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Jejunoileal atresia: a case-series of 63 neonates and risk factors to mortality

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Abstract

Introduction: Jejunoileal atresia (JIA) among neonates is still a condition which has huge morbidity and mortality, particularly in the developing world. We share a case series of JIA in context of their presentation, management, and outcome.

Materials and methods: This study was conducted at Children's Hospital and Institute of Child Health, Lahore, over 1 year. We included all patients presenting with JIA, and their demographic details, presentation, investigations, treatment strategies, and the outcome were noted at a pre-designed proforma. All data were analyzed using SPSS version 26.

Results: A total of 63 neonates with JIA were included. Most of them (79.4%) presented after 48 h of life, and the mean age at presentation was 5.68 ± 4.75 days. There were 37 male patients (58.7%), and 51 (81%) were full-term. The most common presenting complaint was not being able to pass meconium (88.9%). Type III atresia was the most common subtype (41.3%). Most of them underwent resection without tapering. The mean hospital stay was 12.81 ± 6.53, and it was significantly longer among those who underwent re-exploration ($P = 0.034$). Twenty-three patients (36.5%) expired within 6 months of follow-up. The only significant factor for mortality was the presence of short bowel syndrome ($P = 0.030$). All other demographic and management factors did not alter the mortality rate.

Conclusion: Management of surgical neonates is a difficult job in developing countries with limited resources. There is a high mortality rate of neonates following JIA surgeries, and surgeons in these countries must fight on many fronts to improve the outcome.

Keywords: Neonates, Intestinal, Jejunal atresia, Ileal atresia, Pakistan

Introduction

Gastrointestinal (GI) atresia is a commonly encountered entity in the neonates and a major cause for emergency surgery among them. Among GI atresia, the most common is the duodenal atresia, followed by jejunoileal atresia (JIA) and esophageal atresia. JIA is a pertinent condition in neonates with relatively less associated congenital anomalies; however, there are much more varieties and much more surgical options in the case of JIA. It makes JIA a bit more complicated because of a variety of choice of surgical options [1–3]. Also, it is a challenging situation in developing and poor countries because of lack of resources, delayed presentation of neonates, and lack of nursery intensive care unit (NICU) facilities [4, 5]. We aimed to present our experience of neonates presenting with JIA and determine the prognostic factors leading to mortality in our setup.

Materials and methods

This study was conducted at The Children's Hospital and The Institute of Child Health (CHICH), Lahore, after approval of the Ethical review board. CHCIH is the largest center for children in the country and the main referral center from all over the country. The duration of the study was 1 year (January 2020 to December

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2020). All the neonates who underwent surgery for JIA were included in the study. JIA was labeled as per-operative findings narrated by the surgeon. We did not include the other causes of small bowel obstruction like meconium ileus, volvulus without JIA, or total colonic Hirschsprung's disease. Also, those who expired before any intervention were not included in the study. Preterm was defined as gestational age < 37 weeks, and low-birth-weight was labeled if weight was < 2500 g. Mortality was defined as demise within 30 days after the surgery. All the patients were initially resuscitated, and after optimization, surgery was planned. It is a children's hospital and does not have any maternity unit, so all the cases were referred from other centers. We retrospectively went through the record of the operating room and collected all the cases of JIA who had been operated during this period. The charts of these patients were retrieved and all details were collected. The demographic details, presentation, investigations, treatment strategies, and outcome were noted at a pre-designed proforma. All data were analyzed using SPSS version 26. We calculated percentages for categorical variables and mean (SD) continuous variables. The odd's ratio (OR) was calculated for dependent variable (mortality) using binary logistic regression analysis.

Results

A total of 63 neonates presented with JIA in the study duration. Thirteen neonates (20.6%) presented in the first 48 h of life, while 50 neonates (79.4%) presented later. The mean age at presentation was 5.68 ± 4.75 days, and the age range of presentation was 1–25 days of life. Most of them (37; 58.7%) were males. Fifty-one neonates (81%) were full-term, and 37 (58.7%) were born through the vaginal route. Thirteen neonates (20.6%) had low birth weight (< 2500 g). In two patients (3.2%), JIA had been diagnosed antenatally.

Regarding the presentation, the most common presenting complaint was not being able to pass meconium (88.9%), followed by abdominal distention (76.2%) and bilious vomiting (68.3%). On per rectal examination, most of them (60; 95.2%) passed only mucous. Further, contrast enema identified microcolon in 28 patients (44.4%), and in remaining neonates, it was not significant.

All these neonates underwent exploratory laparotomy. The most commonly found type of JIA was type III atresia (26; 41.3%), followed by type 1, type II, and type IV. Among four patients, the volvulus of the intestine was also present along with JIA. The most commonly opted procedure was anastomosis of both portions of the gut without tapering (23; 36.5%), followed by tapering enteroplasty and anastomosis of the gut (15; 23.8%), stoma formation (15; 23.8%), and chimney creation procedures (10; 15.9%). Per-operatively, 15 patients (23.8%) were found to have short bowel, and they were specifically advised to start total parenteral nutrition (TPN) (Table 1).

The mean hospital stay was 12.81 ± 6.53 days, with the range being 5–35 days. Seven patients (11.1%) needed a re-exploration due to any reason. Regarding the mortality within 30 days, 23 patients (36.5%) expired, and 1 patient (1.6%) is still a failure to thrive (Table 1).

Table 1 Demographic details, presentation, management, and outcome of neonates in this study

Age at presentation	
Within 48 h	13 (20.6%)
After 48 h	50 (79.4%)
Gender	
Male	37 (58.7%)
Female	26 (41.3%)
Gestational age	
Pre-term	13 (20.6%)
Full-term	50 (79.4%)
Mode of delivery	
Vaginal route	37 (58.7%)
Cesarean section	26 (41.3%)
Weight	
< 2500 g	13 (20.6%)
≥ 2500 g	50 (79.4%)
Antenatal scan	
Yes	2 (3.2%)
No	61 (96.8%)
Presentation	
Bilious vomiting	43 (68.3%)
Abdominal distension	48 (76.2%)
Inability to pass meconium	56 (88.9%)
Surgical procedure opted	
ETEA without resection	23 (36.5%)
Tapering enteroplasty with ETEA	15 (23.8%)
Ileostomy	15 (23.8%)
Bishop-Koop procedure	7 (11.1%)
Santulli procedure	3 (4.8%)
Type of atresia	
Type I	14 (22.2%)
Type II	12 (19%)
Type III	26 (41.3%)
Type IV	11 (17.5%)
Short bowel syndrome	
Yes	15 (23.8%)
No	48 (76.2%)
Outcome	
Expired	23 (36.5%)
Alive	40 (63.5%)

Also, we tried to look for the factors which may lead to mortality among these patients. The only significant factor was the presence of short bowel syndrome (OR 3.643, $P = 0.036$). Mortality was higher among patients where tapering enteroplasty or chimney procedure was opted, but it was not significant. The age of presentation later than 48 h of life, male gender, type of atresia, associated volvulus or hospital stay had no significant effect on mortality ($P = 0.870$). All these details are summarized in Table 2.

Table 2 Logistic regression for factors leading to mortality

Variables	Number of patients with mortality (%)	OR: (95% CI): *P* value
Age at presentation		
Within 48 h	5/13	Reference
After 48 h	18/50	1.111: (0.316–3.908): 0.870
Gender		
Male	11/37	0.494: (0.174–1.403): 0.185
Female	12/26	Reference
Gestational age		
Pre-term	5/13	0.788: (0.218–2.846): 0.716
Full term	15/50	Reference
Mode of delivery		
Vaginal route	13/37	
Cesarean section	10/26	0.867: (0.307–2.450): 0.787
Surgical procedure opted		
ETEA without resection	6/23	Reference
Ileostomy	6/15	0.971: (0.222–4.243): 0.968
Bishop-Koop procedure	4/7	1.833: (0.392–8.566): 0.441
Santulli procedure	3/3	3.667: (0.557–24.132): 0.177
Type of atresia		
Type I	3/14	Reference
Type II	5/12	0.227: (0.040–1.299): 0.096
Type III	9/26	0.595: (0.114–3.102): 0.538
Type IV	6/11	0.441: (0.105–1.854): 0.264
Short bowel syndrome		
Yes	9/15	3.643: (1.091–12.168): 0.036
No	14/48	Reference
Volvulus		
Yes	1/4	0.561: (0.055–5.727): 0.625
No	22/59	Reference
Re-exploration		
Yes	3/7	1.350: (0.274–6.644): 0.712
No	20/26	Reference
Hospital stay		1.058: (0.977–1.146): 0.163

Discussion

In developing countries, delayed presentation and mortality are quite common in surgical neonates. In this series, 79.4% of the neonates presented after 48 h of life. Authors have reported similar conditions from Nigeria, who reported delayed presentation in 63.2% of cases [6]. Another study from Ethiopia reported 72% of cases having delayed presentation with GI atresia [7]. It is a different situation in developed countries, where authors report an early presentation [8, 9]. There are many reasons for this delayed presentation. First and foremost are the healthcare facilities for mother and child in developing countries. Most of the children are born in the peripheral centers without optimal neonatal care, and once they are referred to us, it is already more than 48 h passed [6, 7].

Gender distribution was almost equal in both genders in this study. No particular gender propensity has been noted in JIA in previously reported reports from Nigeria and Spain [6, 8].

An antenatal scan was done in only two patients (3.2%) in this series, and only their parents knew about the condition prenatally. Similar proportions have been narrated from other developing countries. However, in developed countries, it has been reported in up to 86.6% of cases [8]. Virgon C et al. conducted a meta-analysis over the data and reported the accuracy of prenatal ultrasound for the diagnosis of JIA varying 10–100%, with an overall prediction being 50.6%. Also, they narrated that the accuracy for diagnosing jejunal atresia is higher than ileal atresia (66.3% vs. 25.9%) [10]. Although ultrasound machines are available everywhere in Pakistan, the proportion of these conditions being diagnosed is very low. A recent huge population-based study concluded that almost 50% of the women visit healthcare workers during pregnancy two or fewer times [4]. There may be various reasons for this problem. Most probably, there is a lack of trained radiologists, and at most of the centers, gynecologists or technicians are performing antenatal scans. These people are not trained to diagnose JIA and other GI anomalies [11]. Also, most centers have poor resolution probes and old machines, which probably cannot pick up these conditions [6].

We found that the most common type of JIA was type III, followed by type I, type II, and type IV. Most of the

previous reports have narrated similar findings [6, 12]. However, one center had reported type II being most common type [13]. Another study from Pakistan had also reported type III as the most common subtype [12]. This variability in the frequency among types of JIA highlights that globally, it varies from one area to another. It may be another important area of future studies to investigate this aspect and determine the reason for this variance.

An important aspect in the management of surgical neonates in developing countries is the high mortality rate. In this series, we found mortality in 36.5% of cases. When we analyzed the data of JIA cases and their mortality, we found it 34.2% [6], 25% [7], 33% [14], 37% [15] from developing countries. However, in developed countries, it is clearly less than 10% [8]. The obvious reason for high mortality is increased complication rate, higher wound infection rate, sepsis, delayed presentation of the neonates, lack of NICU services and TPN, and hypothermia. Another report from Pakistan narrated the mortality in 42.5% of cases with GI atresia [12]. Chaudhry et al. reported complications in 40.8% of all children presenting with intestinal obstruction. These higher numbers show the conditions of the facilities and the level of care being provided [5]. The outcome can be improved with the provision of peri-operative anesthesia services, good post-operative care, trained NICU staff, good nutritional service, and a team-based approach to these cases [16].

The only variable which had significant effect on mortality was the presence of short bowel syndrome. The mean hospital stay in patients with short bowel syndrome was 16.6 + 7.67 days and 11.63 + 5.71 days (P 0.009) in those without SBS. All of the patients with SBS were started with TPN within the hospital. As we do not have the facility for home TPN, so usually, patients are discharged from the hospital, once they are tolerating oral feed and are stable. As mentioned, that in our setup, it makes a lot of sense because of poor provision of resources and no facility to provide home-based TPN. Because of these scarce resources, almost all such patients succumb to death and have no good outcome. Another interesting finding was that those who underwent some chimney procedure (like Bishop-koop or Santulli), had higher mortality. Most of the literature shows a better outcome with chimney formation procedures [17, 18]. We went through the charts of these patients to find out the reason, but could not find any pertinent reason for this phenomenon in our patients. However, we noticed that most of these procedures were opted when level of atresia was too proximal in the jejunum. We do not understand the reason for this observation; nonetheless, we speculate that it may be because of complications of a stoma, which may be too proximal in the jejunum. Another similar finding has been shared recently where authors found higher short-term and long-term complications among those who underwent stoma than those who underwent anastomosis [19].

Although a major limitation of this case series was being a single-center report, it highlights the situation and difficulties we observe in managing JIA in our setup. It also highlights a high mortality rate. We need to focus on the peri-operative care and provision of nutritional services to these neonates to improve the outcome.

Abbreviations

JIA: Jejunoileal atresia; GI: Gastrointestinal; NICU: Nursery intensive care unit; CHICH: The Children's Hospital and The Institute of Child Health; OR: Odd's ratio.

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Authors' contributions

Muhammad Saleem and Naeem Liaqat: conceptualization, methodology, software. Jamal Butt, Imran Hashim, Asim Raza, and Atika Urooj: data curation, writing—original draft preparation. Asif Iqbal: visualization, investigation, writing manuscript, proofreading. Muhammad Saleem: supervision. Naeem Liaqat and Asif Iqbal: software, validation: Naeem Liaqat, Imran Hashim, Asim Raza, and Atika Urooj: writing—reviewing and editing. All authors read and approved the manuscript.

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Declarations

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Yes, obtained.

Consent for publication

We give consent.

Competing interests

All authors declare that they have no competing interests.

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