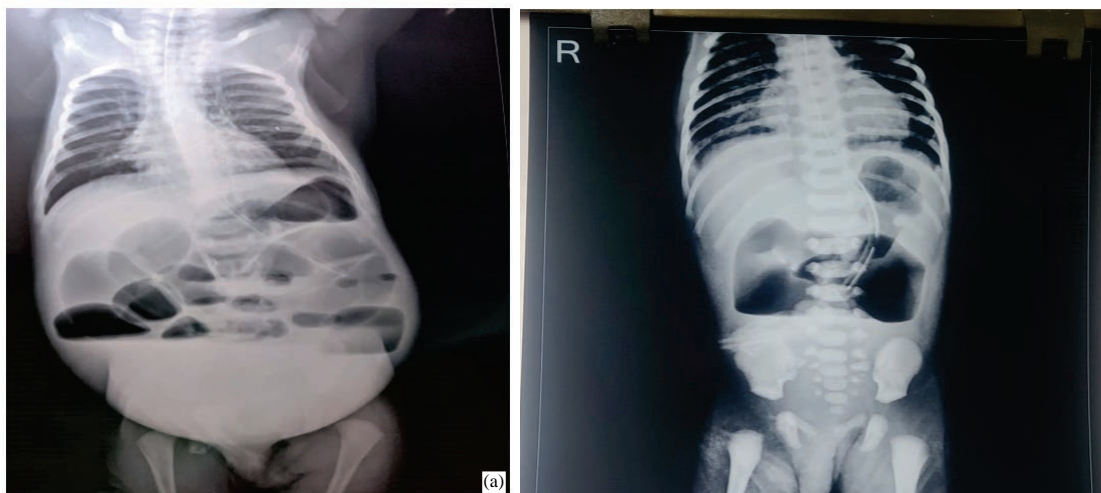


Fig. 3(a-b): (a) Multiple ileal atresia radiography: Multiple bubble sign and (b) Duodenal atresia: Double bubble sign

Table 1: Jejunal atresia patient with anti-mesenteric tapering enteroplasty with trans-anastomotic tube

Type	Cases	Surgical procedure	No.	Survival rates (%)
DA	44	Kimura's duodenoduodenostomy	40	22 (55)
		Excision of duodenal web	4	4 (100)
JIA	60	Resection anastomosis	48	18 (37.5)
		Tapering enteroplasty with transanastomotic tube	4	4 (100)
		Ileostomy	8	7 (87.5)
CA	4	Resection anastomosis	1	0 (0)
		Resection and colostomy	2	1 (50)
		Resection anastomosis with proximal ileostomy	1	1 (100)
Multiple atresia				
DA+IA	3	Resection and anastomosis with kimura's	3	0 (0)
DA+JA	1	Kimura's with resection and anastomosis	1	0 (0)
Total	112		112	58 (58.1)

- In cases of DA, Double-bubble sign on X-ray abdomen was found, while in perforated duodenal web, distal gas with double-bubble was seen. In triple atresia patients, shows double-bubble sign with red rubber catheter in upper oesophageal pouch. In perforated duodenal web patient upper-GI gastrografen contrast study was performed because of diagnostic dilemma due to presence of gas in distal bowel on X-ray. In JIA, Tripple bubble sign and multiple dilated bowel loops and multiple air-fluid levels seen. Gastrografen enema was performed in cases of ileal atresia (Fig. 3a-b)
- Surgical treatment done in all cases after resuscitating the patient and proper investigation. We surprising result found in jejunal atresia patient with anti-mesenteric tapering enteroplasty with trans-anastomotic tube have 100% survival rates. Another procedure are followings (Table 1):
- There are postoperatively many complications seen. Most common complication is functional obstruction followed by SSI and anastomotic leak. Mostly reoperation required in JIA cases (Table 2)

Table 2: Postoperatively complications

Complications	Cases	Percentages	Management
Surgical site infection	7	21.2	Dressing
Burst abdomen	2	6	Secondary suturing
Anastomotic leak	7	21.2	No intervention = 3 RA = 1 EL with ileostomy = 3
Proximal perforation	3	9	Primary repair with ileostomy = 2 Primary repair = 1
Functional obstruction	9	27.3	Conservative = 7 EL with re-anastomosis = 2 conservatively
Aspiration pneumonia	5	15.15	
Total	33		

- Returning of bowel function is 4.3 days in DA, 6.2 days in JIA and 8.4 days in colonic atresia observed
- Mean hospital stay for duodenal atresia is 11.4 days, for jejunal atresia 12.8 days and for colonic atresia 11.5 days for survivors
- Overall survival rates of surgery of intestinal atresia after surgery is 51.8%. for duodenal atresia 59.1%, jejuno-ileal atresia: 50% and for colonic atresia: 50%
- One months follow up also taken in most operated patients which comes with mainly cough and cold n = 7 [6.25%], fever n = 5 [4.46%], functional obstruction n=2 [1.78%] and with sepsis in n = 1 [0.9%] found

DISCUSSIONS

Congenital duodenal obstruction (CDO) is a common surgical anomaly in newborns, that can be diagnosed prenatally and requires careful planning for surgical repair after birth. This chapter focuses on the surgical management duodenal atresia, a common form of CDO. With the rising popularity of laparoscopic surgery, duodenal atresia, one of the forms of CDO, also can be managed laparoscopically. However, use of a laparoscopic approach in these patients requires advanced experience in minimally invasive surgery and special equipment to perform such a demanding procedure in a limited operative space.

IA is one of the most common causes of neonatal intestinal obstruction. In detection of duodenal atresia than JIA or CA, Prenatal ultrasonography is more reliable. Basu and Burge reported that 31% of patients with small bowel atresia could be diagnosed on prenatal ultrasound^[6].

Pre-operative management in all patients require includes, primary resuscitation, correction of dehydration and electrolyte abnormalities., echocardiography and ultrasonography of the abdomen should be performed in all patients because of high incidence of cardiac and renal anomalies associated with DA^[7]. In this study only n = 10 [22.7%] DA patients associated with cardiac anomalies and only n = 4 [9%] associated with imperforated anus. similarly in a series of 138 cases of DA, 38% cardiac anomalies, 14% renal anomalies, 6% EA with TEF and 5% imperforated anus found^[5].

In duodenal atresia, trachea-oesophageal fistula associated with 6.8% [n = 3], anorectal malformation associated with 6.8% [n = 3] and anorectal malformation with trachea-oesophageal both associated with 4.5% [n = 2] cases.

After making definitive diagnosis we perform surgical procedure on basis of type of atresia, degree of dilatation of proximal segment, patients' condition. In DA, Kimura's diamond shaped duodeno-duodenostomy performed. In type-I DA, duodenotomy with excision of web is an option which is performed less commonly due to chances of damage to ampulla of Vater^[5]. Only 4 cases are managed by this procedure

Duodeno-jejunostomy is another surgical option in difficult cases, because of patient anatomy particularly in small and premature children. Anti-mesenteric tapering duodenoplasty is advised as a useful technique, only done for managing duodenal motility disorder related to mega-duodenum^[10].

In this study peri-operative mortality is almost same for DA, JIA and CA. Outcome of IA is still not satisfactory in developing countries^[10].

Returning of bowel function is 4.3 days in DA, 6.2 days in JIA and 8.4 days in colonic atresia observed. This implies that bowel function returns earlier in DA than in JIA^[5,6]. Excision or tapering of proximal dilated bowel may help in earlier return of bowel function.

The re-operation rate of around 10% is quite high and was for anastomotic leakage and anastomotic stricture. Most of the re-operations were for JIA.

mean hospital stay for duodenal atresia is 11.4 days, for jejunal atresia 12.8 days and for colonic atresia 11.5 days for survivors. This difference in duration of hospital stay could be explained by earlier return of bowel function in DA as well as the absence of re-operation and less complications in most cases of DA^[15].

Functional obstruction and anastomotic leaks are the major surgical complications.

One month follow up also taken in most operated patients which comes with mainly cough and cold n = 7 [6.25%], fever n = 5 [4.46%], functional obstruction n = 2 [1.78%] and with sepsis in n = 1 [0.9%] found. Average follow-up duration of 30 days is low when compared with developed countries, it can be enhanced to detect mortality and morbidity^[13].

Surgical outcome in developed and developing countries is not because of only surgical techniques, rather because of advancement in the following: availability of good primary health care with early referral and proper transportation system, availability of neonatal surgeons, parallel growth of neonatal anaesthesia, expert neonatologist, TPN and neonatal ICU with trained personnel^[16-18].

In developing countries, delayed in making diagnosis, late arrival at hospital, poor primary health care management, lack of equipped neonatal ICU with trained personnel, overcrowding which leads to cross infection and septicemia and less availability of TPN, are some primary reasons for high mortality rates. Cardiac anomalies are more commonly associated with DA than JIA and are one of the major obstacles to successful outcome in case of DA^[13].

CONCLUSION

We concluded that survival rates of neonates with intestinal atresia is poor in developing country, high volume tertiary care centre in India. It is not only cause of surgical technique and expertise but multifactorial causes like delayed presentation, high patients load at hospitals, overcrowding in the ICU and septicemia. It leads to short term survivals failure of patients. Late presentation is common in this series but does not appear to negatively affect the outcome as meticulous pre-operative resuscitation is emphasized. A high proportion of the mortalities had re-operation for anastomotic leak. From this study we found that we improve surgical outcomes by improving prenatal diagnosis, early referral to higher centre, planned delivery in centres, establishment of neonatal surgical intensive care unit, encouraging sub-specialization in neonatal anaesthesia, early involvement of paediatrics surgeon in postnatal assessment of neonates and modification of some surgical technique [like antimesenteric tapering enteroplasty with tube in jejunal atresia patients], use of TPN and adequate investigations for congenital cardiac anomalies, may improve the outcome.

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