RESEARCH ARTICLE

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Does Localization Have Clinical Significance in Small Bowel Atresia?

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Objective: Small bowel atresia is a common cause of intestinal obstruction in newborns. In this study, we aimed to compare the effects of localization on treatment, follow-up, mortality and morbidity.

Materials and Methods: 40 patients operated for small bowel atresia between 2011 and 2021 were analyzed. Data on gender, time of diagnosis, birth week, clinical features, accompanying anomalies, operative findings, hospitalization, complications, nutrition and development were compared.

Results: 21 duodenal, 7 proximal-jejunoileal and 12 distal-jejunoileal atresia were found. 28 patients were diagnosed prenatally. Duodenoduodenostomy was performed in 15 duodenal atresia, tapering+duodenoduodenostomy in 3 and duodenojejunostomy in 3 patients. Ostomy was performed in 1 proximal jejunoileal atresia and 6 distal jejunoileal atresia, all others were treated with anastomosis. In all groups, most common cause of mortality was sepsis and most common cause of additional surgery was obstruction. Time to full nutrition was 12(7-27) days in duodenal atresia, 56(10-123) days in proximal jejunoileal atresia and 29.5(9-673) days in distal jejunoileal atresia. The mean follow-up period was 1136.1 days. In the long-term there was developmental delay in jejunoileal atresia.

Conclusion: In proximal small bowel atresia, additional anomalies were more frequent, time to full nutrition and hospitalization was shorter; in distal atresia, time to total nutrition and hospitalization was longer, developmental delays were frequent. This maybe explained by involvement of a longer proximal bowel segment in distal atresia. Ileal atresia should be followed closely and appropriate projections should be made to families for the future according to atresia location

Key Words: *Small bowel atresia, Jejuno-ileal atresia, congenital anomaly, outcome*

İnce Barsak Atrezilerinde Lokalizasyonunun Klinik Önemi Var Mıdır?

Amaç: İnce barsak atrezisi, yenidoğan barsak obstrüksiyonunun sık sebeplerindendir. Çalışmamızda bu konjenital anomalinin lokalizasyonunun tedavi, takip, mortalite ve morbiditeye etkilerini karşılaştırmayı amaçladık.

Gereç ve Yöntem: 2011-2021 arasında duodenal, proksimal jejunoileal ve distal jejunoileal atrezi nedeniyle opere edilen 40 hastaya ait veriler incelendi, cinsiyet, tanı zamanı, doğum haftası, klinik özellikler, eşlik eden anomaliler, ameliyat bulguları, yatış süresi, komplikasyonlar, total beslenmeye geçiş süreleri ve büyüme gelişme ile ilgili veriler karşılaştırıldı.

Bulgular: Çalışmaya dahil edilen 40 hastanın 23'ü kız, 17'si erkekti. 21'inde duodenal, 7'sinde proksimal jejunoileal, 12'sinde ise distal jejunoileal atrezi mevcuttu. 28 hastaya prenatal tanı konulmuştu. Duodenal atrezi nedeniyle opere edilen 15 hastaya duodenoduodenostomi, 3 hastaya tapering+duodenodudenostomi, 3 hastaya duodenojejunostomi uygulandı. Proksimal jejunoileal atrezi nedeniyle opere edilen 1 ve distal jejunoileal atrezi nedeniyle opere edilen 6 hastaya ostomi açılırken diğer hastalar rezeksiyon+primer anastomoz ile tedavi edildi. Tüm gruplarda en sık mortalite sebebi sepsis, en sık ek cerrahi sebebi obstrüksiyondu. Full beslenmeye geçiş duodenal atrezilerde 12(7-27), proksimal jejunoileal atrezilerde 56(10-123), distal jejunoileal atrezilerde ise 29.5(9-673) gündü. Ortalama takip süresi 1136.1 gündü. Uzun dönem takiplerde duodenal atrezilerde boy, kilo persantili normal sınırlardayken özellikle distal jejunoileal atrezilerde gelişim geriliği mevcuttu.

Sonuç: Çalışmamızda proksimal ince barsak atrezilerinde full beslenmeye geçiş ve hastanede yatış süresi daha kısa, distal atrezilerde ise total beslenmeye geçiş süresi ve hastanede yatış süresi daha uzun, uzun dönemde gelişim geriliğinin daha sık olduğu görüldü. Bu durum distal atrezilerde daha uzun bir barsak segmentinin etkilenmesiyle açıklanabilir. Büyüme ve gelişme açısından ileal atreziler yakın takip edilmeli, atrezinin yerine göre aileler bilgilendirilmelidir

Anahtar Kelimeler: *İnce barsak atrezisi, jejuno-ileal atrezi, konjenital anomali*, uzun dönem sonuçlar

Introduction

Congenital small bowel atresia (duodenal, jejunal and ileal atresia) is a well-recognized, frequent cause of neonatal intestinal obstruction and can occur at any point in the gastrointestinal tract (1). Incidence of intestinal atresia ranges from 1.3 to 3.5/ 10,000 births (2, 3). Intestinal obstruction often manifests itself with different signs and symptoms including prenatal polyhydramnios, distended abdomen, bilious emesis, feeding intolerance and failure to pass meconium within first 24 hours of life (4). Resection of the effected segment and primary anastomosis with or without enteroplasty

is the typical surgical techniques of choice for proximal atresia. In distally located, more complex atresia, creating a decompressing stoma is preferable (5).

Over the last years, increased antenatal diagnosis, early interventions, availability of highly qualified intensive care units, parenteral nutritional support, preservation of intestinal length and improvement in management of associated anomalies have significantly improved the prognosis of intestinal atresia (6). Predicting the clinical outcome of infants with intestinal atresia may be difficult. Although short bowel length after atresia repair is a well-known cause for poor outcome, it is not clear whether the prognosis depends on the type, location or other associated anomalies (7).

In the current literature, there is no study establishing a connection between the site of intestinal atresia and its long-term impact on children's physical growth. In this study, we aimed to reveal this relationship and to compare the effects of atresia localization on treatment, follow-up, mortality and morbidity of the patients.

Materials and Methods

Research and Publication Ethics: The study was started after getting permission from the Tepecik Training and Research Hospital local ethics committee (No:2021/07-03).

All newborns admitted with small bowel atresia over a period of 10 years, from January 2011 to December 2021, were included in the study. Patients with concomitant abdominal wall defects were excluded from the study. Data were collected from patients' hospital records included antenatal history, patient demographics, gestational age, diagnosis, associated anomalies, operative findings, preferred surgical management and outcome. The patients were grouped according to location of the atresia (duodenal, proximal jejunoileal and distal jejunoileal) to analyze the relationship between management and outcome.

These patients were invited to participate in a telephone survey. Patients' body weight and heights were questioned. The records of the patients followed up in our center were obtained from the center records. Patients' body weight-for-age (BWA) and height-for-age (HFA) z-scores were calculated using the World Health Organization (WHO) growth curves. According to WHO definitions, a BMI z-score <-2 SS was considered short stature and a BMI Z-score <-2 SS was considered underweight (8). The sample size was calculated using the G*Power software (version 3.1.9.2). G*Power software estimated statistical power of 96.8% with a significance level of α=0.05 for a total of 27 respondents.

For discrete and continuous variables, descriptive statistics (mean, standard deviation, median, minimum, and maximum values) were calculated. In addition, the homogeneity of variances, which is one of the prerequisites of parametric tests, was checked using Levene's test. The assumption of normality was tested using the Shapiro-Wilk test. To compare the differences between the two groups, an independent sample t-test was used when the parametric test prerequisites were fulfilled, and the Mann-Whitney U test was used when such prerequisites were not fulfilled. The chi-squared test was used to determine the relationships between the two discrete variables. When the expected sources were less than 20%, values were determined using the Monte Carlo simulation method to include these sources in the analysis. Age was determined as covariates (to be excluded), and the groups were compared using covariance analysis. Kruskal-Wallis, Mann-Whitney U, One-way ANOVA tests were used where indicated and written throughout the text. Statistical analysis was performed using SPSS 25.0 (IBM Corp., Armonk, New York, USA).

P values less than 0.05 were considered statistically significant.

Results

The data of 40 patients operated for small bowel atresia were retrospectively analyzed. Demographic characteristics of the patients are given in Table 1. Of the 40 patients, 23 (57.5%) were girls and 17(42.5%) were boys. The mean birth weight was 2640 g [range: 1010-3970] and 17 patients (42%) were preterm.

Table 1. Patient demographics

	Duodenal N=21	**Proximal Jejunoileal N=7**	**Distal Jejunoileal N=12**	***P***
Prenatal diagnosis	14(66)	6(85)	8 (66)	0.126
Additional Congenital anomalies	16(76)	5 (71.4)	9 (75)	
Cardiac	15	4	7	0.835
GIS	2	1	2	
GUS	1	-	-	
Male/Female	12:9	1:6	4:8	
Preterm (<37w)	12(57)	1(15)	4 (34)	
Birth weight (gr)	2770±520	2150±860	2520±560	0.078
GIS Complication Surgery	2 (9)	5(71)	7 (58)	0.018
Mortality	2(14)	1 (14)	3 (16)	0.862
Follow-up (mo)	51±44	44±20	44±43	0.907

Values are expressed as mean±SD or number (percent)

Table 2. Short-term outcomes based on location of the atresia

Variable	Duodenal (N=21)	Proximal Jejunoileal (N=7)	Distal Jejunoileal (N=12)	*p*
Length of stay (d)	19.5(8-185)	83(15-156)	38(10-673)	0.001
Time to full enteral nutrition (d)	12 (7-37)	56(10-123)	29.5(9-673)	0.013

Data are medians (IQR).

Statistically significant comparing proximal jejunoileal to duodenal and proximal jejunoileal to distal jejunoileal atresia

Table 3. Long-term outcomes based on location of atresia

	Duodenal N=14	Proximal Jejunoileal N=5	Distal Jejunoileal N=8	*P*
Z-Score Height for age*	0.17±1.2	-0.75±0.52	-2.1±0.83	0.003
Z-Score Weight for age	0.28±1.35	-1.37±0.58	-2±1.02	0.059

Date are mean ± SD

*Statistically significant comparing distal jejunoileal to duodenal and distal jejunoileal to proximal jejunoileal

The atresia was located in duodenum in 21 (52.5%), proximal jejunoileal in 7(17.5%) and distal jejunoileal in 12(30%) patients. 28 patients (70%) were diagnosed by prenatal USG. The most prominent findings in prenatal USG were polyhydramnios 6(28%) and double-bubble appearance 9(32%) in duodenal atresia, increased echogenicity 4(57%) and polyhydramnios 3(42%) in proximal jejunoileal atresia and dilatation of intestinal anus 4(33%), presence of weak meconium signal 3(25%) in distal jejunoileal atresia.

The number of accompanying anomalies was higher in duodenal atresia (76%). Cardiac (ASD, VSD, PDO) and gastrointestinal anomalies (annular pancreas, malrotation, antenatal volvulus) were the most common accompanying anomalies in all groups. Duodenoduodenostomy was performed in 15 patients with duodenal atresia, tapering+duodenoduodenostomy in 3 patients and duodenojejunostomy in 3 patients. Temporary ostomy was performed in 1 patient operated for proximal jejunoileal atresia and 6 patients operated for distal jejunoileal atresia, while the other patients were treated with resection+primary anastomosis. The ostomies were closed after a mean of 54 days [range:20-96]. In all groups, the most common cause of mortality was sepsis and the most common cause of additional surgery was obstruction. Time to full nutrition was 15±9 days in duodenal atresia, 56±43 days in proximal jejunoileal atresia and 102±203 days in distal jejunoileal atresia (p=0.013). The mean follow-up period was 44±41 months (Table 2).

Data from twenty-seven of thirty-four living patients were evaluated for long term development. As the growth of the patients was evaluated, short stature or underweight was not found in any of the 14 patients with duodenal atresia. Only 1 (20%) of 5 patients with proximal jejunoileal atresia was underweight, while 6 (75%) of 8 patients with distal jejunal ileal atresia had short stature and 4 (50%) were underweight. When the Z-scores of BWA and HFA of the patients were compared, a statistically significant difference was found between groups (p<0.001 both for BWA and HFA) (Table 3). Figure 1 depicts number of patients with normal Z scores (between -2 and +2) in each group both for BWA and HFA.

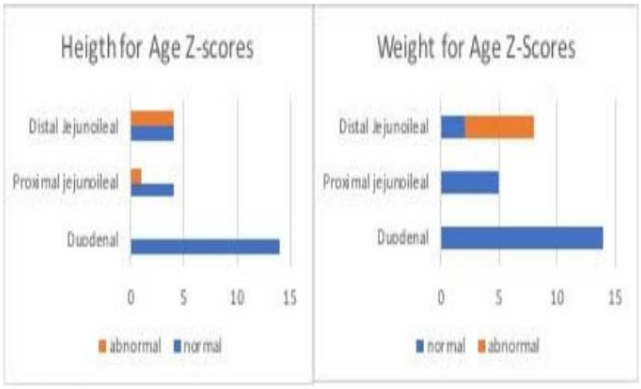

Figure 1. Height for age and Weight for age Z-scores

Discussion

Evaluation of intestinal atresia is usually initiated before birth. In our series the major diagnostic tool was antenatal ultrasound performed between 28-31 weeks of gestation, advanced imaging with antenatal MRI was also used in complex cases. Our accuracy of 70% in detecting small bowel atresia compares highly favorable with other similar series of 23-45% accuracy rate (7). Our hospital is a major antenatal diagnostic center with highly experienced perinatologists, this may contribute to our high diagnostic rates. Radiologic findings vary depending on the location of atresia. Diagnosis of duodenal atresia was mostly based on image of double bubble on ultrasound, in some cases polyhidramiosis was also noted. Jejunoileal atresia was diagnosed by dilated intestinal loops and polyhydramniosis, the number of dilated loops and echogenicity of the bowels increased as the atresia located more distally. In our series there was no correlation between polyhydramniosis and location of the atresia.

Furthermore, our rates of antenatal detection did not differ between groups which is different than the current literature stating high antenatal diagnostic rates for more proximal atresia (9). As accepted in literature, duodenal atresia is more commonly associated with comorbidities whereas jejunoileal atresia is more often an isolated finding (10). interestingly, in our series, there was no significant difference in additional congenital anomalies between groups. In all groups cardiovascular comorbidities were the most common, followed by gastrointestinal and genitourinary anomalies. Although cardiovascular comorbidity was common in all groups, only 2 patients (1 duodenal atresia and 1 distal jejunoileal atresia) had complex cardiac anomalies and both died due to cardiac reasons. Further research is needed to explain our higher incidence of associated anomalies in both proximal and distal jejuno-ileal atresia groups (71% vs 75%).

The surgical approach in intestinal atresia depends on the location of the lesion, the anatomy, intraoperative condition, and the remaining bowel length (4). In our center, surgical repairs were performed through a right upper quadrant transverse laparotomy in the post-natal period as soon as the patients were stable. Although surgeon's individual preference plays an important role in the selection of operative approach, for duodenal atresia, treatment involved diamond shaped duodenoduodenostomy or duodenojejunostomy and in certain cases with extensive dilatation a prior tapering was also included. For proximal and distal jejunoileal atresia first choice of operation was resection of atretic loop and primary anastomosis but in cases with significant luminal disparity Santulli or Mikulicz type enterostomies were also performed. In our cohort around 30% of the neonates required re-laparotomy due to surgical complications after initial surgery. This number is higher than similar series in literature (4, 7). Both distal and proximal jejunoileal atresia has higher complication surgery rates than duodenal atresia but there was no statistical difference in between proximal and distal jejunoileal atresia. In all groups, bowel obstructions due to post-operative adhesions were the major reason of re-operation followed by anastomotic dehiscence. In our series there was no correlation between type of surgery and post-operative complications.

The mortality associated with neonatal intestinal obstruction ranges between 15% to 45% depending on the developmental status of a nation (11). In this series, overall mortality was 15%. In a similar cohort, Tongsin et al. demonstrated greater intrauterine growth retardation in neonates with jejunal as compared to ileal atresia and linked this to increased mortality (12). In our series, we have not found any significant correlation between mortality and prematurity or birth weight. The location of atresia did not directly correlate with mortality rates, cause of mortality was mostly associated with additional congenital anomalies in duodenal atresia and sepsis due to anastomotic complications in distal jejunoileal atresia.

Our data indicates an average of 2 weeks for infants with duodenal atresia and 3 to 4 weeks for proximal and distal jejunoileal atresia for full enteral nutrition. There was no statistically significant difference between proximal and distal jejunoileal atresia, both needed significantly longer time to full enteral nutrition compared to duodenal atresia and we believe this was the main reason for longer length of hospital stay (LOS) in both groups. International studies show a median LOS between 16 and 32 days in intestinal atresia (13, 14) our median of 24 days is consistent with literature. In regard to location of the atresia, duodenal atresia patients had significantly shorter LOS when compared to both proximal jejunoileal and distal jejunoileal atresia. In a recent national cohort study Schmedding at al. found that patients who needed a stoma had significantly longer LOS (15) we also found that regardless of the place of atresia, presence of enterostomy significantly increases duration of hospitalization (median 28 vs 43 days. p=0.042).

Growth measurements are valued as the most important components of the nutritional assessment in children. A growth standard refers to a dataset and related growth charts that reflect a goal for the population. The WHO charts are considered a growth standard because they describe the growth of healthy children under optimal nutritional and environmental conditions (16). Z-scores, that are units of standard deviation (SD) from the population mean, are used as reference lines in growth charts. The normal range is generally defined as between -2 SD and +2 SD (Z-scores between -2.0 and +2.0). There are only a few data documenting the longer-term outcomes of intestinal atresia. Shibuya et al. (17) reports that in intestinal atresia, within two years of surgery accelerated growth to regain normal size occurs in children. In a similar cohort in 2018 Peng et al. (18) reported that intestinal atresia patients who recover from the initial operation and survive the short-term postoperative period, have a good chance of having normal growth and development. Burjonrappa et al. revealed that, due to greater adaptability and increased surface area for absorption of the ileum, jejunal atresia offers better outcome when compared to ileal atresia (19). While the effect of genetic make-up and environmental factors cannot be denied, we found out that despite similar gestational age, birth weight and additional comorbidities in the long-term there is significant developmental delay in distal jejunoileal atresia. Again, the numbers in this study are relatively small, but regardless the surgical approach; as the atresia located distally, although catch-up growth in weight is achieved in time, short stature remains constant.

The main limitation of this study is low sample size which could be explained by the rarity of the disease. Several factors, like specific operative details and other missing data points are subject to reporting and observer bias. Furthermore, it is a single-center study with retrospective data collection, some patient records were lost to follow up. Future multicentered studies with larger sample size investigating impact of location of intestinal atresia on long term results are advocated.

As a result, although initially there was no significant difference in prematurity, birth weight and addition congenital anomalies; in the long term distal jejunoileal atresia patients demonstrate signs of developmental delay. We believe distal atresia patients may benefit from more aggressive nutritional support in order to enhance their potential to achieve a normal growth. Multicentered studies with large numbers regarding long term effects of site of atresia are needed for providing reliable data for antenatal counselling.

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