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Documentation of small intestine atresias: a single-institution experience in Turkey (22 cases)

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Aim: To investigate the associated anomalies and causes of death in a neonatal autopsy series with intestinal atresias over an 8-year period from the archives of our department. Intestinal atresia, meaning the complete absence of a segment of the intestine or complete occlusion of the intestinal lumen, is the most common cause of gastrointestinal obstruction in neonatal autopsy series. There is an approximately equal sex distribution.

Materials and methods: From the archives of our records, 22 cases with intestinal atresia were reevaluated. The patients' demographic data were noted. Anomalies associated with intestinal atresia were grouped into systemic categories and the main causes of death were documented.

Results: There was a female predominancy with an average life span of 10 days. Neonates with prematurity or low birth weight made up the vast majority of the series (95.5%). Duodenal atresia was seen in 11 cases (50%). The anomaly most frequently associated with intestinal atresia was detected in the gastrointestinal system, as well (77.2%). The most common cause of death was bacterial sepsis, at 50%.

Conclusion: Intestinal atresias are one of the significant anomalies among perinatal autopsy cases and need to be carefully investigated by pathologists.

Key words: Intestinal atresia, autopsy

Türkiye'den tek merkeze ait ince barsak atrezilerinin dökümantasyonu (22 olgu)

Amaç: Barsağın bir segmentinin tamamen yokluğu veya barsak lümeninin tamamen tıkanması anlamına gelen intestinal atrezi, yenidoğan otopsi serilerinde gastrointestinal obstrüksiyonun en sık nedenidir. Her iki cinsiyette hemen hemen eşit oranda görülür. Bu çalışmada; amacımız departmanımızın sekiz yıllık arşivindeki intestinal atrezili neonatal otopsi serisinde eşlik eden anomaliler ve ölüm sebebini araştırmaktı.

Yöntem ve gereç: Arşivimizde kayıtlı 22 intestinal atrezili olgu yeniden değerlendirildi. Olguların verileri kaydedildi. İntestinal atrezi ile ilişkili anomaliler sistemik kategoriler içinde gruplandırılarak başlıca ölüm nedeni dökümante edildi.

Bulgular: Kız cinsiyet baskın olup ortalama yaşam süresi on gündü. Olguların büyük bir çoğunluğunu prematür ve düşük doğum ağırlıklılar oluşturmaktaydı (% 95,5). On bir olguda (% 50) duodenal atrezi vardı. En sık görülen eşlikçi anomali yine gastrointestinal sistemde idi (% 77,2). En sık ölüm nedeni bakteryel sepsisti (% 50).

Sonuç: Barsak atrezileri perinatal otopsilerde patologlar tarafından dikkatle araştırılması gereken önemli doğumsal anomalilerdendir.

Anahtar sözcükler: Barsak atrezileri, otopsi

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Introduction

Intestinal atresia (IA) is the most common cause of gastrointestinal (GI) obstruction in children. IA is the complete absence of a segment of the intestine or the complete occlusion of the intestinal lumen. There is an approximately equal sex distribution. The incidence rate is about 1 in 1000 live births. There is an almost equal ratio among atresias of the duodenum, jejunum, and ileum. Multiple jejunoileal atresias (multiple JIA) are found in approximately 10% of cases. Colonic atresia occurs less frequently. Due to the fact that the duodenum is the only segment of the small bowel that is obliterated at some stage of its normal development, it is appropriate to invoke the failure of recanalization of the lumen as one of the mechanisms for duodenal atresia (DA). Most multiple JIAs result from intrauterine vascular accidents with infarction and subsequent resorption of the necrotic segment; they are associated with vascular insults such as intrauterine malrotation, volvulus, intussusception, internal hernia, and constricting gastroschisis (1,2).

Congenital anomalies are seen with IA; in about 40%-50% of all cases of DA, there are associated anomalies. Extraintestinal anomalies are more commonly associated with DA than with the distal IAs. DA is associated with Down syndrome, annular pancreas, congenital heart disease, malrotation, and esophageal atresia (3).

Our aim was to investigate the associated anomalies in a series of 22 neonatal autopsy cases with IA.

Materials and methods

IA autopsy cases were retrieved from the archives of the autopsy section of the pathology laboratory of İzmir Tepecik Training and Research Hospital for an 8-year period (1998-2005). Gender, birth weight, gestational age, associated anomalies, performed surgeries, and life duration were noted for each case. Anomalies were grouped into categories as being associated with the gastrointestinal system (GIS), cardiovascular system (CVS), genitourinary system (GUS), central nervous system (CNS), respiratory system (RS), and others. The cause of death was also assessed.

Results

Out of 268 neonatal autopsy cases, 22 had IAs (8.2%). There were 9 males and 13 females. The mean birth weight was 2073 g (1300-4100 g) and the mean gestational age was 32.6 weeks (24-39 weeks). The prematurity rate was 81.8% (18 cases), whereas the rate of low birth weight was 13.6% (3 cases). One case showed normal gestational age and weight. The average life span was 10 days (1-60 days).

It was seen that 11 cases (50%) had DAs, 6 had JAs (27%), 3 (13.5%) had ileal atresias, and 2 had multiple JIAs (9%) (Table 1). Out of 22 patients, 14 received a corrective operation; the DA corrective operation was the most common in our series (50%), whereas 5 operations were for JAs and 2 were for ileal atresias. The most common GIS anomaly was esophageal atresia with or without tracheoesophageal fistula (EA ± TEF) (31.8%). The most frequent CVS anomaly was ventricular septal defect (VSD) (9%). There were 3 genitourinary anomalies, including renal aplasia, ureteral duplication, and uterus unicollis (13.6%). Meanwhile, 2 newborns had lung disorders such as a lobulation anomaly (3 lobes in the left lung) and right lung aplasia, 1 had a cleft lip and palate, and 1 had an extremity anomaly (Table 2). The systemic anomalies were seen mostly in DA patients, while 11 patients were associated with at least 1 GIS anomaly (64.7%). The most common anomaly was EA ± TEF. All of the CVS and RS anomalies were detected in DA patients. The GUS anomalies were most frequent in the DA group (66.7%), as well (Table 3).

Bacterial sepsis was the most common cause of death, at 50%; 9 of the newborns lost to bacterial sepsis (64.2%) were in the surgical intervention group (Table 4).

Table 1. Types of atresias (n: number of the cases).

Types of intestinal atresia	n
Duodenal atresia	11
Jejunal atresia	6
Ileal atresia	3
Jejunoileal atresia	2
Total	22

Table 2. Systemic findings from the cases with intestinal atresia. GIS: gastrointestinal system, CVS: cardiovascular system, GUS: genitourinary system, RS: respiratory system, EA ± TEF: esophageal atresia with or without tracheoesophageal fistula, n: number of cases.

Systemic anomalies	Type (n)	Number of cases
GIS	EA ± TEF (7) Congenital hypertrophic pyloric stenosis (2) Meckel's diverticulum (2) Midgut malrotation (2) Annular pancreas (1) Anorectal aplasia (1) Pancreatic heterotopia in stomach (1) Gastroschisis (1)	17
CVS	Ventricular septal defect (2) Atrial septal defect (1) Aortic arch anomaly (1) Single atrium (1)	5
GUS	Renal aplasia (1) Ureteral duplication (1) Uterus unicollis (1)	3
RS	Lobulation anomaly of the lung (1) Lung aplasia (1)	2
Others	Cleft lip and palate (1) Pes equinovarus (1)	2

Table 3. Distrubition of the systemic anomalies according to the atresia types. GIS: gastrointestinal system, CVS: cardiovascular system, GUS: genitourinary system, RS: respiratory system, n: the number of the cases, EA ± TEF: esophageal atresia with or without tracheoesophageal fistula, VSD: ventricular septal defect, ASD: atrial septal defect.

Type of atresia	GIS (n)	CVS (n)	GUS (n)	RS (n)	Other (n)
Duodenal atresia	EA ± TEF (4) Anorectal aplasia (1) Congenital hypertrophic pyloric stenosis (2)Annular pancreas (1) Meckel's diverticulum (1) Pancreatic heterotopia in stomach (1) Midgut malrotation (1)	VSD (2)ASD (1) Single atrium (1) Aortic arch anomaly (1)	Left renal aplasia (1) Uterus unicollis (1)	Lobulation anomaly of the lung (1) Lung aplasia (1)	
Ileal atresia	EA ± TEF (1) Midgut malrotation (1)				
Jejunal atresia	EA ± TEF (1) Meckel's diverticulum (1)		Ureteral duplication (1)		
Jejunoileal atresia	EA ± TEF (1) Gastroschisis (1)				Cleft lip and palate (1) Pes equinovarus (1)

Table 4. The main causes of death (RDS: respiratory distress syndrome).

Causes of death			
Sepsis	Bacterial	11	
	Mycotic	1	*Candida Actinomyces*
RDS		4	
Pneumonia	Nonspecific	1	
	Viral	1	
	Aspiration	1	
	Klebsiella	1	
Severe cardiac malformation	Aortic arch anomaly	1	
Total		22	

Discussion

IA is a well-defined cause of bowel obstruction in neonates. It accounts for 95% of GI obstructions. Although the management of newborns with IA has improved in terms of operative technique (4), it can be a causative factor in death for neonates because of associated anomalies, low birth weight, prematurity, and superinfections. The goal of the current study was to investigate the associated anomalies. We also examined the basic causes of death of the newborns.

Our autopsy series from an 8-year period showed a rate of 8.2% IA cases out of 268 neonates with a predominance in females (59%), compatible to the results of previous reports (4,5).

Many infants with DA are premature (35%) (2,6,7). The prematurity incidence in this study was 72% (16 cases). The low birth weight incidence is approximately 25%-50% (6). The low birth weight incidence was 81% (18 cases) in the current report.

There is an approximately equal occurrence of atresia in the duodenum, jejunum, and ileum (1,8). DA was the most common type of atresia in the current series (50%).

In about 40%-50% of all cases of DA, there is an associated congenital anomaly. These include mostly GI and CVS disorders, such as malrotation, EA ± TEF, biliary atresia, and different types of congenital heart diseases (9-16). Even though they are more rare than DAs, JAs and multiple JIAs can also present associated anomalies (2). Choudry et al. reported that from 57% of associated congenital anomalies with DA and a series of multiple JIAs, 52% of the cases had associated congenital anomalies (3,17). Out of 22 cases from our recorded neonates, 17 (77%) had gastrointestinal anomalies including esophageal atresia, congenital hypertrophic pyloric stenosis, Meckel's diverticulum, TEF, annular pancreas, gastroschisis, and malrotation. We observed CVS anomalies with IAs including VSD, atrial septal defect (ASD), aortic arch anomaly, and a single atrium in 5 cases (22.7%).

Interestingly, 3 cases were presented with GUS disorders such as left renal aplasia, ureteral duplication, and uterus unicollis (13.6%), which is not a usual finding.

We also noted 2 cases with respiratory anomalies with IA; 1 had a lobulation anomaly of the lung (3 lobes in the left lung) and the other had aplasia of the lungs (9%). The other findings noted were 1 cleft lip and palate and 1 pes equinovarus.

The mean life span was 10 days. Half of the cases were lost in the first week (11 cases), 27% in the second (6 cases), 9% in the third (2 cases), 4.5% after 1 month (1 cases), and 9% after 2 months (2 cases). Although associated congenital anomalies are risk

factors in the morbidity, the most serious cause of mortality in the series was superinfections resulting from sepsis.

It is well known that surgical intervention is an incontestable risk factor, especially for those newborns that have additional serious risks such as prematurity and low birth weight, as seen in our series. On the other hand, operation is the first choice for treatment in IA.

The conclusions of our study can be listed as follows:

1) The most commonly associated anomalies with IA were GIS anomalies.
2) Low birth weight and prematurity were serious risk factors for death in cases with IAs.
3) The most common cause of death was bacterial sepsis.
4) Surgical intervention might be an additional factor contributing to the occurrence of bacterial sepsis in those newborns that already display serious risk factors such as low birth weight and prematurity.

IA is a significant GI tract anomaly among the perinatal autopsy cases in our institutional archives. Although perinatal screening tests today are very well developed, IA can still be a cofactor in mortality in neonates because of the underlying risks.

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