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Research Article

Duodenal Atresia- Clinical Presentation and Management in Tertiary Care Centre

Atul Khare *

Department of surgery, Government Medical College Shadol, India.

***Corresponding Author:** Atul Khare, Department of surgery, Government Medical College Shadol, India.

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Abstract

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. This study focuses on the surgical management of duodenal atresia. 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. In this study, we analysed the short term outcome of surgical management of intestinal atresia in our unit.

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

Materials and methods: All patients of intestinal atresia admitted in our centre in neonatal unit in one-year period from march 2021 to march 2022 were included, the 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 with clinical assessment, an upright X-ray abdomen was taken and Decision for surgery was finalised.

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: 64 patients were male and 48 of them were female, M:F ratio was 1.3:1. Average weight of atresia was 2.14 kg [ranges from 1.1- 3.3kg] and mean age of presentation was 6.54 days [ranges from one day to 32 days]. Patients having age more than one week were only 28 [25%] in number. Cases were highest in December and lowest in the month of April. Muslim population were mostly associated with duodenal atresia. Intestinal atresia included different variants as follows, duodenal atresia- [n=44], Jejuno-ileal- [n=60], Colonic- [n=4], Multiple atresia- [n=4]. In duodenal atresia the distribution was as follows, 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] were recorded. In colonic atresia type-1 [n=3], type-2 [n=0] and type-3 [n=1] cases were recorded. In DA vomiting was the chief complaint while in JIA and IA abdominal distension, bilious vomiting and failure to pass meconium were the chief complains. All patients required surgical repair. A surprising result was found in jejunal atresia patients treated with tapering enteroplasty with trans-anastomotic tube with 100% survival rates. Most common complication was functional obstruction followed by SSI and anastomotic leak. Most of the patients required reoperation in JIA cases. Return of bowel function was seen in 4.3 days in DA, 6.2 days in JIA and 8.4 days in colonic atresia. Mean hospital stay for DA was noted to be 11.4 days, for jejunal atresia it was around 12.8 days and for CA it was found to be 11.5 days for survivors. Overall survival rates of surgery of intestinal atresia after surgery was 51.8%, among which DA accounted to 59.1%, jejuno-ileal atresia- 50% and for CA- 50%. During the one month follow up period,

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%] were found.

Conclusions: Short term survival of neonates with intestinal atresias in our unit is still poor when compared with statistics from developed countries. Thus efforts are being made to improve the surgical outcome and to understand difficulties faced by surgeons with the help of this study.

Key Words: ileal atresia; exploratory laparotomy; neonatal intestinal obstruction

Introduction

Congenital duodenal obstruction (CDO) is a common surgical anomaly in newborns [1, 2] that can be diagnosed prenatally and requires careful planning for surgical repair after birth. This study focuses on the surgical management of duodenal atresia, a common form of CDO. With the rising popularity of laparoscopic surgery, duodenal atresia, one of the forms of CDO, can also be managed laparoscopically. However, use of a laparoscopic approach in these patients requires enormous experience in minimally invasive surgery along with supply of special equipments to perform such a technically challenging 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 of patients with CDO [9, 10]. The most common associated disease reported is Down syndrome registered in 25–46% of cases [11–13]. 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 [13]. 12% of patients with duodenal atresia have an associated anomaly of the biliary tract, such as biliary atresia [21, 22]. 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 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 as 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.

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 atresia 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, jejunoileal region and colon. DA mostly associated with other congenital anomalies, most commonly down syndrome and associated with imperforated anus. Jejunoileal atresia’s occurs from Treitz ligament to ileocecal valve anywhere and mainly associated with cystic fibrosis and malrotation. Colonic atresias 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.

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 a 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’’.

Patients and Methods

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

Patients who died before the operation or before making any definitive diagnosis, with volvulus, complicated meconium ileus, gastroschisis and who left or got discharged against medical advice, were excluded.

After making our presumptive diagnosis with clinical assessment, an upright X-ray abdomen was taken and confirmed to have intestinal atresia. Patients were then vitally stabilised by intravenous fluid and antibiotics. Nasogastric (NG) tube was inserted in all patients. After making definitive diagnosis, plan for surgery was made. Postoperatively antibiotics were continued and NG tube was removed after peristalsis was re-established or when the NG aspirate was found to be gastric and < 15ml/day, 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

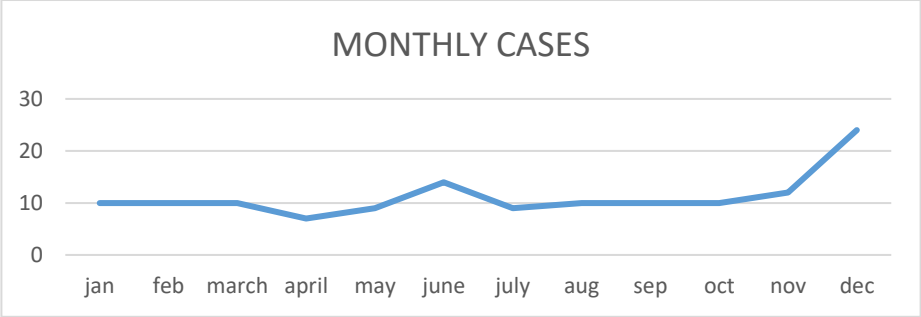
A total of 136 patients were studied, in which 6 patients expired before operation, 7 patients went LAMA, 5 patients came with previous history of operation, 5 patients came with atresia with meconium ileus, and 1 patient came with atresia associated with gastroschisis. These patients were excluded from the data, thus we analysed only 112 patients who required operation. We found these results from our study-

1] males were 64 and females were 48 patients, M/F ratio was 1.3:1.

2] average weight of atresia was 2.14 kg [ranges from 1.1- 3.3kg]

3] mean age of presentation was 6.54 days [ranges from one day to 32 days]. Patients having age more than one week were only 28 [25%].

4] monthly distribution of cases was highest in December and lowest in April months.

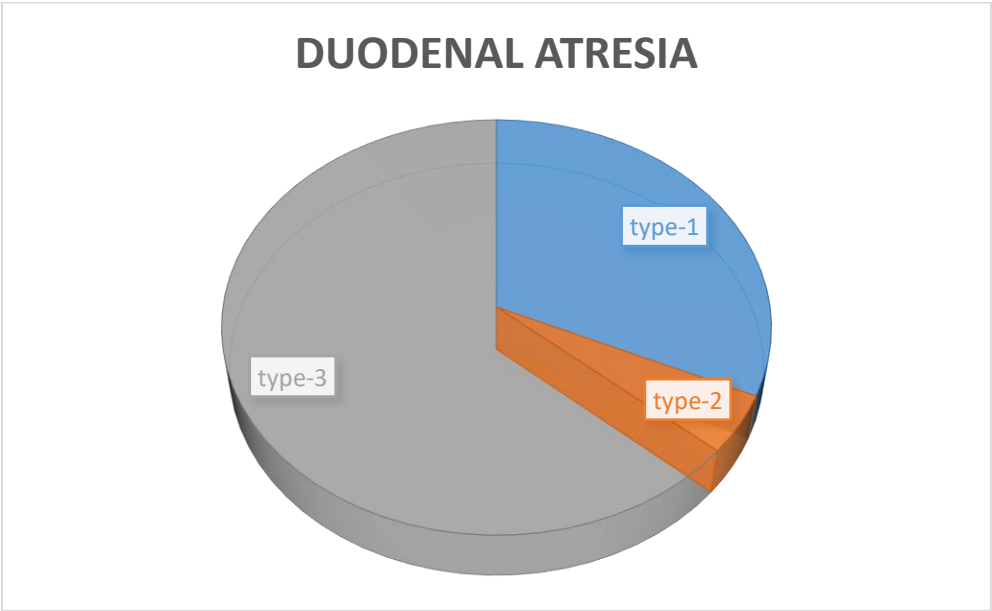


5] Muslim population was found to be mostly associated with duodenal atresia while Hindu population were mostly associated with all intestinal atresias.

6] 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

7] Intestinal atresia included duodenal atresia in [n=44], Jejuno-ileal in [n=60], Colonic in [n=4], Multiple atresias in [n=4].

8] 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.



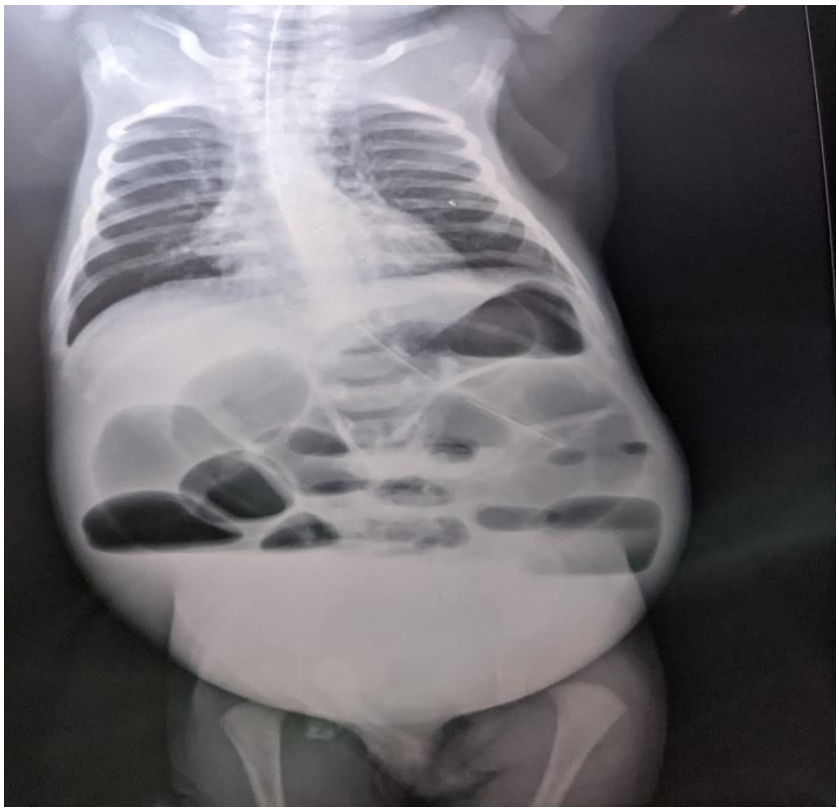
9] In duodenal atresia, trachea-oesophageal fistula associated with [n=3], anorectal malformation associated with [n=3] and anorectal malformation with trachea-oesophageal both associated with [n=2] cases.

10] 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.

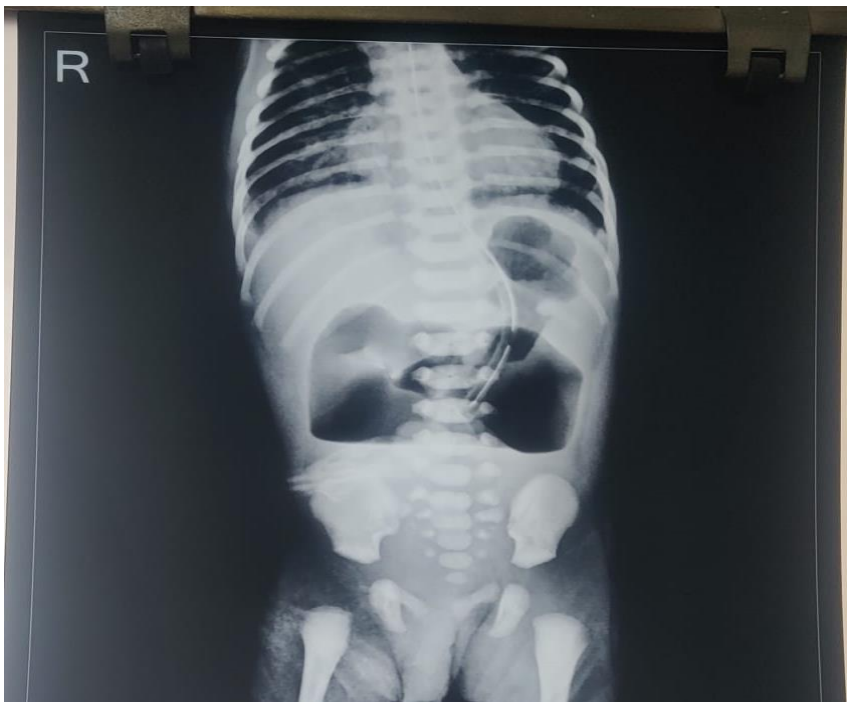
11] in colonic atresia type-1 [n=3], type-2 [n=0] and type-3 [n=1] cases are recorded.

12] in DA vomiting is chief complain while in JIA and IA abdominal distension, bilious vomiting, and failure to pass meconium is chief complains.

13] 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, double-bubble sign with red rubber catheter in upper oesophageal pouch was found. In perforated duodenal web patient upper-GI gastrografin 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 were seen. Gastrografin enema was performed in cases of ileal atresia.



Multiple ileal atresia radiography= multiple bubble sign



Duodenal atresia= double bubble sign

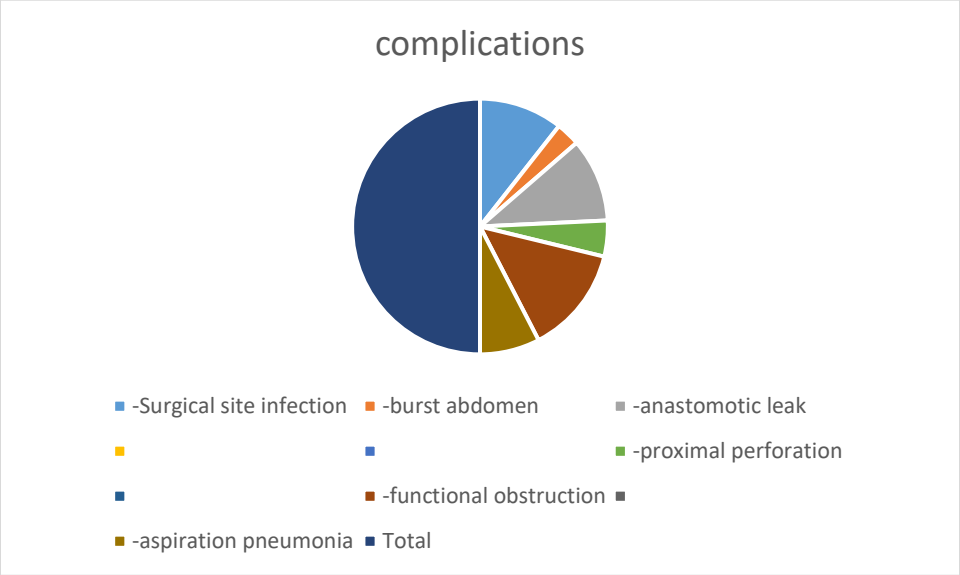
14] Surrgical treatment was done in all cases after resuscitating the patient and proper investigations. A surprising result was found in jejunal atresia patients with anti-mesenteric tapering enteroplasty with trans-anastomotic tube showing 100% survival rates. Other procedures used were as follows-

Type	Cases	Surgical procedure	Number	Survival rates
DA	44	-Kimura's dudeno-dudenostomy -excision of duodenal web	40 4	22 [55%] 4 [100%]
JIA	60	-resection anastomosis -tapering enteroplasty with trans-anastomotic tube	48 4	18 [37.5%] 4 [100%]

		-ileostomy	8	7 [87.5%]
CA	4	-resection anastomosis -resection and colostomy -resection anastomosis with proximal ileostomy	1 2 1	0 [0%] 1 [50%] 1 [100%]
Multiple atresia -DA+IA -DA+JA	 3 1	 -resection and anastomosis with kimura's -kimura's with resection and anastomosis	 3 1	 0 [0%] 0 [0%]
Total	112		112	58 [58.1%]

15] there were many postoperative complications. Most common complication being functional obstruction followed by SSI and anastomotic leak. Mostly reoperation was required in JIA cases.

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
-aspiration pneumonia	5	15.15%	conservatively
Total	33		



16] return of bowel function was seen in 4.3 days in DA, 6.2 days in JIA and 8.4 days in colonic atresia.

17] mean hospital stay for duodenal atresia was 11.4 days, for jejunal atresia 12.8 days and for colonic atresia 11.5 days for survivors.

18] Overall survival rates of surgery of intestinal atresia after surgery was 51.8%. for duodenal atresia 59.1%, jejuno-ileal atresia- 50% and for colonic atresia- 50%.

19] one months follow up also taken in most operated patients which mainly showed 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%] .

Discussion

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 of 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 [11].

Pre-operative management was needed in all patients which included, 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 [12]. 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 performed surgical procedure on the basis of type of atresia, degree of dilatation of proximal segment and patients' condition. In DA, Kimura's diamond shaped duodeno-duodenostomy was performed. In type-I DA, duodenotomy with excision of web is an option which was performed less commonly due to chances of damage to ampulla of Vater. [5] Only 4 cases were 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 was almost same for DA, JIA and CA. Outcome of IA is still not satisfactory in developing countries [16].

Return of bowel function was seen in 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 rates of around 10% is quite high and was mainly for anastomotic leakage and anastomotic stricture. Most of the re-operations were for JIA cases.

Mean hospital stay for duodenal atresia was 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.

Functional obstruction and anastomotic leaks were the major surgical complications.

One month follow up also taken in most operated patients of which complaints were 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 should be extended to detect mortality and morbidity.[15]

Difference in surgical outcomes in developed and developing countries is not only because of surgical techniques, but also 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, use and availability of TPN and state of art neonatal ICU facilities with trained personnel.

In developing countries, delay in making diagnosis, late arrival at hospital, poor primary health care management, lack of equipped neonatal ICU with trained personnel, overcrowding leading to cross infection and septicaemia, 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 [15]

Conclusion

We concluded that survival rates of neonates with intestinal atresia is poor in developing country, in a high volume tertiary care centre in India. It is not only because of surgical techniques and expertise but due to multifactorial causes like delayed presentation, high patients load at hospitals, overcrowding in the ICU and septicaemia. It leads to failure of short term survivals 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 were seen in cases with re-operation for anastomotic leak.

From this study we found that we can improve surgical outcomes by improving prenatal diagnosis, early referral to higher centre, planned delivery in centres, establishment of neonatal surgical intensive care units, encouraging sub-specialization in neonatal anaesthesia, early involvement of paediatrics surgeons in postnatal assessment of neonates and modification of some surgical techniques [like antimesenteric tapering enteroplasty with tube in jejunal atresia patients], use of TPN, and adequate investigations for congenital cardiac anomalies, which may improve the outcome.

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