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Evaluation of prenatal-postnatal outcomes and risk factors in fetal jejunoileal atresia

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ABSTRACT

Aims: The aim of the current study is to reveal ultrasonographic and clinical features, evaluation of prenatal-postnatal outcomes and risk factors of fetal jejunoileal atresia.

Methods: This retrospective study evaluated all cases of fetal jejunoileal atresia identified between 2018 and 2024 at a tertiary centre of maternal-fetal medicine. In all cases, the pediatric surgery team confirmed the definitive diagnosis of jejunoileal atresia during the postnatal period. The cohort was divided into two groups, the "poor outcome" and "good outcome", in order to compare and evaluate risk factors determining the outcome.

Results: During the study period, a total of 18 cases were recorded. All cases in the cohort managed to reach live birth. 15 (83.3%) neonates survived after the surgical procedure, whereas 3 (16.7%) neonates were deceased during the post-operative period. Short bowel syndrome was diagnosed in 2 (11.1%) living cases during postoperative follow-ups. ≤80 cm intact bowel length predicts poor outcome with 80% sensitivity and 92.3% specificity (p<0,015). The distance of the most proximal point of atresia to Treitz ≤40 cm predicts poor outcome with 80% sensitivity and 69.2% specificity (p<0,025).

Conclusion: Fetal jejunoileal atresia still has high mortality and morbidity rates despite improved technology, surgical techniques and advanced postoperative care. Therefore, it is very important that the delivery and particularly surgical procedure should be performed by an experienced surgical team in well-equipped centers.

Keywords: Fetal jejunoileal atresia, prenatal diagnosis, outcome, risk factors

INTRODUCTION

Jejunoileal atresia is an extremely rare condition, with a prevalance of between 0.3 and 1.1 per 10.000 live births in Europe.[1] A significant portion of affected infants are born prematurely.[2,3] Commonly associated anomalies include cardiac and abdominal wall defects, and cystic fibrosis. The initial surgical intervention typically involves either primary anastomosis (with or without resection) or the creation of an ostomy. The outcomes for children with this condition can vary; some may thrive, while others may experience significant complications, including difficulties with feeding, short bowel syndrome, or liver failure.[4,5]

The prenatal identification of fetal gastrointestinal disorders plays a crucial role in the management following birth. By facilitating a pre-arranged approach to these conditions, it is possible to recommend delivery at a facility equipped with a pediatric surgical team at the appropriate gestational age, thereby reducing the long-term outcomes of the anomaly.[6]

Inaccurate prenatal counseling can adversely affect the psychological well-being of parents and diminish the overall quality of the pregnancy experience. The sensitivity of prenatal ultrasound in detecting small bowel atresia (SBA) has been noted to vary significantly, primarily influenced by the level of the obstruction. The diagnostic accuracy for jejunal and ileal atresia has been reported to range from 25% to 50%.[7]

The presence of bowel dilation and polyhydramnios during the third trimester of pregnancy are recognized as major findings of jejunal and ileal atresia; however, their sensitivity and specificity are limited. This is due to the fact that other conditions, including meconium ileus, colonic atresia, Hirschsprung's disease, and imperforate anus, may mimic comparable ultrasound characteristics.[8]

The aim of the current study is to reveal ultrasonographic and clinical features, evaluation of prenatal-postnatal outcomes and risk factors of fetal jejunoileal atresia.

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METHODS

The study was conducted with the permission of the Clinical Researches Ethics Committee of Zeynep Kamil Women's and Children's Diseases Training and Research Hospital (Date: 20.12.2023, Decision No: 170). All procedures were carried out in accordance with the ethical rules and the principles of the Declaration of Helsinki.

This retrospective study evaluated all cases of fetal jejunoileal atresia identified between 2018 and 2024 at a tertiary centre of maternal-fetal medicine.

Patients with fetuses suspected of having jejunoileal atresia underwent weekly or biweekly ultrasound evaluations to monitor several key parameters. These assessments included the amniotic fluid index, indicators of fetal growth restriction (FGR), the presence of hyperechogenic bowel, and examinations of the stomach, gallbladder, small intestines, colon, anal target sign, as well as the detection of pseudocysts or intra-abdominal calcifications. Additionally, evaluations were conducted to assess for fetal anemia, ascites, intestinal peristalsis, and any associated anomalies. The volume of amniotic fluid was assessed using the amniotic fluid index, with values below 50 mm classified as oligohydramnios and those exceeding 250 mm categorized as polyhydramnios. A dilated intestinal loop is characterized as a hypoechoic, fluid-filled segment of the intestine that measures greater than 7 mm in diameter at its widest point. FGR was identified when the abdominal circumference was below the 3rd percentile in biometric assessments. Intestinal structures that appeared isoechoic in relation to adjacent bony elements were classified as hyperechogenic bowel. A lack of intestinal peristalsis observed for a minimum of 15 minutes during the ultrasonographic evaluation was characterized as decreased intestinal peristalsis. The diagnosis of fetal anemia was made when the peak systolic velocity of the middle cerebral artery exceeded 1.5 multiples of the median (MoM).

Genetic counseling was provided to all patients, and fetal karyotyping was offered. For those patients who did not have karyotyping conducted during the prenatal period, both karyotyping and cystic fibrosis mutation analysis were carried out after birth.

In each case, the pediatric surgery team confirmed the definitive diagnosis of jejunoileal atresia during the postnatal period. Details about the intraoperative findings were gathered from the surgical records and the surgical team.

The latest information regarding the cases was obtained through assessments carried out by specialists in paediatrics and paediatric gastroenterology, in addition to direct discussions with the parents concerned.

The cohort was divided into two separate groups: "poor outcome" and the other as "good outcome." This classification was intended to enable a comparison of outcomes and to evaluate the factors affecting these results. The two groups were compared in terms of demographic data, clinical characteristics, ultrasonographic findings, and postnatal outcomes.

All examinations were performed by experienced maternal-fetal medicine specialists utilizing a 5 MHz convex abdominal transducer (VOLUSON E6, GE).

Descriptive data are presented as median/25-75% interquartile range or numbers and %.

Statistical Analysis

Statistical analyses were performed using the Mann-Whitney U test for continuous data and the Fisher's exact test for proportions. To assess the performance of distance of atresia to Treitz and remaining intact bowel length in predicting poor and good outcome, receiver operating characteristic (ROC) analysis was performed. In all statistical analyzes the significance level (p-value) was determined at 0.05. The study data were analyzed using IBM SPSS statistics version 22.0 (IBM Corporation, Armonk, New York, United States) and MedCalc statistical software version 19.2.

RESULTS

From 2018 to 2024, our perinatology center monitored a total of 18 cases of fetal jejunoileal atresia, all of which were confirmed through postnatal pediatric surgical intervention. The median age of the cohort was 28 years old and the median gestational age for the first admission was 29.5 weeks. The most common reason for referral to our centre was dilated intestinal loops in 13 cases (72%). Hyperechogenic bowel in 2 cases (11%), FGR in 1 case (5.6%), fetal hydrops in 1 case (5.6%) and multiple structural anomaly in 1 case (5.6%) were the other reasons respectively. Clinical characteristics and prenatal-postnatal outcomes of the cohort were summerized in **Table 1**.

Throughout the prenatal monitoring, 4 cases showed no indications of intestinal obstruction. In one of these cases, there were no ultrasonographic findings at all. In the remaining three cases, the only ultrasonographic observation was a hyperechogenic bowel. Jejunoileal atresia was suspected in 14 cases based on ultrasonographic evaluations. Among these 14 cases, the median gestational age at which the diagnosis was suspected was 33 weeks, with the earliest gestational age of suspected jejunoileal atresia being 26 weeks (**Table 1**).

All patients except one received genetic counseling and were offered genetic diagnostic tests but prenatal genetic analysis was performed in only 2 patients via amniocentesis. Trisomy 18 was identified in one of these cases with jejunal atresia in addition to hypoplastic left heart syndrome and omphalocele. No karyotype abnormalities were detected in the other case that underwent amniocentesis and in the remaining 16 patients underwent postnatal genetic analysis (**Table 1**).

Ultrasonographic findings suggestive of fetal jejunoileal atresia were summarized in **Table 1**. Associated structural anomalies (hypoplastic left heart syndrome and omphalocele) were detected in 2 cases: hypoplastic left heart syndrome and omphalocele in 1 case and left isomerism in the other case. In 1 case (5.6%), FGR was detected before 32 weeks of gestation which were considered as early-onset FGR with umbilical artery end-diastolic flow loss.

Table 1. Demographic, clinical and ultrasonographic characteristics and prenatal-postnatal outcomes of the cohort (n=18)

Age, years (median/range)	28 (19-38)
Parity (median/range)	2 (1-4)
Consanguineous marriage (n,%)	5 (27.8)
GA at prenatal diagnosis, weeks (median/range)	33 (26-37)
Karyotype result	
Normal (n,%)	17 (94.4)
Trisomy 18 (n,%)	1 (5.6)
Ultrasonographic features	
FGR (n,%)	5 (27.8)
Polyhydramnios (n,%)	6 (33.3)
Oligohydramnios (n,%)	1 (5.6)
Dilated stomach (n,%)	3 (16.7)
Hyperechogenic intestines (n,%)	5 (27.8)
Dilated intestinal loops (n,%)	14 (77.8)
Ascites (n,%)	1 (5.6)
Decreased intestinal peristalsis (n,%)	4 (22.2)
Meconium pseudocyst (n,%)	1 (5.6)
Associated anomaly (n,%)	2 (11.1)
Prenatal diagnosis	
Jejunoileal atresia (n,%)	14 (77.8)
No diagnosis (n,%)	4 (22.2)
Mode of delivery	
Vaginal (n,%)	4 (22.2)
Cesarean (n,%)	14 (77.8)
GA at delivery, weeks (median/range)	37 (30-40)
Birthweight, gram (mean±SD)	2381±879
Postnatal surgery	18 (100)
Delivery to surgery, days (median/range)	2 (1-73)
Long term outcomes of live birth	
Neonatal death (n,%)	2 (11.1)
İnfant death 2-4 months (n,%)	1 (5.6)
Short gut syndrome (n,%)	2 (11.1)
Normal (n,%)	13 (72.2)
Cystic fibrosis (n,%)	2 (11.1)

Abbreviations: GA: Gestational age, FGR: Fetal growth restriction, SD: Standard deviation

All cases in the cohort managed to reach live birth. In the postnatal period, 8 of the neonates who were evaluated as intestinal obstruction were operated within the first 24 hours. Ten neonates were operated on after postnatal day 1, at the latest on day 73. Intraoperative features of the cases were summarized in **Table 2**. Accordingly, the median distance from Treitz ligament to the atresia was 45 cm (IQR 22.25-82.5) and the median length of intact bowel was 102.5 (IQR 78.25-122.5) cm in the cohort. During the postnatal period, cystic fibrosis was identified in two cases following the diagnosis of jejunoileal atresia. In six cases, thickened meconium was observed during surgical procedures; however, only one of these cases was subsequently confirmed to have cystic fibrosis after.

15 neonates survived after the surgical procedure, whereas 3 neonates were deceased during the post-operative period. Short bowel syndrome was diagnosed in 2 living cases during postoperative follow-ups. According to these findings, 5 cases including 3 deceased cases and 2 cases with short bowel syndrome were categorized as the "poor outcome" group, as 13 cases who had totally normal findings in postoperative period were categorized as the "good outcome" group to evaluate risk factors.

To evaluate prenatal and postnatal risk factors leading to poor outcome; demographic and clinical characteristics, ultrasonographic and surgical findings of both groups were analyzed. It was determined that there were no significant differences between two groups. On the other hand, receiver-operating characteristic (ROC) analysis was performed to determine a cut off value for the distance of the atresia to Treitz and intact bowel length (**Figure**). Accordingly, ≤80 cm intact bowel length predicts poor outcome with 80% sensitivity and 92.3 % specificity (p<0,015). The distance of the most proximal point of atresia to Treitz ≤40 cm predicts poor outcome with 80% sensitivity and 69.2% specificity (p<0,025).

DISCUSSION

Jejunoileal atresia frequently leads to neonatal intestinal obstruction, often resulting from intrauterine mesenteric vascular incidents. Recent developments in surgical methods, enhanced perinatal and postoperative care, along with the provision of artificial nutrition, have contributed to a decline in the overall mortality rate to 11-16% over the past few decades.[9] In more recent studies, mortality rates have been reported between 4% and 9%.[3,10,11] Mortality rate in our cohort was 16.6% as in line with the literature. Strategies aimed at

Table 2. Types of atresia, intraoperative findings, surgery type and complications

	Type 1	Type 2	Type 3a	Type 3b	Type 4
n, %	6 (33.3)	2 (11.1)	6 (33.3)	2 (11.1)	2 (11.1)
Segment with atresia					
Jejunal (n)	3	0	1	0	0
Ileal (n)	2	1	1	1	0
Jejunoileal (n)	1	1	4	1	2
Findings					
Malrotation (n)	4	0	1	2	1
Perforation (n)	1	0	2	0	0
Thick meconium (n)	2	1	1	1	1
Surgery type					
Resection+PA (n)	6	1 (ji)	6	2	1
Resection+stoma (n)	0	1 (i)	0	0	1
Complications					
Anastomosis leakage (n)	0	1	1	0	0
Adhesive obstruction (n)	2	0	0	1	0
Short bowel syndrome (n)	0	1 (ji)	0	0	1
Mortality (n)	1 (ji)	0	2 (1 j, 1 ji)	0	0

Abbreviations: i: ileal, j: jejunal, ji: jejunoileal

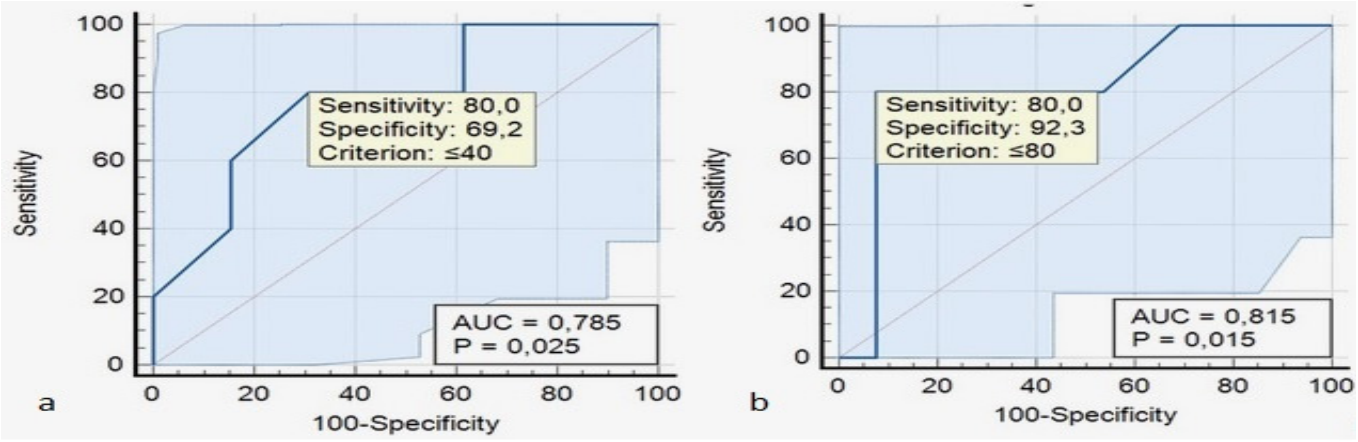

Figure. Receiver-operating characteristics analysis determined a cut-off for a) distance of the most proximal point of atresia to Treitz and b) remaining intact bowel

preserving bowel length and function have an impact on the length of time patients require total parenteral nutrition (TPN), with this duration being inversely related to the length of the remaining small bowel.[12] Extended reliance on artificial nutritional support increases the risk of complications, such as sepsis,[13] which was the cause of death in three cases within our cohort. It has been reported that associated anomalies can impact the outcome.[2] In our study, we observed that associated anomalies influenced two cases (11.1%): the first case involved hypoplastic left heart syndrome and omphalocele associated with trisomy 18, while the second case presented with left isomerism. As in the first case associated anomalies significantly contributed to the death of the newborn, in the second case did not affect the outcome. As previously reported in the literature, chromosomal abnormalities are observed at very low rates in jejunoileal atresia.[14] In our study, aneuploidy was detected in 1 case (5.5%) which is in line with the literature. Cystic fibrosis (CF) is estimated to impact between 5% and 24% of cases diagnosed with JIA.[3,15] In alignment with the findings of the current study, CF was identified in two of our cases, representing 11.1%, as determined through postnatal genetic analysis. JIA, particularly types IIIb and IV, along with associated complications such as volvulus, complicated meconium ileus, and meconium peritonitis, can lead to considerable intestinal loss, resulting in short bowel syndrome.[11] It is estimated that this occurs in 43% of cases.[16] In current study, during the long-term follow-up, two patients (11.1%) were identified as having short bowel syndrome and required nutritional support at home. One patient was born at term and underwent resection anastomosis due to type 2 atresia; however, a second surgical intervention was necessary 38 days later due to an anastomotic leakage, leading to another resection. The other patient was born at 35 weeks and received resection anastomosis for type 4 atresia, with a subsequent diagnosis of cystic fibrosis confirmed through genetic testing.

The prenatal ultrasound findings indicative of JIA including hyperechoic bowel, dilated fetal loops, and polyhydramnios, were commonly observed in our study. However, these findings did not influence the outcomes when compared between poor outcome and good outcome groups. There were also no differences in demographic and clinical characteristics, ultrasonographic and surgical findings between two groups.

CONCLUSION

In summary, it is well established that bowel loss and the resulting prolonged reliance on artificial nutrition are significant contributors to morbidity and mortality in patients with JIA, ultimately affecting their long-term prognosis and quality of life. Jejunoileal atresia still has high mortality and morbidity rates despite improved technology, surgical techniques and advanced postoperative care. Therefore, it is very important that the delivery and particularly surgical procedure should be performed by an experienced surgical team in well-equipped centers.

ETHICAL DECLARATIONS

Ethics Committee Approval

The study was conducted with the permission of the Clinical Researches Ethics Committee of Zeynep Kamil Women's and Children's Diseases Training and Research Hospital (Date: 20.12.2023, Decision No: 170).

Informed Consent

Because the study was designed retrospectively, no written informed consent form was obtained from patients.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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