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Original Research Article

Jejunoileal Atresia: A 5 Year Institutional Review of Surgical Outcomes, Prognostic Factors and the Impact of Delayed Presentation in a Resource-Limited Setting

Arkaprovo Roy[1], Rajarshi Kumar[2], Aloke Kumar Sinhababu[3]

[1]MS General Surgery, Department of General Surgery, North Bengal Medical College and Hospital, Siliguri, West Bengal 734012

[2]M.Ch (Pediatric Surgery), Department of Pediatric Surgery, North Bengal Medical College and Hospital, Siliguri, West Bengal 734012

[3]MBBS, DCH, MS (General Surgery), M.Ch (Pediatric Surgery), Department of Pediatric Surgery, North Bengal Medical College and Hospital, Siliguri, West Bengal 734012

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Corresponding Author: Dr. Arkaprovo Roy

Conflict of interest: Nil

Abstract:

Introduction: Jejunoileal atresia (JIA) is a frequent cause of intestinal obstruction in the newborn and it occurs in 1-3 per 10,000 live births. The condition is caused by vascular accident in utero. The antenatal diagnosis is difficult even though prenatal imaging has improved. Neonates with JIA normally manifest within the first 24-48 hours of birth with biliary emesis and inability to empty meconium with or without abdominal distension. The gold standard is surgical resection of the atresia segment and end to end anastomosis. When neonates are in severe sepsis or in poor general state or in severe bowel distension, other surgical methods- stoma or a chimney procedure (e.g Santulli) can be used. Survival rates in developed countries are more than 90% due to early diagnosis, neonatal intensive care advancements, pediatric anaesthesia expertise and access to total parenteral nutrition (TPN).

Aims and Objectives: This paper seeks to carry out an institutional review based on cases of jejunoileal atresia that have been conducted over a period of 5 years basing on short term outcomes and survival influencing factors.

Materials and Methods: A retrospective study was conducted on neonates admitted with jejunoileal atresia who were operated between January 2020 to December 2024. The data that was recorded include the demographics, clinical presentation, and preparation, surgical details and outcomes along with associated anomalies if any. To establish the significant predictors of survival, statistical analysis was done.

Results: 33 neonates with jejunoileal atresia were operated with a male-female ratio of 1.36:1. 79 % were low birth weight, and 64 % were very low birth weight. The average age of presentation was around 9.5 days. Our study indicated the timing of presentation and birth weight as significant factors of survival. Even though the overall survival which was 72 % at the time of initial discharge had declined to 64 % at least 6 months follow up, the switchover to Santulli (chimney) procedure in the final 2 years yielded a survival of more than 90 %

Conclusions: Neonates with late presentation and low birth weight did poorly. High index of suspicion of intestinal obstruction in neonates who have biliary emesis is essential for early diagnosis. Survival might be enhanced by antenatal diagnosis by intervening in time. A chimney operation such as Santulli, which was found to have a higher survival rate than primary anastomosis in neonates of unfavorable general condition and massive luminal disparity, has turned out to be saviour in resource-limited tertiary centres.

Keywords: Jejunoileal atresia, delayed presentation, Santulli procedure.

Introduction

Jejunoileal atresia (JIA) is a frequent cause of intestinal obstruction in the newborn and it occurs in 1-3 per 10,000 live births. [1,2] The condition is caused by vascular accident in utero. The antenatal diagnosis is difficult even though prenatal imaging has improved. [3,4] Neonates with JIA normally manifest within the first 24-48 hours of birth with biliary emesis and inability to empty meconium with or without abdominal distension. [5,6] The gold standard is surgical resection of the atresia segment and end to end anastomosis. When neonates are in severe sepsis or in poor general state or in severe bowel distension, other surgical methods- stoma or a chimney procedure (e.g Santulli) can be used. [7] Survival rates in developed countries are more than 90% due to early diagnosis, neonatal intensive care

advancements, pediatric anaesthesia expertise and access to total parenteral nutrition (TPN). [8,9]

Rationale for the study

Although the world has made some improvements in the neonatal surgery, the treatment of JIA in the resource-poor countries is complicated and the best surgical approach to be used in the neonates is still under contention. [10,11] The study was carried out in a tertiary care centre with a wide catchment area.

We aimed at examining the results of neonates with JIA in the last 5 years with emphasis on how the age of presentation, birth weight, various surgical interventions influence survival and determine the factors that lead to morbidity and mortality in our context. The assessment of these factors will allow us to give evidence-based recommendations to enhance the survival and quality of care of neonates with JIA in analogous healthcare settings.

Material and Methods

Study Area: North Bengal Medical College Department General Surgery

Study Population: This was a retrospective institutional review evaluating cases of jejunoileal atresia (JIA) operated between January 2020-December 2024 at a tertiary care referral centre serving neonates from North Bengal, Bihar and even cross border patients from neighbouring countries.

The centre is a resource constrained one as there is limited access to antenatal diagnosis, neonatal intensive care beds and total parenteral nutrition (TPN).

Inclusion Criteria: Neonates with confirmed JIA at exploratory laparotomy at our tertiary care centre between January 2020 to December 2024 with a minimum of 6 months post discharge follow up. The diagnosis of JIA was strictly intra operative.

Exclusion Criteria: Neonates with incomplete medical records (missing operative details or follow up) Patients lost to follow up before 6 months post discharge.

Data Collection: The relevant information was extracted using a structured data abstraction form in hospital records, operative notes, neonatal, intensive care records and follow up charts.

The variables obtained were:

1. Demographic: male to female, gestational age at birth, birth weight
2. Clinical presentation: age at admission, main symptoms,antenatal diagnosis, sepsis at admission
3. Operative: site & type of atresia, presence of associated anomalies
4. Surgical interventions: primary anastomosis, two-step procedure (stoma or Chimney procedure e.g. Santulli)
5. Postoperative outcomes: complications, NICU stay duration, TPN availability, survival at discharge, re hospitalisation post discharge, survival at minimum of 6 months follow up

Successful outcome was defined as survival of the baby at the time of discharge and on short term follow up of a minimum of 6 months.

Statistical Analysis: Data were entered into Excel and analyzed using SPSS and GraphPad Prism. Numerical variables were summarized using means and standard deviations, while categorical variables were described with counts and percentages. Two-sample t-tests were used to compare independent groups, while paired t-tests accounted for correlations in paired data. Chi-square tests (including Fisher's exact test for small sample sizes) were used for categorical data comparisons. P-values ≤ 0.05 were considered statistically significant.

Results

Table 1: Demographic Characteristics and Survival Outcomes of Neonates with Operative JIA Diagnosis

Variable	Value
Total neonates operated	33
Diagnosis	Operative only
Overall survival at discharge	72%
Overall survival at 6 months	64%
Statistical methods used	Chi-square test, t-test, logistic regression
Sex distribution	
Male	19 (58%)
Female	14 (42%)
Male: Female ratio	1.36 : 1

Table 2: Distribution of Cases, Survival Rates, and Statistical Analysis by Birth Weight, Age of Presentation, Surgical Procedure, and Type of Atresia

		No. of cases (n=33).	Survival rate (%)	P Value
Birth weight category	VLBW (<1.5 kg).	21(64%).	52%	0.038
	LBW (1.5 kg-2.8 kg).	5(15%).	60%	
	NBW (>2.8kg).	7(21%).	100%	
Age of presentation	<2 days	5(15%)	100%	0.041
	2-4 days	4(12%).	75%	
	4-7 days	5(15%).	60%	
	>7 days	19 (58%).	53%	
Surgical procedure	Primary anastomosis	13(39%).	36%	0.0217
	Stoma	6(18%).	20%	
	Santulli procedure	14(42%).	94%	
Type of Atresia	Jejunal Atresia	13 (38%)	Not specified	0.031
	Ileal Atresia	19 (58%)	Not specified	
	Multiple Atresia	1 (3%)	0	

Table 3: Distribution of Postoperative Complications in Study Patients

Postoperative Complication	Number of Cases	Percentage (%)
Sepsis	9/34	28
Wound infection	4/33	12
Anastomotic leak	4/33	12
Functional obstruction	6/33	18

Table 4: Distribution Of Mean NICU stay and time to full enteral feeding:

Surgical procedure(days)	Mean NICU stay (days)	Time to full enteral feeding (days)
Primary anastomosis	28.5+-5.2.	16.5+-3.1
Santulli procedure	22.3+-4.1.	12.3+-2.5
Stoma formation	19.7+-3.8.	7.8+-1.9

Table 5: Independent Predictors of Mortality from Logistic Regression Analysis Mortality predictors

Predictor	Odds Ratio (OR)	p-value	Interpretation
Low birth weight (<1.5 kg)	2.31	0.026	Significant independent predictor
Delayed presentation (>7 days)	1.98	0.038	Significant independent predictor
Primary anastomosis vs Santulli procedure	3.12	0.012	Significant independent predictor

In our study, a total of 33 neonates were operated on, with diagnoses managed through operative intervention only. The overall survival rate at discharge was 72%, which decreased to 64% at six months follow-up. The sex distribution showed a predominance of males, with 19 (58%) male neonates and 14 (42%) females, resulting in a male-to-female ratio of 1.36:1.

In our study of 33 neonates, survival rates varied significantly across different birth weight categories, with very low birth weight (VLBW, <1.5 kg) infants showing a survival rate of 52%, low birth weight (LBW, 1.5–2.8 kg) infants 60%, and normal birth weight (NBW, >2.8 kg) infants having 100% survival (p=0.038). Age at presentation also influenced survival outcomes, where neonates presenting before 2 days of age had a 100% survival rate, compared to 75% for 2–4 days, 60% for 4–7 days, and 53% for those presenting after 7 days (p=0.041). Surgical procedure impacted survival significantly as well; primary anastomosis was associated with a 36% survival rate, stoma formation with 20%, and the Santulli procedure showed the highest survival rate at 94% (p=0.0217). Regarding the type of atresia, 38% had jejunal atresia, 58% had ileal atresia, and 3% had multiple atresias, though survival rates for these groups were not specified. The difference in survival rates between jejunal and ileal atresia was not statistically significant (p=0.031).

In our study, postoperative complications were observed with varying frequencies among the neonates. Sepsis was the most common complication, occurring in 28% (9 out of 34) of cases and was the leading cause of mortality. Wound infections and anastomotic leaks were each seen in 12% (4 out of 33) of patients. Functional obstruction was noted in 18% (6 out of 33) of cases, representing a significant postoperative challenge. In our study, the surgical procedure significantly influenced both the mean NICU stay and the time to achieve full enteral feeding. Neonates undergoing primary

anastomosis had the longest mean NICU stay of 28.5 ± 5.2 days and required an average of 16.5 ± 3.1 days to reach full enteral feeding. Those treated with the Santulli procedure had a shorter NICU stay of 22.3 ± 4.1 days and achieved full enteral feeding in 12.3 ± 2.5 days. Stoma formation was associated with the shortest NICU stay of 19.7 ± 3.8 days and the quickest time to full enteral feeding at 7.8 ± 1.9 days.

In our study, logistic regression analysis identified several significant independent predictors of mortality in neonates. Low birth weight (<1.5 kg) was associated with a 2.31- fold increased odds of mortality (p=0.026). Delayed presentation beyond 7 days also significantly increased mortality risk with an odds ratio of 1.98 (p=0.038). Additionally, undergoing primary anastomosis compared to the Santulli procedure was linked to a higher mortality risk, with an odds ratio of 3.12 (p=0.012).

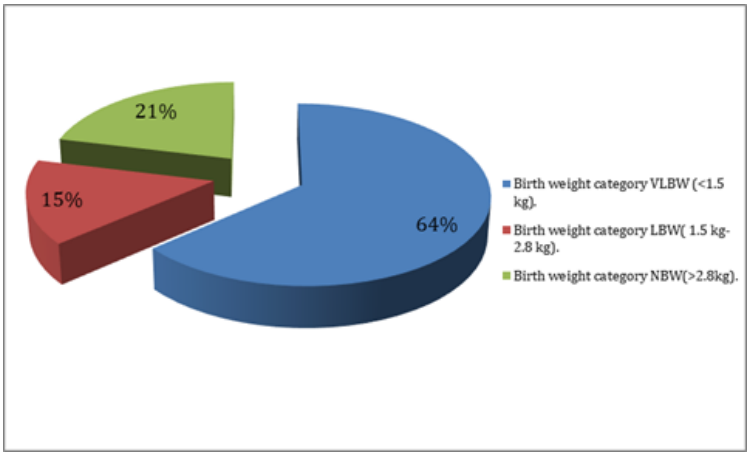

Figure 1: Distribution of Birth weight category

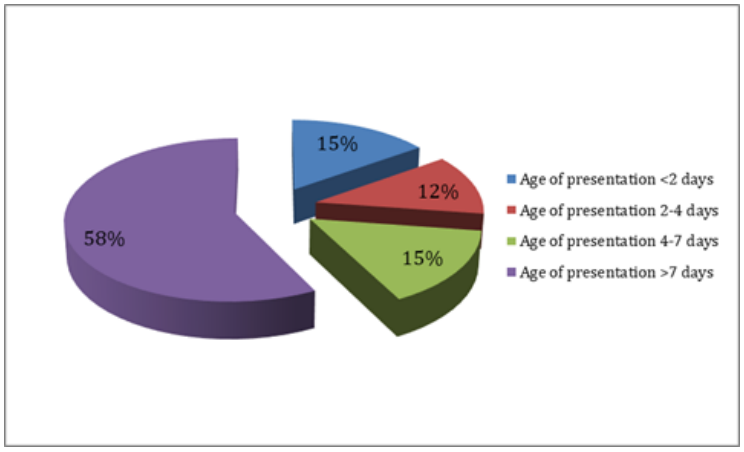

Figure 2: Distribution of Age of presentation

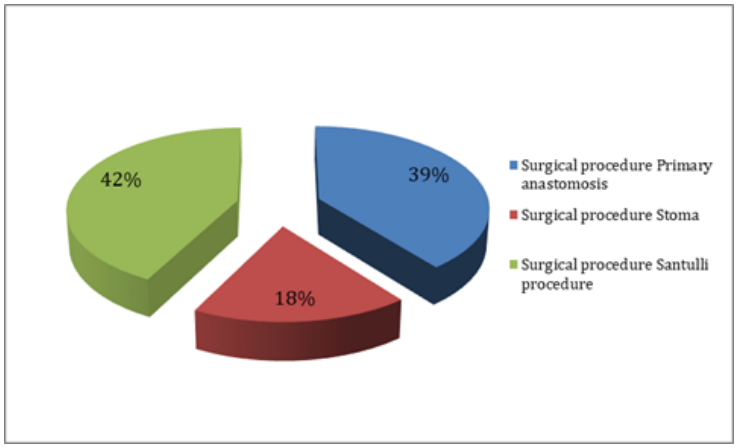

Figure 3: Distribution of Surgical procedure

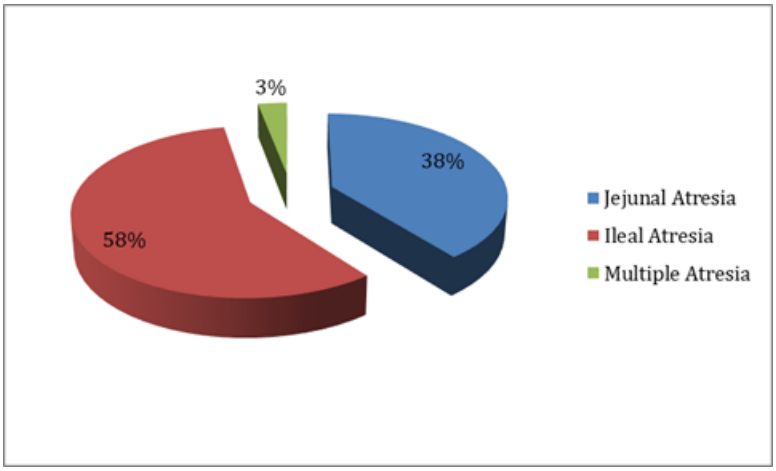

Figure 4: Distribution of Type of Atresia

Surgery trends: During the first 34 months of the 5 year study period, in most cases either primary anastomosis or stoma procedure was done depending upon the overall condition of the neonates. During the final 26 months of the research, most of them underwent Santulli procedure. This change was associated with better results.

Discussion

JIA is among the most frequent causes of neonatal intestinal obstruction that constitutes a substantial percentage of neonatal surgical emergency and needs urgent identification and treatment to avoid morbidity and mortality. [12] JIA is geographically distributed with some incidences indicating that 1 in every 1500 live births has JIA whereas other studies indicate that 1 in 10,000 births have JIA. JIA was the 4 th most prevalent cause of neonatal intestinal obstruction in our centre following Hirschsprung disease, anorectal malformation and malrotation.

Although there have been advances in antenatal screening especially in developed countries, prenatal diagnosis of JIA is not optimal especially in resource-limited countries. In our institution, out of the 33 patients who were operated upon in the study period, no patient had an antenatal diagnosis.

Etiology: The main causes of JIA are intrauterine vascular insult resulting in an ischemic necrosis of a portion of the intestine, the resorption and atresia formation. [15] The theory of this vascular accident was originally described by Louw and Barnard (1955) which showed that reduced mesenteric blood flow in the fetal life leads to ischemic injury and subsequent formation of atresia. [12] JIA may be an isolated condition or it may be accompanied by other birth defects including gastroschisis, malrotation. In our study, 5 neonates were reported to have associated anomalies.

Classification

JIA is classified based on the extent of luminal disruption and mesenteric defect:

Type 1: mucosal web obstructs the lumen, bowel remains continuous

Type 2: fibrous cord connects two blind ended bowel segments.

Type 3a: a complete discontinuity of the bowel

Type 3b: apple peel atresia: extensive atresia with a spiralized remnant of the distal nowel around the mesenteric artery

Type 4: multiple atresias occurring along the length of the small intestine.

The classification helps in surgical decision-making. More severe atresia types 3b & 4 are associated with a higher risk. [14]

Clinical presentation: In the majority of newborns with JIA, obstructive symptoms are shown during the first 24-48 hours of the neonatal age. Most frequent manifestations can be characterized by: vomiting with bilious content (classic manifestation of intestinal obstruction), no production of meconium, distension of the abdomen (more in case of distal atresias). [13] In approximately 25-50 percent cases, maternal polyhydramnios is identified as a result of the inability of the fetus to absorb amniotic fluid that he/she swallowed through the intestines. [14] Neonates who pass non-meconium small amounts of mucus can in some instances be misinterpreted to have passed meconium. [13]

Diagnosis: The preoperative suspicion of JIA is based on clinical evaluation and a straight X-ray of the abdomen showing multiple air fluid levels. The final diagnosis of JIA was always intra operative in our study.

Management

Surgery is the mainstay of JIA. The possible options are:

1. Primary anastomosis: is the gold standard, and is the preferred technique in uncomplicated cases

2. Tapering enteroplasty and anastomosis: applied in the cases of considerable proximal vowel dilatation.
3. Stoma procedure: indicated in neonates, who are critically ill or unstable; gut is gangrenous and abdomen contaminated).
4. Chimney procedure (Santulli or Bishop-Koop): goes together with a temporary enterostomy and an anastomosis as a means of early feeding and of guarding the anastomosis where the overall condition of the neonate is not good, there is great luminal discrepancy, and the presentation is delayed with the neonate having long parenteral fluids.

Prognosis and survival outcomes: JIA continues to be an important cause of morbidity and mortality in the newborn. The key to better outcomes is early detection, surgical treatment and good after-care, including nutrition. Prenatal detection enhancement as well as optimization of neonatal intensive care facilities are important in reducing mortality and long run outcomes. [14]

Trends in the survival between the developed and developing countries vary greatly. The survival rates of JIA have been in excess of 90 percent among developed nations as a result of prenatal diagnosis as well as due to the development of specialised neonatal surgical procedures, enhanced our abilities in neonatal intensive care and the provision of the total parenteral nutrition.[15] In resource-poor conditions, however, the death rates are high beyond 30-40% because of delayed diagnosis/presentation, missing neonatal surgical skills, poor neonatal intensive care services and scarcity of TPN. [3]

Various important findings were obtained during the retrospective review that can be added to the literature on the topic:

Consequences of delayed presentation: The average age of presentation was 9.5 days + 3 days implying a late diagnosis in the majority of the cases. Only 15 percent of them presented in the first two days of life and 58 percent presented after 7 days. Later presentation was also correlated to poorer survival (p=0.041, statistically significant), making the early detection and referral even more important. It was likely that the morbidity and mortality was higher as a result of prolonged starvation and metabolic derangements and more susceptible to sepsis. [14] As in a retrospective study in Nepal, the late presentation in most of the cases was associated with incorrect interpretation of mucus passage after birth as meconium by parents and primary care givers. [15,17] Also, many of the deliveries are still performed at home or in inaccessible places which led to delay in diagnosis and subsequently delay in referral.

Effect of birth weight: The study involved 79 percent LBW and 64 percent VLBW. There was a high degree of correlation between birth weight and mortality (p=0.038, statistically significant). This observation was corroborated by the other research studies which also documented low birth weight as a major risk factor of high mortality among neonates with JIA as they have a poor physiological reserve and increased susceptibility to infections. [14]

Surgery and surgical outcomes: The type of surgery that was determined by the overall patient condition also played a prominent role. A chi-square test ($x^2 = 7.66$, $p = 0.0217$) showed that there was a statistically significant difference in survival between surgical procedures and the Santulli procedure showed the highest survival (94%) compared to the primary anastomosis procedure and stoma procedure.

This confirms the results of other studies that have already indicated better results with a Chimney procedure that provides the advantage of both anastomosis and diversion stoma especially in neonates having unstable conditions or in those who are malnourished in a resource constrained environment [16,17] In developed countries or where the conditions are optimal and resources are sufficient, primary anastomosis will continue to be the gold standard and the most definitive surgical procedure.

As other factors like low birth weight and delayed presentation are not always in our control, a shift to Santulli as a primary procedure in majority of cases in the last 26 months have resulted in survival of more than 80% in the last 2 years suggesting that tailored surgical approaches based on patient condition lead to better outcomes.

Improvement of outcomes should be done using the following strategies:

1. Antenatal screening: antenatal anomaly scans may enable early detection of intestinal atresia permitting planned delivery in centres capable of performing neonatal surgery.
2. Early postnatal diagnosis: it is the responsibility of healthcare providers to have a high index of suspicion of intestinal obstruction in neonates with bilious vomiting, irrespective of the presence of abdominal distension which may preclude delay in diagnosis and treatment.
3. Timely referral and intervention: early referral to tertiary care centres and early surgical intervention is important particularly in low birth weight neonates.
4. Customized surgical technique: it is important to adopt a chimney technique (e.g Santulli) in neonates with unstable or malnutrition conditions since it is linked to higher survival outcomes.
5. Improved neonatal care: the investment in the NICU facilities, the availability of TPN and

specialised neonatal care can greatly minimize postoperative mortality and morbidity.

Conclusions

Jejunoileal atresia is a serious problem in the field of neonatology especially in resource limited countries. In this context, delayed presentation and low birth weight are shown in our study to be related to poor prognosis of JIA. Of the surgical procedures Santulli procedure proved to be better than primary anastomosis in our settings hence the need to adopt a case by case surgical approach depending on the condition of the patient.

The priority in future should be to strengthen antenatal diagnosis and increase the availability of specialised neonatal care to reduce the survival disparity between developing and developed countries.

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