Diplomarbeit

Postoperative Outcome after Small Bowel Atresia

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Graz, am ……

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Zusammenfassung

Hintergrund

Trotz der verbesserten pränatalen Diagnostik, intensivmedizinischen Neonatologie und operativen Errungenschaft, sind Dünndarmatresien noch immer mit längerem Krankenhausaufenthalt und teilweise mit Sterblichkeit vergesellschaftet. Obwohl sie bereits genau untersucht wurden, ist es jedoch immer noch unklar, welche Faktoren den klinischen Verlauf beeinflussen. Deshalb macht sich diese Studie zur Aufgabe die einwirkenden Faktoren zu ermitteln.

Methoden

Insgesamt wurden 86 Fälle mit Dünndarmatresien, die von 1975 bis 2009 an der Universitätsklinik für Kinder- und Jugendchirurgie der Medizinischen Universität Graz behandelt wurden, erforscht und analysiert.

Ergebnisse

Von den 40 Mädchen und 46 Jungen hatten 40 eine Duodenalatresie, 22 sowohl eine Jejunal- als auch eine Ileumatresie und 2 multiple Atresien des Jejunums und Ileums. Die Geschlechtsverteilung wies ein Verhältnis von männlich : weiblich = 1,15 : 1 auf. Bei 34 Kindern (40%) wurde die Dünndarmatresie pränatal diagnostiziert. Insgesamt betrug das Durchschnittsgestationsalter 36,38 Wochen, wobei 43% der Kinder Frühgeborene waren und 20% unter 2000g wogen. Die Mortalitätsrate lag bei 10,5% und setzt sich aus sieben Kindern mit Duodenalatresien und zwei Kindern mit Ileumatresien zusammen. Es wurden keine chirurgischen Komplikationen verzeichnet, die zum Tode führten. Sechs dieser Kinder waren unreif und sieben hatten assoziierte Anomalien. Unmittelbare Todesursachen waren Sepsis, kongenitale Herzerkrankungen und Kurzdarmsyndrom. Zu den häufigsten assoziierten Anomalien zählten mit 45% bei allen Kindern gastrointestinale Malformationen wie Malrotation, Ösophagus-/Kolon-/Analatresie, Gastroschisis und Meckel-Divertikel. Kinder mit Duodenalatresien wiesen in 43% Trisomie 21 und in 38% kongenitale Herzerkrankungen auf und zuletzt zeigten Neugeborene mit Jejunalatresien in 23% Mukoviszidose als Ursache. Anastomosenstenose, Sepsis und Ileus/Subileus führten die Liste der postoperativen Komplikationen an.

Schlussfolgerung

Die Inzidenz der Dünndarmatresien stieg in den letzen 15 Jahren stetig an, allen voran die Inzidenz der Duodenalatresien mit assoziierter Trisomie 21. Die Mortalitätsrate blieb in den letzten 35 Jahren mit einem Median von 2 Kindern pro Jahrzehnt konstant. Die Gesamtmortalität ist abhängig von Frühgeburtlichkeit, assoziierten Anomalien und Lokalisation der Atresie und korreliert nicht mit verzögerter Diagnosestellung, pränataler Diagnose und Trisomie 21. Zudem gibt es keinen Zusammenhang zwischen postoperativem Outcome und Lokalisation der Atresie. Kinder mit Jejunalatresien leiden häufiger an Frühkomplikationen als Kinder mit anderen Dünndarmatresien, während Kinder mit Duodenal- und multiplen Atresien mehr Langzeitkomplikation aufweisen, die im Allgemeinen häufiger vorkommen, als in der Literatur beschrieben. Mehr als ein Drittel der Kinder mit Dünndarmatresien hatten in ihren ersten beiden Lebensjahren gastrointestinale Probleme in Form von Ileus/Subileus Symptomatik und ausbleibender Gewichtszunahme.

Schlagworte: Dünndarmatresie, Duodenalatresie, Jejunalatresie, Ilealatresie, Intestinalatresie

Abstract

Background

Despite improved prenatal diagnosis, neonatal intensive care and better surgical achievements, small bowel atresias are still associated with prolonged hospitalization and at times mortality. Although they are extensively investigated, it's not yet possible to define clearly which factors affect the clinical course. The aim of this study is to identify factors which influence morbidity and mortality.

Methods

86 cases of small bowel atresia treated from 1975 to 2009 at Department of Pediatric and Adolescent Surgery at Medical University of Graz were retrospectively analyzed.

Results

Out of 40 girls and 46 boys, 40 infants had a duodenal atresia, 22 either a jejunal either an ileal atresia, and two had multiple atresias localized in jejunum and ileum. The sex ratio showed a male : female ratio of 1.15 to 1. 34 children with atresia were diagnosed prenatally (40%). Overall the average gestational age was 36.38 weeks, whereat 43% were premature infants and 20% weighed under 2000g. The mortality rate is 10.5% and is consists of seven infants with duodenal atresia and two infants with ileal atresia. There were no surgical complications, which led to death. Six of these children were premature and seven had associated anomalies. The leading cause of death was sepsis, congenital heart disease and short bowel syndrome. Among the associated anomalies there were 45% gastrointestinal malformations as malrotation, esophageal/colon/anal atresia, gastroschisis and Meckel-diverticulum. Newborns with duodenal atresia had in 43% trisomy 21 and in 38% congenital heart disease and finally infants with jejunal atresia showed in 23% mucoviscidosis as a cause. Anastomotic stenosis, sepsis and ileus/subileus led the list with postoperative complications.

Conclusion

Incidence of small bowel atresia increased steadily during the last 15 years, especially the incidence of duodenal atresia with associated trisomy 21. The mortality rate during the last 35 years remained constant with a median of two

dead infants in a decade. Overall mortality is related to premature birth, associated anomalies and location of atresia and isn’t dependent on delayed diagnosis, prenatal diagnosis or trisomy 21. There is no correlation between postoperative outcome and location of atresia. Infants with jejunal atresia have more early complications than infants with small bowel atresia, while infants with duodenal and multiple atresias have more long-term complications, which are in this group of patients higher when compared with the literature. More than one third children with congenital small bowel atresias had during their first two years of life gastrointestinal problems related to either ileus/subileus symptomatology or lack of weight gain.

Keywords: small bowel atresia, duodenal atresia, jejunal atresia, ileal atresia, intestinal atresia

1. Introduction

Despite improved prenatal diagnosis, neonatal intensive care and better surgical strategies, small bowel atresia is still associated with prolonged hospitalization and at times mortality. It's not yet possible to make a clear statement which factors affect the clinical course.

The underlying surgical problem comes from the caliber differences of the bowel segments with a large bowel segment proximal of the atresia and a small bowel segment distally. Thus, special anastomotic techniques are necessary and postoperative stepwise increments of enteral nutrition take longer until an adequate oral feeding is achieved.

The aim of this retrospective study was to describe small bowel atresias treated in Department of Pediatric and Adolescent Surgery at Medical University of Graz during the last 35 years. This study intends also to identify factors, which have an influence on the outcome of babies.

1.1. Anatomy

The small intestine has a form of a loopy thin walled canal, which ranges from the pylorus to large intestine with the ileo-caecal valve as its lower end. It accomplishes the majority of the chemical and mechanical digestion and absorption. The bowel length is about 304 ± 44 cm in the newborn and in premature children it is about 142 ± 22 cm. The small bowel is arranged in the central and lower part of the abdominal cavity surrounded by the large intestine. Three different sections of the gut can be described: the duodenum, the jejunum and ileum.[1,16]

1.1.1. Duodenum

The duodenum is subdivided in four parts:

- Pars superior
- Pars descendens
- Pars inferior

- Pars ascendens

It is fixed by the dorsal layer of peritoneum and the ligamentum suspensorium duodeni Treitz.[2]

Pars descendens has two papillae for the secretions of bile and pancreatic juice:

- Papilla duodeni minor
  Here ductus pancreaticus accessorius ends
- Papilla duodeni major Vateri
  Here ductus choledochus and ductus pancreaticus end

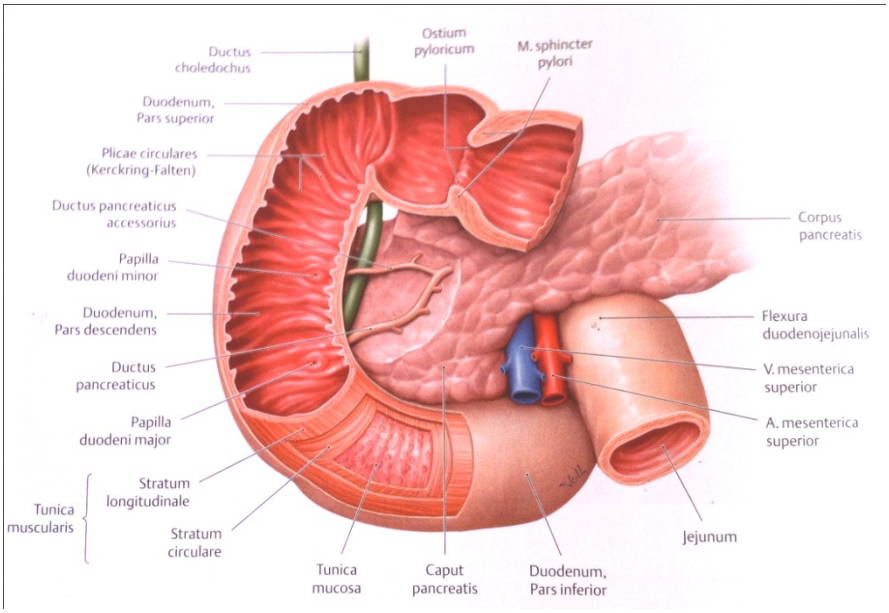


Figure 1: Anatomy of the duodenum. Data from Michael Schünke, Erik Schulte, Udo Schumacher, Markus Voll, Karl Wesker. Prometheus: Hals und Innere Organe. Stuttgart: Georg Thieme Verlag KG; 2005. p. 188.

The oral two centimeters of pars superior are located intraperitoneally and the rest of duodenum is located retroperitoneally. The pancreatic head lies within the concavity of duodenal c-loop. The congenital malformation of the pancreas annulare (incomplete rotation) causes in most cases duodenal stenosis or atresia.[2]

The duodenum ends in flexura duodenojejunalis (Treitz's flexur), which assigns the transition from the duodenum into the jejunum. The small bowel distal of the Treitz's flexur is again flexible in motion.[2]

Duodenum is supplied by arteria gastroduodenalis and arteria mesenterica superior and the nerves branch from the solar plexus.[1]

1.1.2. Jejunum and Ileum

Generally the jejunum and ileum have the same wall structure, however there are specific differences between them in folding relief and in vascularisation. Within the jejunum the plicae circulares are arranged closer than in the ileum and the ileal submucosa has numerous lymphoid follicle for the immunological reaction.

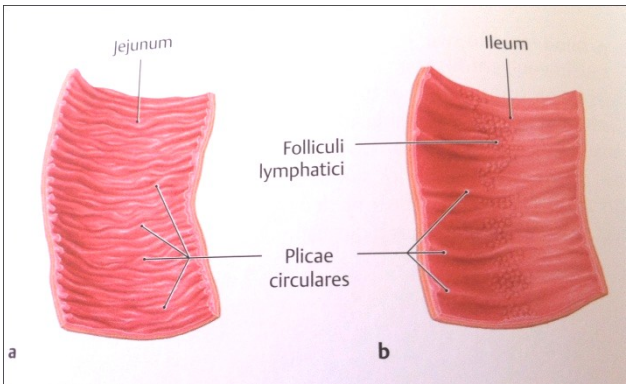


Figure 2: Wall structure of the jejunum and the ileum. Data from Michael Schünke, Erik Schulte, Udo Schumacher, Markus Voll, Karl Wesker. Prometheus: Hals und Innere Organe. Stuttgart: Georg Thieme Verlag KG; 2005. p. 190.

Moreover the jejunum is wider, thicker, more vascular, deeper in color and heavier. The jejunum is arranged in the umbilical and left iliac regions whereas the ileum fills in the umbilical, hypogastric, right iliac and pelvic regions. Both are fixed to the posterior abdominal wall by the mesentery that extends from the superior mesenteric vessels and the plexuses of sympathetic nerves down to the right abdominal part close to the ileocaecal valve.[1,2]

1.2. Intestinal Atresia

This term describes basically a partial (stenosis) or complete (atresia) obstruction, the latter with a congenital absence of the bowel lumen. It can be diagnosed by prenatal ultrasonography. Causes of intestinal atresia are ischemic event and failure of lumen. Louw and Barnard found in 1955 that most jejunoileal atresias are caused by a late intrauterine mesenteric vascular catastrophe by subjecting dog fetuses in the course of gestation. They proposed that “a ligation of the feeding vessels of foetal bowel may end in disappearance of the infracted part of the small bowel”. With the consideration that the vascular origin wasn’t able to cause necrosis, but an atresia by a functional problem with resultant defective peristalsis.[3,5,17]

1.2.1. Duodenal Obstruction

1.2.1.1. History

The duodenum is most frequently concerned by intestinal obstruction during the embryology. The first reported case of duodenal atresia was described by Calder in 1733 and the first successful operation was accredited to Vidal and Ernst in 1905 and 1914. Cordes showed in 1901 the typical clinical findings and in 1929 Kaldor succeeded by collecting 250 patients with duodenal atresia reported in the literature. A review by Webb and Wangensteen demonstrated only nine survivors in 1931.[5]

At the beginning of the twentieth century congenital duodenal obstruction was a lethal disease because of inadequate nutrition, lack of surgical knowledge, associated congenital anomalies and substandard neonatal care skills. With the introduction of a side-to-side anastomosis and advancement in intensive unit care the survival rate reaches today nearly 100%. Historically, when a duodenal atresia was found associated with trisomy 21, the obstruction was neglected from treatment in many cases. Not until the 1970s were the social and medical ethics of the universal operative approach for these infants changed.[3,5]

1.2.1.2. Epidemiology

The incidence of duodenal atresia has been accounted for 1 in 5,000 to 10,000 births. While no specific genetic abnormality is established, there are a high number of associated anomalies and the incidence in siblings and offspring indicates genetic association in some cases.[5]

Around half of all newborns with duodenal obstruction have a congenital anomaly of another organ system. In the following table the most frequently associated anomalies are listed:

Associated Anomalies	
Down syndrome	**28 %**
Annular pancreas	**23 %**
Congenital heart disease	**23 %**
Malrotation	**20 %**
Esophageal atresia/ tracheoesophageal fistula	**9 %**
Genitourinary anomalies	**8 %**
Anorectal anomalies	**4%**
Other bowel anomalies	**4%**
Other anomalies	**11 %**

Table 1: List of associated anomalies; Data from Sweed Y:Duodenal obstruction. In Puri P (ed): Newborn Surgery, 2nd ed, London, Arnold, 2003, p 423

1.2.1.3. Etiology

Ladd classified duodenal obstructions in intrinsic and extrinsic lesions. The following table contains the most common causes:

Ladd classification	
Intrinsic lesion	**Extrinsic lesion**
Duodenal atresia	Annular pancreas
Duodenal stenosis	Malrotation
Duodenal web	Peritoneal bands
	Anterior portal vein

Table 2: Ladd classification: Several congenital lesions whether intrinsic or extrinsic can cause complete or partial obstruction

In the sixth week during embryonic development bowel epithelium proliferates at rapid rate and occludes the intestinal lumen. Over the following weeks the intestinal lumen reopens by apoptosis of the epithelium. If this process fails it causes a partial or complete atresia, a membrane or a membrane with a hole.[5] Thus, an intrinsic obstruction can vary among atresia, stenosis, perforate or imperforate web. The intrinsic blockage occurs in 85% near the junction of pars superior and pars descendens of the duodenum. In the case of a persistent web the bowel becomes dilated due to the gastric secretion and swallowed amniotic fluid until it becomes the shape of a “windsock”.[3,5]

Duodenal stenosis or atresia can be caused by external compression or a malrotation with the Ladd’s band fixing the malrotated coecum lying in the upper position to the liver, thereby compressing the duodenum from outside. The result is a partial obstruction. Since air can pass into the distal intestine it is often characterized by a delay of diagnosis and treatment.[3]

The most common cause of a duodenal atresia is a pancreas anulare: The developing pancreas is composed of two pouches, a larger dorsal and a smaller ventral. If the ventral part does not rotate completely, it results in a ring of pancreatic tissue, which surrounds the duodenum and causes total obstruction.[5]

Finally, an anterior portal vein crossing the anterior surface of the descending part can cause an obstruction. This rare anomaly is often associated with a complex congenital heart disease.[5]

1.2.1.4. Classification

Gray and Skandalakis have divided the duodenal atresia into three types:

- Type 1 (92 %)
  This type has an obstructing web consisting of mucosa and submucosa with no muscularis. The web can vary in thickness from one to several millimeters. A complete layer causes an atresia, while perforate web allows some passage of air and fluid but causes in most cases a significant stenosis. In this case the diameter of the obstruction affects the degree of the stenosis and is indirectly proportional to the severity of the clinical presentation. The arteria mesenterica superior is intact.
  A “windsock” is a variant membrane anomaly and can lead to a thin and elongated membrane. In these cases the level of the stenosis seems more distally than it actually is.[5]
- Type 2 (1%)
  The two ends of the duodenum are connected by a short fibrous cord. The arteria mesenterica is intact.[5]
- Type 3 (7%)
  The two ends of the duodenum have no connection to each other, the consistency is total interrupted.[5]

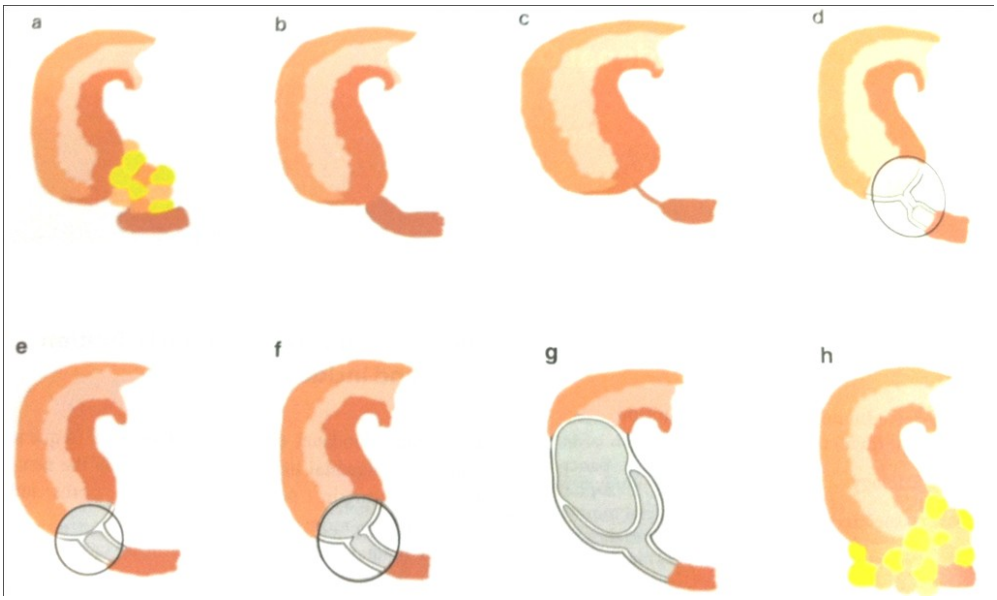


Figure 3: Classification of duodenal atresia. Data from Puri P, Höllwarth M. Pediatric surgery: Diagnosis and management. Berlin: Springer; 2009. p. 384
a) blind ends separated by a gap
b) two ends in apposition
c) ends joined by a fibrous cord
d) duodenal stenosis
e) complete duodenal membrane
f) perforated diaphragm
g) wind-sock web
h) annular pancreas

1.2.1.5. Clinical presentation

The symptoms of duodenal obstructions vary from polyhydramnios, distention of stomach, feeding intolerance, bilious vomiting and aspiration.
Progress in prenatal care allows today to diagnose the majority of cases with duodenal obstructions prior to child's birth by prenatal ultrasonography. Identification of maternal polyhydramnios and distention of stomach are indicators of upper intestinal obstruction. This allows parents and all institutions to prepare for prompt care after birth, but hasn't changed the incidence of associated life-threatening anomalies and the survival rate.[4]

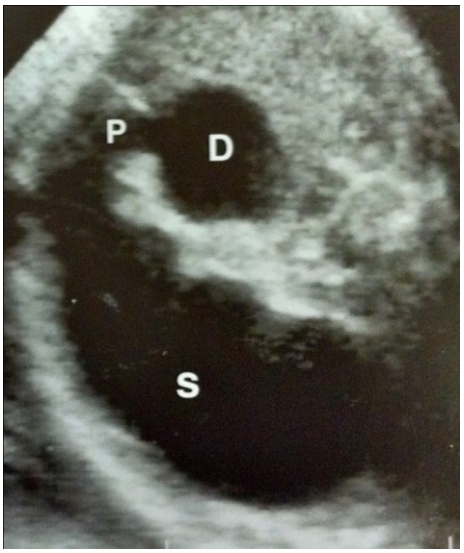


Figure 4: Utrasonography. Data from Puri P, Höllwarth M. Pediatric surgery: Diagnosis and management. Berlin: Springer; 2009. p. 385
"Double bubble sign" of a 24-week gestatinal age fetus.
S=stomach, P=pylorus, D=duodenum

After birth the clinical presentation is normally designated by feeding intolerance and beginning of vomiting within the first 24 h of life. In over 80 % the obstruction is located in the postampullary region, thus vomit is mostly bile stained, therefore repeated bilious emesis is a usual symptom of duodenal obstruction.[4,5]

1.2.1.6. Diagnostic investigation

A plain radiograph of the abdomen in upright position shows the characteristic "double bubble" sign, which is a dilated stomach and pars superior of the duodenum filled with intraluminal air and fluid, but generally no distal air pattern. In this case the plain film is diagnostic and no further imaging of the gastrointestinal tract is required. If the duodenal obstruction is of partial nature a plain film of the abdomen will also show a double bubble sign with some air in the distal intestine.

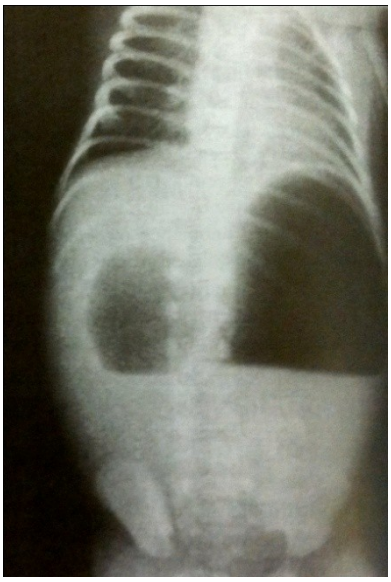


Figure 5: Abdominal radiograph. Data from Puri P, Höllwarth M. Pediatric surgery: Diagnosis and management. Berlin: Springer; 2009. p. 386
Distended stomach and duodenum with double bubble sign with no air beyond the duodenum.

Placement of a nasogastric tube can be used not only as therapy treatment but also as diagnostic aid. A return of more than 30 ml of fluid indicates an intestinal obstruction. The intrinsic obstruction would show a smooth rounded termination in pars descendens and a distal “beaking” effect in pars inferior would indicate a volvulus. In cases, in which the double bubble appearance is not clearly visible, injection of 30 to 60 ml of air trough the nasogastric tube can boost this characteristic imaging sign.[3,4,5]

Incomplete duodenal obstruction may have insignificant symptoms and is sometimes diagnosed after months or even years.

Furthermore a careful physical evaluation for associated anomalies should be indispensable. That should include cardiac and renal ultrasonographic examinations.[4]

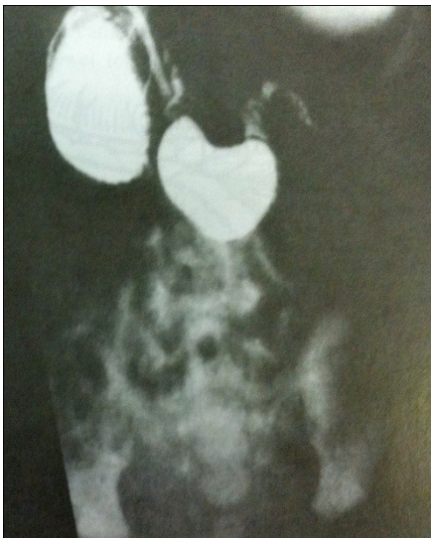


Figure 6: "Wind-sock" web. Data from Puri P, Höllwarth M. Pediatric surgery: Diagnosis and management. Berlin: Springer; 2009. p. 389
Contrast study showing dilated duodenum with duodenal membrane ballooned distally

1.2.1.7. Treatment

Duodenal atresia is a relative emergency and should be corrected surgically during the first days of life. Before the child is transferred into the operating room, his hemodynamic and electrolyte status should be stable.[4]

1.2.1.7.1. Preoperative preparation and intraoperative bedding

Standard treatment includes nasogastric or orogastric tube decompression, fluid and electrolyte replacement and systemic antibiotics at least 30 min before the operation. Body heat should be preserved by overhead warming lights, warming blankets and room temperature of about 24 °C.

1.2.1.7.2. Laparotomy, preparation of the abdominal wall and inspection of the abdominal cavity

The surgical procedure starts with a transverse supra-umbilical abdominal incision 2 cm above the umbilicus, which range from the midline to the right upper quadrant.

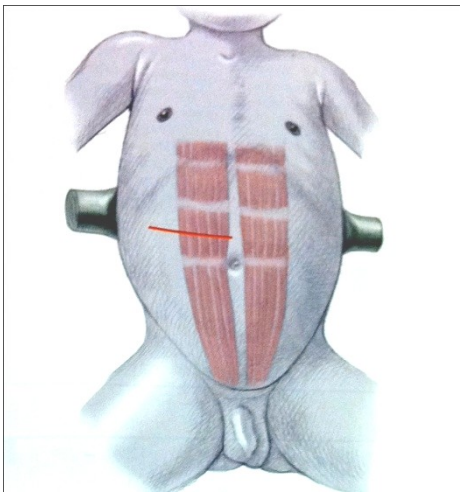


Figure 7: Transverse supra-umbilical abdominal incision. Data from Puri P, Höllwarth M. Pediatric surgery. Berlin: Springer; 2006. p. 205

After opening the abdominal cavity, the bowel is inspected for the presence of other anomalies. To get a better view of the normally dilated and thickened pars superior of the duodenum, the liver is carefully folded up and colon ascendens and flexura coli dextra are moved downward. Nowadays, the procedure is performed laparoscopically in many centers.[4]

1.2.1.7.3. Mobilization of the duodenum

In a first step the duodenum is mobilized and freed it from the retroperitonal attachment. This step is done with caution, especially it's important to avoid carefully a lesion of the ampulla Vateri or the common bile duct. The nasogastric tube is pushed forward into the dilated duodenum and helping to identify the leading part of the obstruction, especially in the case of a "windsock" abnormality. If an annular pancreas is found as cause for the obstruction, the pancreatic tissue should never be divided but bypassed. The duodenum beyond the obstruction is small and decompressed. The need for mobilization of the distal duodenum depends on the location of the atresia and the gap between the two bowel segments. In the case the gap is larger, the surgeon divides the ligament of Treitz, so that the distal duodenum can be mobilized behind the mesenteric vessels to allow an anastomosis without tension.[4]

1.2.1.7.4. Anastomosis

The procedure of choice for infants with duodenal obstruction is the duodeno-duodenostomy. The two applied techniques are either side-to-side duodeno-duodenostomy or a “diamond-shaped” anastomosis – a proximal transverse to distal longitudinal anastomosis.[4]

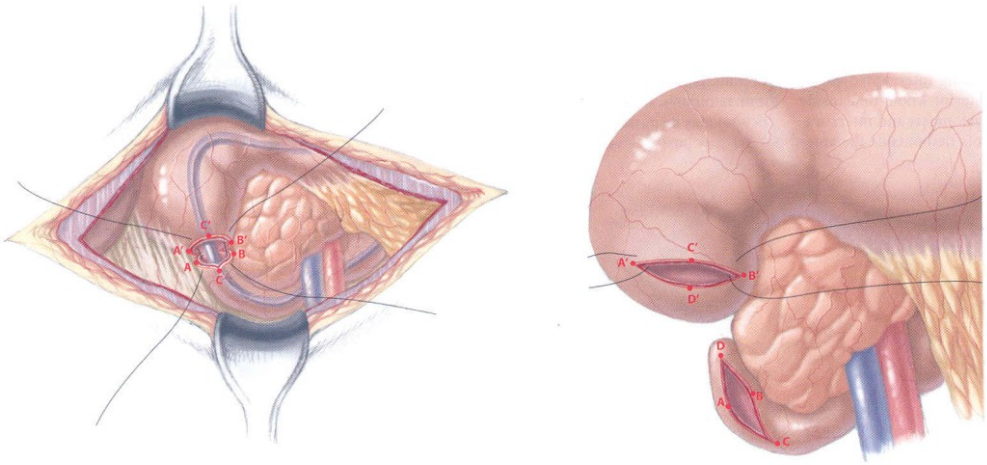


Figure 8 Side-to-side Duodeno-duodenostomy and “diamond-shaped” anastomosis. Data from Puri P, Höllwarth M. Pediatric surgery. Berlin: Springer; 2006. p. 207

1.2.1.7.4.1. Duodeno-duodenostomy of a type 1 atresia

A duodenal web is detected by pushing down the tube into the proximal dilated duodenum. The surgeon places two stitches at the anterior dilated duodenum wall. Afterward he performs a 2.5-3 cm long longitudinal incision above the “transitional zone” between the expanded proximal and the tight distal duodenum portion. Two more stitches are affixed to the margins of the duodenal incision.

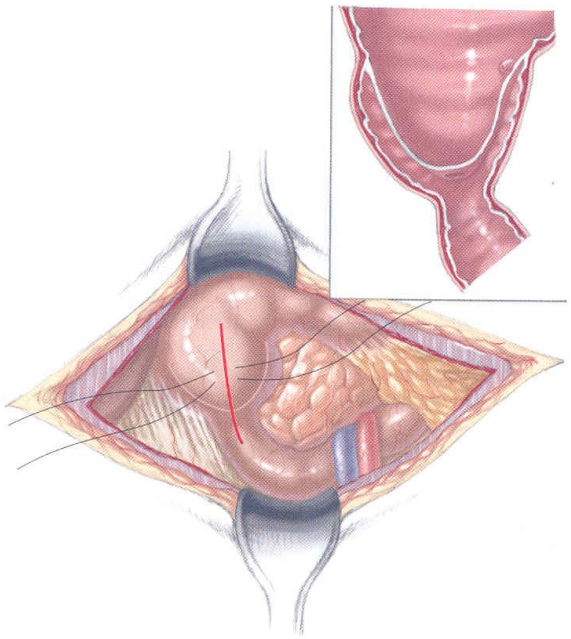


Figure 9 Duodenal web. Data from Puri P, Höllwarth M. Pediatric surgery. Berlin: Springer; 2006. p. 209

The windsock should be well examined inasmuch as the visible transition zone could be several centimeters distal than it appears to be. The ampulla Vateri can be located in pars descendens anteriorly, posteriorly or with twice apertures into the membrane or close to it. Because of this close relationship the situs should be well examined, before performing the resection of the web. Next the surgeon places a single 4/0 Vicryl stitches at the centre of the web and opens up the membrane along its lateral side in order to resect it with a 2-3 mm wide edge. Especially to the medial side of the web should be paid attention in order to avoid damage to ampulla Vateri. Subsequently the duodenum transversely is closed with interrupted 5/0 absorbable sutures. Before its closure, the patency of the distal duodenum should be assured by inserting a catheter through the duodenotomy.[4]

1.2.1.7.4.2. Duodenoduodenostomy of a type 2 and 3 atresia

A Diamond-shaped duodeno-duodenostomy is supposed to allow earlier feeding and good long-term results. Via two traction sutures the proximal duodenum is pulled down to superimpose the distal duodenum. Then the surgeon makes a transverse incision in the distal part of the proximal duodenum and a longitudinal incision in the portion of duodenum distal to the obstruction. A single layer

anastomosis is performed with interrupted 6/0 Vicryl sutures with posterior knots tied inside the posterior wall and with anterior knots tied outside the anterior wall. Before closing the anterior part of the anastomosis, a transanastomotic feeding tube passed from a gastrostomy through the anastomosis downward to the jejunum to allow an early post-operative enteral feeding.[4]

1.2.1.7.4.3. Duodeno-jejunostomy and Gastro-jejunostomy

Duodeno-jejunostomy has the same postoperative results as a duodeno-duodeno-stomy. Gastro-jejunostomy exhibits frequent late complications of marginal ulceration and blind loop syndrome and should not be performed.[5]

1.2.1.7.5. Tapering duodenoplasty

When there is a huge dilatation of the proximal duodenum a tapering duodenoplasty can accelerate the postoperative return of effective peristalsis. This can be performed by suture plication or by resection with the aid of a GIA stapler or needle-tip electrocautery and suture closure. The surgeon performs the tapering on the anterior or anterolateral surface to avoid damage to ampulla Vateri.[5]

1.2.1.7.6. Laparoscopy

There are several recent reports of successfully performed laparoscopic procedures; however, it is not possible to compare the long-term outcomes of this technique with the classic open ones, because the number of patients who have undergone this surgical strategy is limited.[5]

1.2.1.7.7. Postoperative management

A gastrostomy is performed and a gastric tube is inserted to evacuate the excess of fluid, if necessary. The second small tube which has been inserted through the anastomosis in the distal small bowel allows early intestinal nutrition with small increments of volume. As soon as the reflux through the gastric tube gets less and less, oral feeding can be instituted. Postoperative intravenous therapy and antibiotics are provided.[4]

1.2.1.8. Outcome

Long-term outcome after surgery is very good with survival rates reaching 100%. Most of the infants remain asymptomatic and with normal growth. Kokkonen et al. studied 41 patients aged 15-35 years and showed in his study that development and growth were satisfactory. Except a few patients, the great majority remained symptom-free although some gastrointestinal disturbances are common.[4,15] Certainly the prognosis is dependent on various accessory circumstances; especially associated anomalies may downgrade the prognosis essentially. The same is true for premature birth, low birth weight and late diagnosis.The mortality is mainly related to associated anomalies with the leading cause of death especially in infants with trisomy 21 and a complex cardiac anomaly.[4,15]

1.2.2. Jejunal-ileal atresia

1.2.2.1. History

The first reported case of ileal atresia was described by Goeller in 1684 and the first successful repair was accredit to Fockens in 1911.[5]
In 1951 Evans succeeded in identifying 1498 patients with gastrointestinal atresia as reported in his review, however only 139 infants have suvived.[5]

In the fifties congenital jejunoileal atresia had a survival rate of 85% to 90%, but during the last 50 years survival has reached nearly 100% due to improved

surgical techniques, intravenous nutrition and advancement in intensive unit care and pediatric anesthesia.[3]

1.2.2.2. Epidemiology

Jejunoileal atresia and stenosis are a leading cause for congenital intestinal obstruction. Complete atresia occurs in 95% and is caused by closure of the intestinal lumen, while stenosis occurs in 5% and is caused by a partial intraluminal closure producing an incomplete intestinal obstruction.[5]
The prevalence rate of jejunoileal atresia has been reported around 1 in 1000 live births.[4]
Epidemiological studies showed a higher prevalence of small bowel atresia in African-American and Far East Asian infants. Furthermore an increasing risk for the development of small bowel atresia is described after maternal use of pseudoephedrine, acetaminophen, ergotamine tartrate and caffeine during pregnancy.[5]
Boys and girls are equally affected and a third of the children are either born prematurely or small-for-date.[4,5]
Jejunoileal atresia is rarely genetically determined: <1% of patients have chromosomal or other associated anomalies, in contrast to 30% of the Down syndrome children suffering from a duodenal atresia or stenosis.[4,5]
Incidence of associated extraintestinal anomalies is low, including biliary atresia, duodenal atresia, colon atresia, gastric atresia, Hirschsprung's disease and arthrogryposis.[3,4,5]

1.2.2.3. Etiology

A major cause of a jejunoileal atresia is an intra-uterine ischemia of the developing bowel with ischemic necrosis of the affected segments, it results in stenosis or atresia. The ischemic hypothesis is supported by other intrauterine associated ischemic pathologies such as intussusceptions, midgut volvulus, internal hernias with incarceration or peritonitis.

Histological and histochemical abnormalities can be shown up to 20 cm proximal of the obstruction whereas the distall bowel is unused but potentially in normal function.[4]

Another theory for genesis of a jejunoileal atresia is a lack of revacuolization of the solid-cord stage of intestinal development. Factors such as trophic and growth hormones within the amniotic fluid influence the small bowel growth and development. Epithelial plugging is also discussed as a rare cause.[3,5]

1.2.2.4. Classification

Jejunoileal atresia are classified into five types:

- Type 1 (23%)
  This type has a transluminal web or short atretic segment resulting in complete intestinal obstruction. The bowel has no mesenteric defect and the length is not shortened.[4]
- Type 2 (10%)
  That type is represented by a blind-ending distended and hypertrophied for several centimeters proximal bowel attached to a collapsed and underdeveloped distal bowel by a fibrous cord. The bowel is without a mesenteric defect and of normal length.[4]
- Type 3a (16%)
  The two ends of the bowel have no connection to each other. The arteria mesenterica superior is V-shaped defected and the bowel length may be foreshortened.[4]
- Type 3b (19%)
  This anomaly is also called “apple peel” and is a proximal jejunal atresia with an associated malrotation, absence of the network of the arteria mesenterica superior and a large mesenteric defect. The distal bowel is coiled around a single perfusing artery outgoing from the right colic arcades and a jejunal arcade of the mesenteric artery in a helical configuration. The bowel is in the majority of cases significantly reduced in length and associated anomalies are common.[4]
- Type 4 (20%)

That type has multiple atresias like a string of sausages or a combination of types 1-3. There is always followed by a reduction in intestinal length.[4]

Stenosis is not included in this classification.

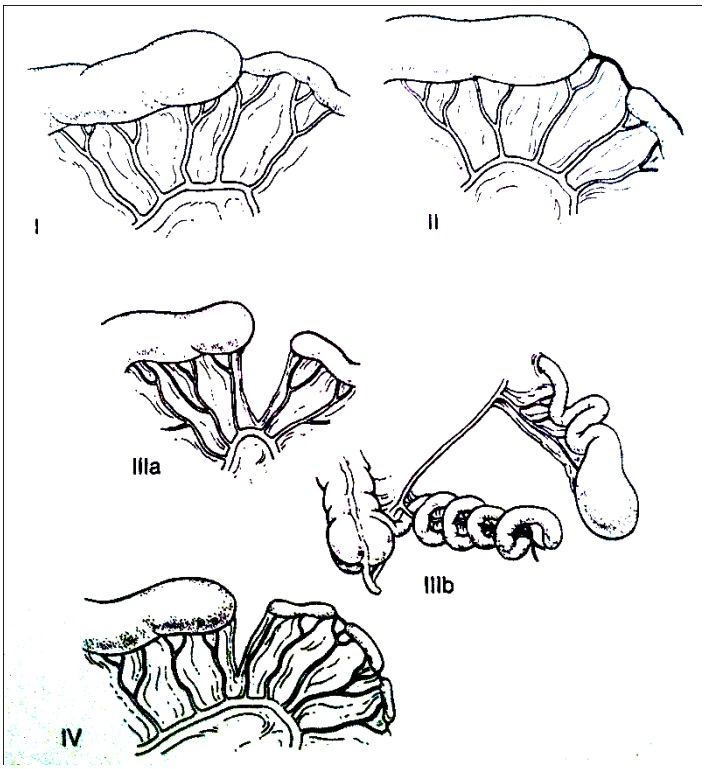


Figure 10: Classification of intestinal atresias. Data from Jay L. Grosfeld, James A. O'Neill, Eric W. Fonkalsrud, Arnold G. Coran. Pediatric Surgery. 6th ed. Edinburgh: Elsevier Mosby; 2006. p. 1276

1.2.2.5. Clinical presentation

The symptoms of jejunoileal atresia are summarized in the following table:

Symptoms	Jejunal atresia(%)	Ileal atresia(%)
Maternal polyhydramnios	38	15
Bilious voiting	84	81
Abdominal distension	78	98
Jaundice	32	20
Failure to pass meconium	65	71

Table 3: Symptoms of jejunoileal atresia[5]

Upper intestinal obstruction causes normally a large intragastric volume at birth. Likewise polyhydramnios, jaundice and bilious vomiting appears earlier in cases with proximal jejunal atresia while significant abdominal distension and late vomiting can be seen in cases with ileal atresia. General distension indicates a lower obstruction whereas proximal jejunal atresia adheres with upper abdominal distension. Typically clinical signs begin 12 to 24 hours after birth.[5]
If the symptoms continue without appropriate diagnostic procedures, intraluminal pressure and/or torsion can induce ischemia, perforation and peritonitis.[4]

1.2.2.6. Diagnostic investigation

Early diagnosis of intestinal atresia is necessary to providethe appropriate therapy. Prenatal ultrasonography identifies duodenal atresia more often than more distal obstructions. Suspicious are multiple distended intestinal loops with pro- and retrograde peristalsis. Postnatally, the diagnosis of jejunoileal atresia can be confirmed by a plain abdominal radiograph. The X-ray in upright position shows distended air and fluid filled loops of the bowel. In cases with high jejunal atresia the X-ray shows only some air-fluid intestinal loops and no gas below the lesion.[5]

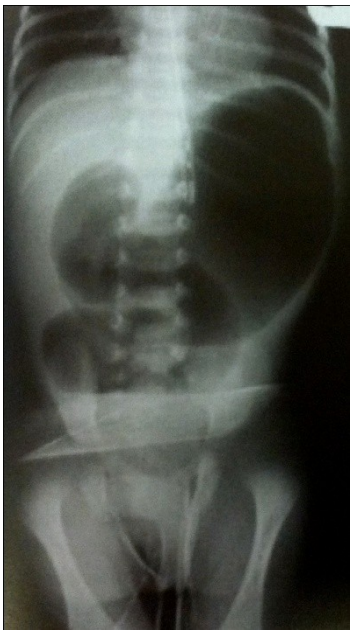


Figure 11: Abdominal radiograph1 . Data from Puri P, Höllwarth M. Pediatric surgery: diagnosis and management. Berlin: Springer; 2009. p. 409

Jejunal atresia showing a few dilated proximal small bowel loops.

Peritoneal calcification is a sign of meconium peritonitis, as a result of intrauterine intestinal perforation.[5]

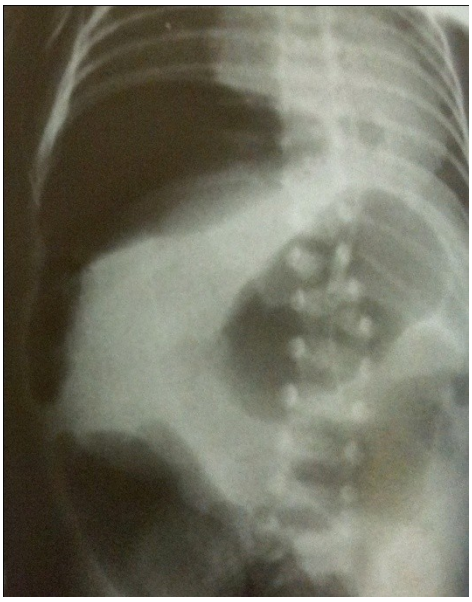


Figure 12: Abdominal radiograph 2. Data from Puri P, Höllwarth M. Pediatric surgery: diagnosis and management. Berlin: Springer; 2009. p. 408
Pneumoperitoneum due to proximal bowel perforation.

In cases of unclear congenital intestinal lesion a contrast enema should be performed in order:

- To differenciate between small or large bowel distention,
- to verify if the colon is used or unused,
- to show the position of the ceacum relating to potential presence of other intestinal anomalies like rotation or fixation anomalies.[5]
-

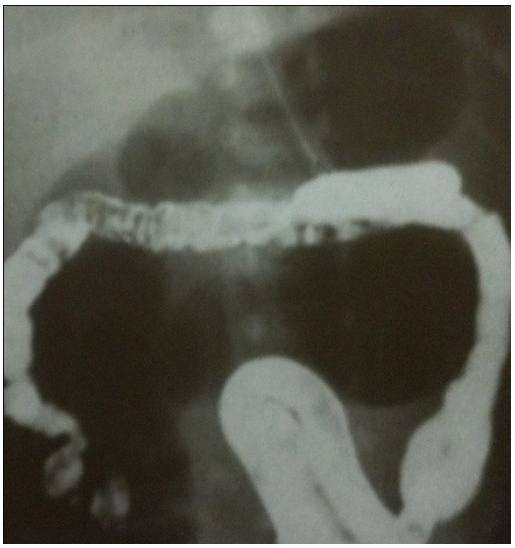


Figure 13: Contrast enema. Data from Puri P, Höllwarth M. Pediatric surgery: diagnosis and management. Berlin: Springer; 2009. p. 410
Normally rotated and patent colon with evidence of proximally small bowel loops.

1.2.2.7. Differential Diagnosis

Infants with congenital intestinal lesion from other causes can show similar symptoms as newborns with jejunoileal atresia. The following table contains the most common differential diagnosis:

Differential diagnosis of jejunoileal atresia
Midgut volvulus
Intestinal stenosis from outside
Meconium ileus
Duplication cyst
Internal hernia
Strangulated inguinal hernia
Hirschsprung's disease
Colonic aganglionosis
Birth trauma
Maternal medications
Prematurity
Hypothyroidism

Table 4: Differential diagnosis of jejunoileal atresia

1.2.2.8. Treatment

Jejunoileal atresia is a relative emergency and should be operated during the first days of life. The hemodynamic and electrolyte status should be stable.[4]

1.2.2.8.1. Preoperative preparation and intraoperative bedding

Nasogastric or orogastric tube decompression, fluid and electrolyte replacement and systemic antibiotics are given at least 30 min before the operation starts. Body heat should be preserved by overhead warming lights, warming blankets and room temperature of about 24 °C.

1.2.2.8.2. Laparotomy, preparation of the abdominal wall and inspection of the abdominal cavity

The surgical procedure starts with a transverse supra-umbilical abdominal incision 2 cm above the umbilicus. After opening the abdominal cavity, the bowel is inspected for the presence of anomalies.[4]
After identification of the pathological type and possible etiology, the patency of the distal small and large bowel should be inspected with saline injection, further distal atretic areas may be found in 6-21%. If a volvulus is present the bowel is untwisted carefully. In a case of short bowel an accurate measurement of residual bowel length proximal and distal to the anastomosis is important. Normally, the total bowel length amounts about 304 ± 44 cm and in premature children it is about 142 ± 22 cm.[4,17]

1.2.2.8.3. Resection

After identification of the atretic region and its proximal distended and distal collapsed bowel the gastrointestinal volume is milked backwards into the stomach and evacuated through a nasogastric tube. A bacteriological specimen is sent for culture.

The proximal dilated atretic segment is resected along the mesenteric bowel border to preserve the mesentery until a normal intestinal diameter has been reached. The distal atretic lesion is incised alongside the antimesenteric border to guarantee that the proximal and distal bowel aperture is of approximately equal size to perform a satisfying anastomosis.[4]

1.2.2.8.4. Anastomosis

The two applied techniques in the surgery are end-to-end and and-to-back (Denis-Browne method) anastomosis with 6/0 absorbable sutures.
In a case with multiple webs an en-bloc resection with a single anastomosis is more productive than multiple anastomosis, if enough bowel length is still available. To avoid the risk of a blind loop syndrome, different techniques such as side-to-side anastomosis, simple transverse enteroplasties, excision of webs and bypassing techniques are not used anymore.
The mesenteric gap is closed by approximating the edges with interrupted sutures, avoiding any ligation of blood vessels. [4]

1.2.2.8.5. Tapering jejunoplasty and plication

When there exists a huge dilatation of the proximal atretic segment a tapering jejunoplasty can accelerate the postoperative return of effective peristalsis. This can be performed by resection with the aid of a GIA stapler or needle-tip electrocautery and suture closure. The indications for tapering are:

- Preservation of bowel length in type 3b or high jejunal atresia
- Reduction of disparity in anastomotic lumen size
- Correction of a failed inversion placation procedure
- Improvement in function of a dilated nonfunctioning mega-duodenum

The plication has in contrast to tapering the benefit of decreasing the risk of leakage from the suture line and preservation of mucosal surface area.[4]
Another technique is antimesenteric seromuscular stripping and inversion plication, which can prevent late insufficiency of plication and preserves a maximum of mucosal surface area for absorption.[4]

1.2.2.8.6. Double tube technique

This technique includes two tubes. The first one is a transanastomotic tube, which splints the anastomosis and enables an earlier beginning with enteral feeding directly into the distal bowel by avoidance the anastomosis. The second tube inserted proximal to the anastomosis has the aim to decompress the accumulating fluid.

1.2.2.8.7. Postoperative Care

Once the output from the proximal tube has declined and the baby has spontaneous bowel motions oral feeding is initiated.[5]

1.2.2.9. Complications

The postoperative complications are anastomotic leaks, stricture formation, transient intestinal dysfunction, blind loop syndrome and short bowel syndrome. Malabsorption and diarrhea can indicate short bowel syndrome, which is the major factor influencing the outcome of infants with jejunoileal atresias. Several operating procedures have been used to improve their outcome.[5]

1.2.2.10. Outcome

The prognosis of jejunoileal atresia depends on the presence of associated anomalies and diseases, prematurity, complications and delay in presentation. Nowadays long-term outcome after surgery has achieved nearly 100% and most affected infants are considered at follow-up as asymptomatic and with normal growth. This is the combined result of optimal intravenous nutrition, improved surgical techniques and advancement in intensive unit care and pediatric anesthesia.[4]

2. Methods

During the last 35 years 86 infants with small bowel atresia were operated in Department of Pediatric and Adolescent Surgery at Medical University of Graz. On the basis of their data, which were completed by hospitalized and ambulant documentation were on hand, a retrospective study has been carried out. For every single infant the following data has been gathered in Microsoft Office Excel table:

- Date of birth
- Gender
- Weight at birth
- Birth size
- Head girth at birth
- Gestational age
- Prenatal diagnosis
- Apgar score
- pH-value of allantoic vein
- Incidence of small bowel atresia
- Location of atresia
- Type of atresia
- Associated anomalies
- Comorbidity
- Operation
- Reoperation
- Duration of stay in hospital
- Duration of stay in intensive care
- Duration of ventilation
- Onset of oral feeding
- Date of total oral feeding
- Complications
- Number of outpatients visits
- Development

At the end of the data inquiry the selected specifications were analyzed via Microsoft Office Excel 2007.

3. Results

3.1. General

In 86 reviewed patients, there were 40 duodenal atresias (45.98%), 22 jejunal atresias (25.29%), 22 ileal atresias (25.29%) and 2 multiple atresias, localized in jejunum and in ileum (3.44 %).

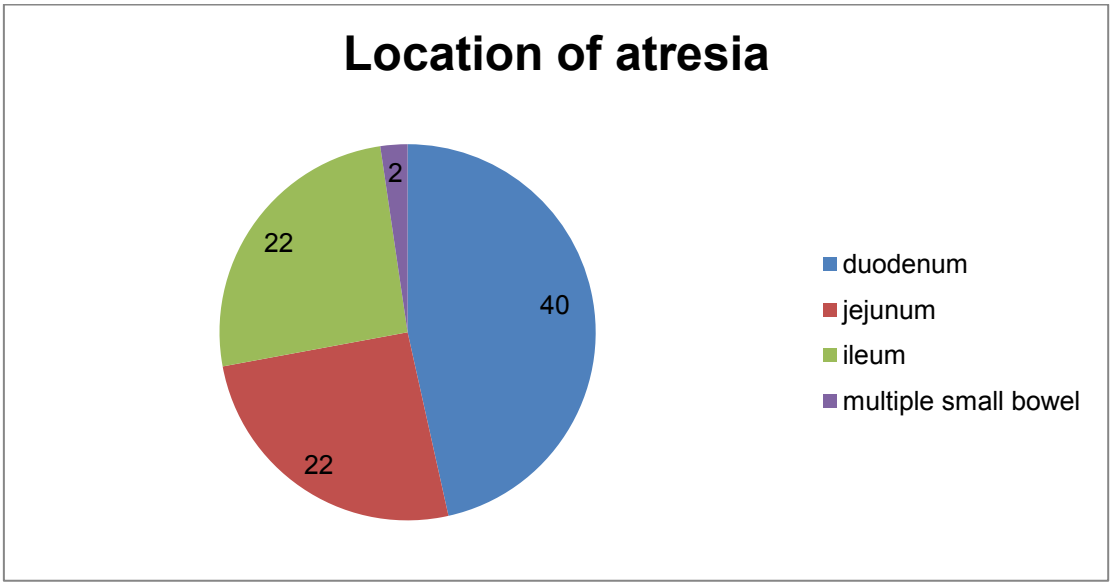


Figure 14: Location of atresia

3.1.1. Weight, size and head circumference at birth and gestational age

The mean birth weight of all 86 newborns within the study was 2487 g ± 670 and its median 2530 g.

The mean body size amounts to 47.5 cm ± 3.5 and its median 48 cm.

In this study the mean head circumference was 32.7 cm ± 2.1 and its median 33 cm.

Finally the mean gestational age accounts for 36.4 ± 2.6 weeks and its median 37.00 weeks. 37 infants were premature (43%).

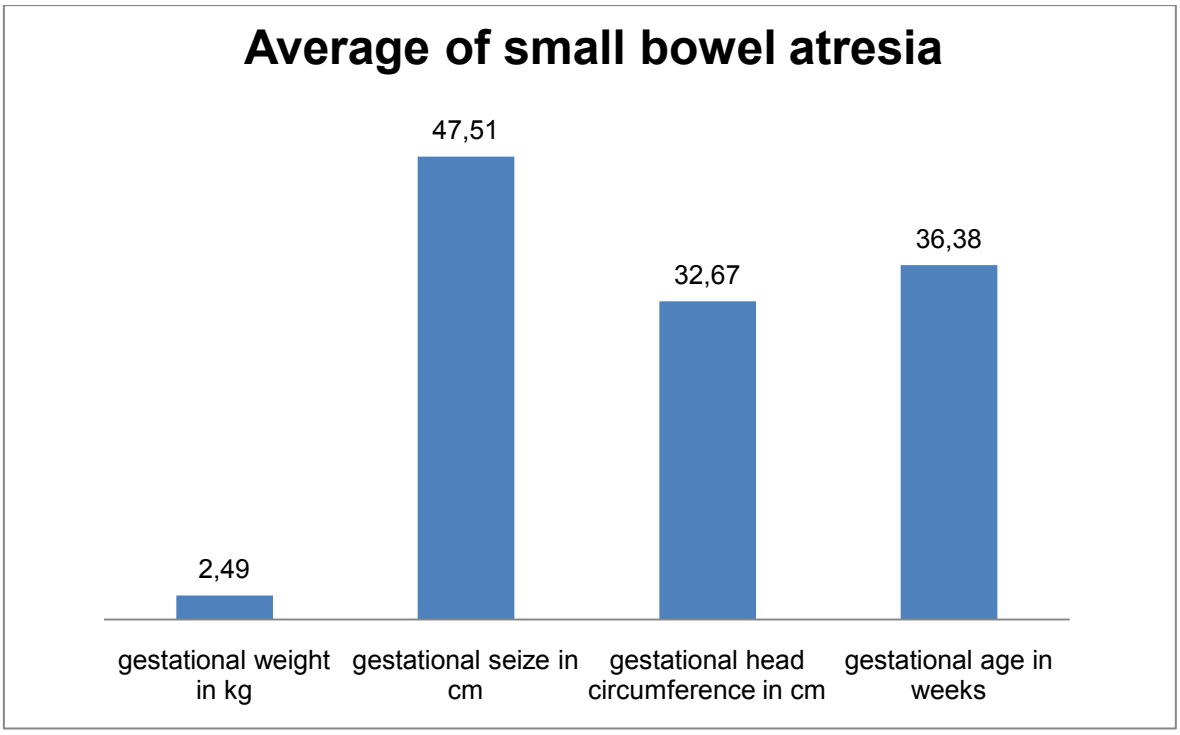


Figure 15: Avarage of gestational weight, body size, head circumference and age

The following figure illustrates the development of gestational age over the past 35 years. Whereas from 1975 to 1979 the mean gestational age amounted 36.4 weeks ± 2.0, it amounted from 1980 to 1984 36.6 weeks ± 3.0, from 1985 to 1989 36.5 weeks ± 1.4, from 1990 to 1994 37.1 weeks ± 2.0, from 1995 to 1999 36.3 weeks ± 2.1, from 2000 to 2004 36.6 weeks ± 3.7 and from 2005 to 2009 35.6 weeks ± 2.8. In conclusion, no significant change over the years can be observed.

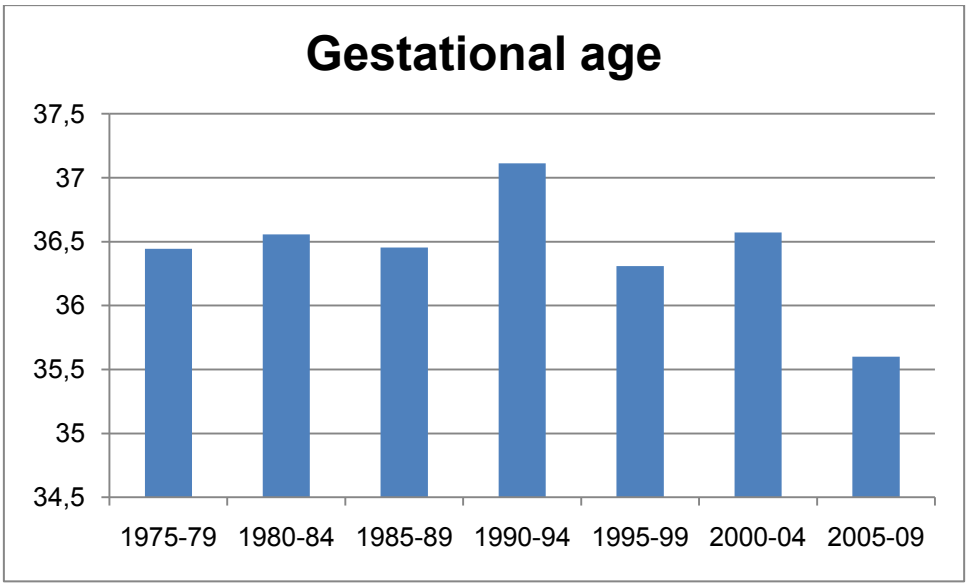


Figure 16: Distribution of gestational age

3.1.2. Gender

Out of 86 newborns 46.51% were girls (n=40) and 53.49% were boys (n=46).

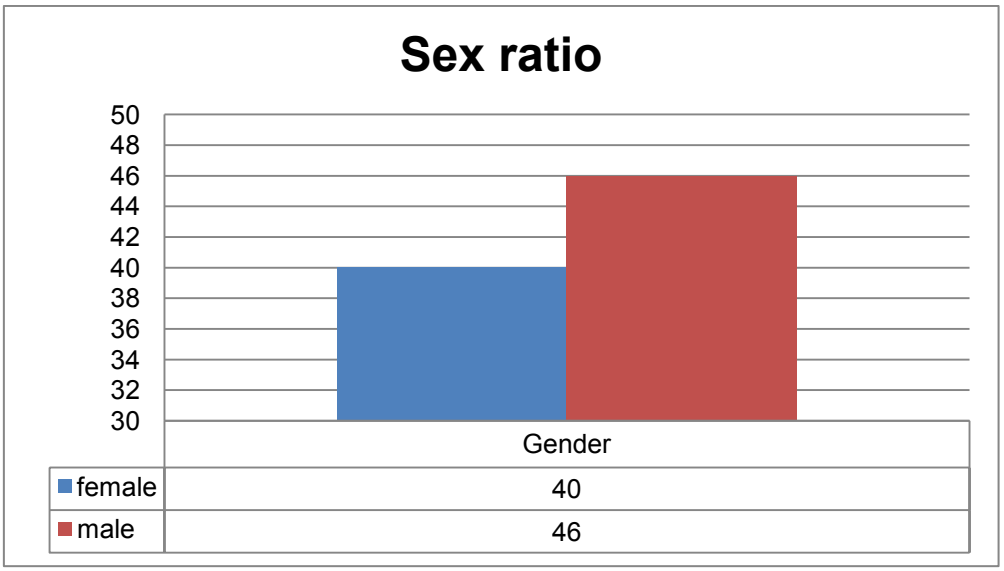


Figure 17: Sex ratio 1

Furthermore half of the girls (n=20) and half of the boys (n=20) out of 40 infants had a duodenal atresia, nine girls (40.91%) and 13 boys (59.09%) of 22 infants had a jejunal atresia and ten girls (45.45%) and twelve boys (54.55%) of 22 infants were born with an ileal atresia.

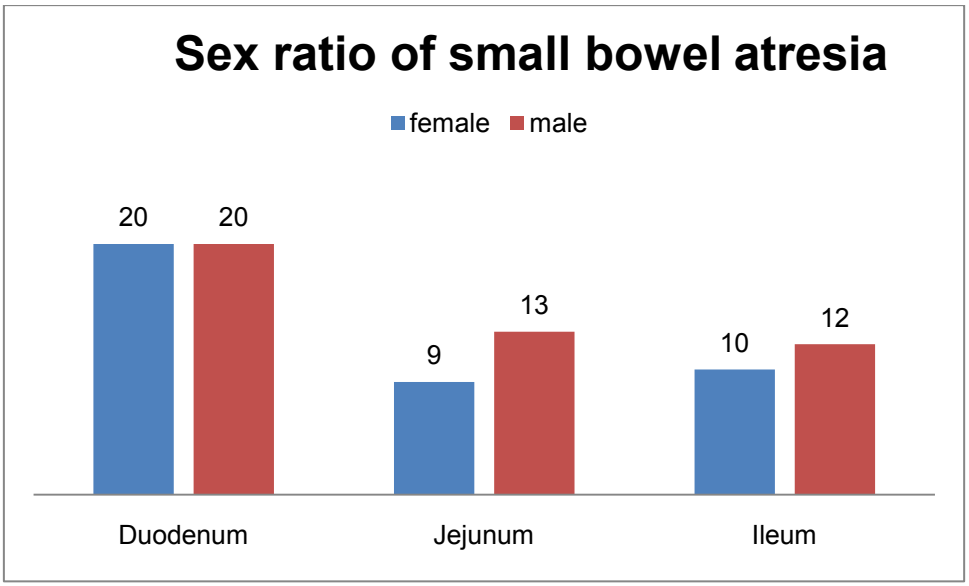


Figure 18: Sex ratio 2

3.1.3. Incidence of small bowel atresia

The following figure shows the incidence of small bowel atresia patients at the Department of Pediatric and Adolescent Surgery at Medical University of Graz during the last 35 years. From 1975 to 1979 there were twelve infants treated, from 1980 to1984 ten infants, from 1985 to 1989 eleven infants, from 1990 to 1994 nine infants, from 1995 to 1999 14 infants, from 2000 to 2004 14 infants and from 2005 to 2009 16 infants.

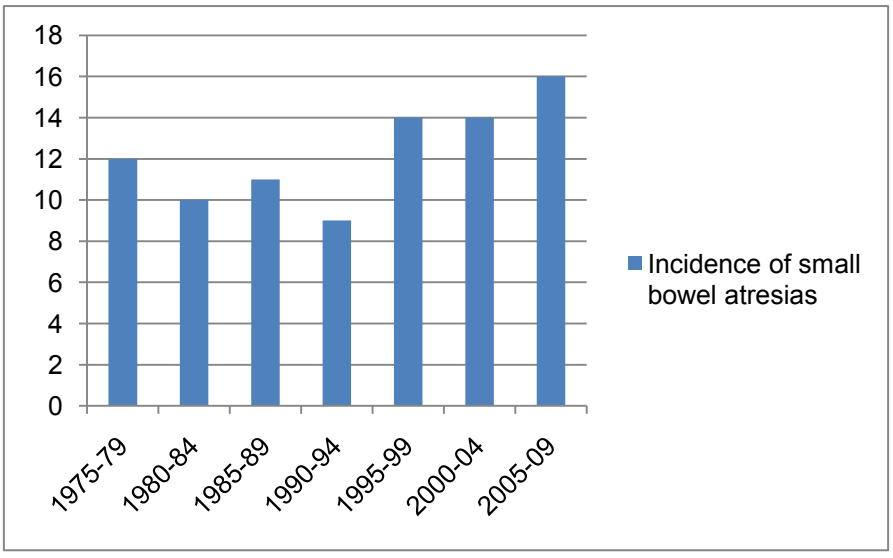


Figure 19: Incidence of small bowel atresia

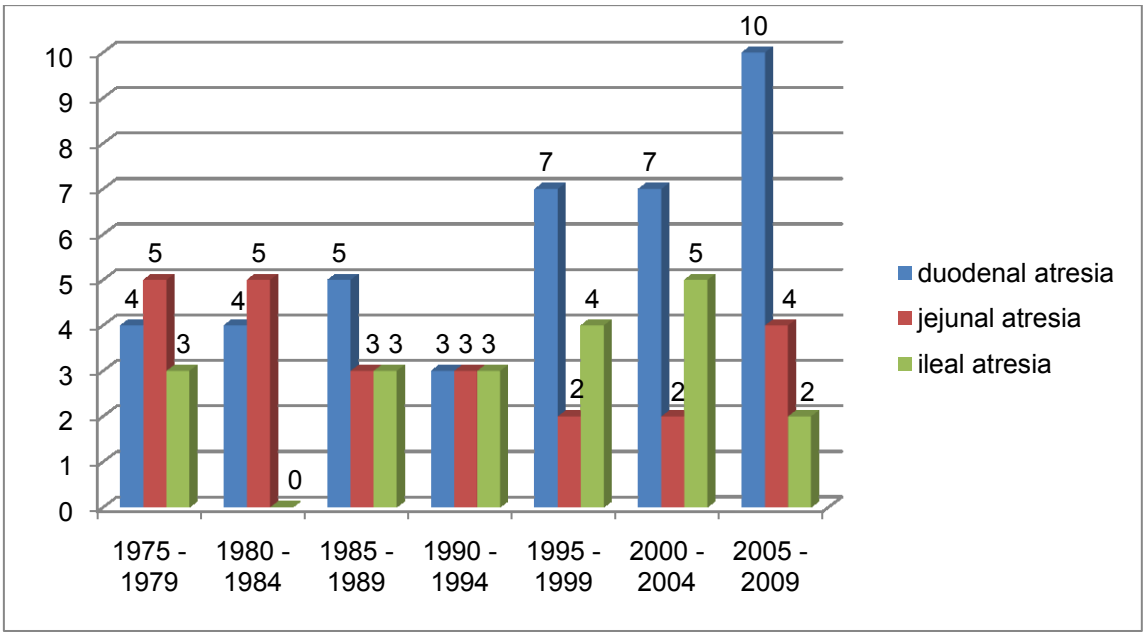


Figure 20: Incidence of small bowel atresia

While the number of cases with jejunal and ileal atresias remained more or less constant over the last 35 years, the number of duodenal atresias increased during the last 20 years from 3 cases from 1990 to 1994 to 7 cases in a period from 1995-1999 and 2000-2004 and reached the highest number with 10 cases from 2005 to 2009.

3.1.4. Location of atresia

3.1.4.1. Duodenal atresia

Of the 40 duodenal atresias, six were located in the proximal part (15.00%), 24 in the pars descendens (60.00%) including 16 cases of associated pancreas annulare and ten in the distal part of the duodenum (25.00%).

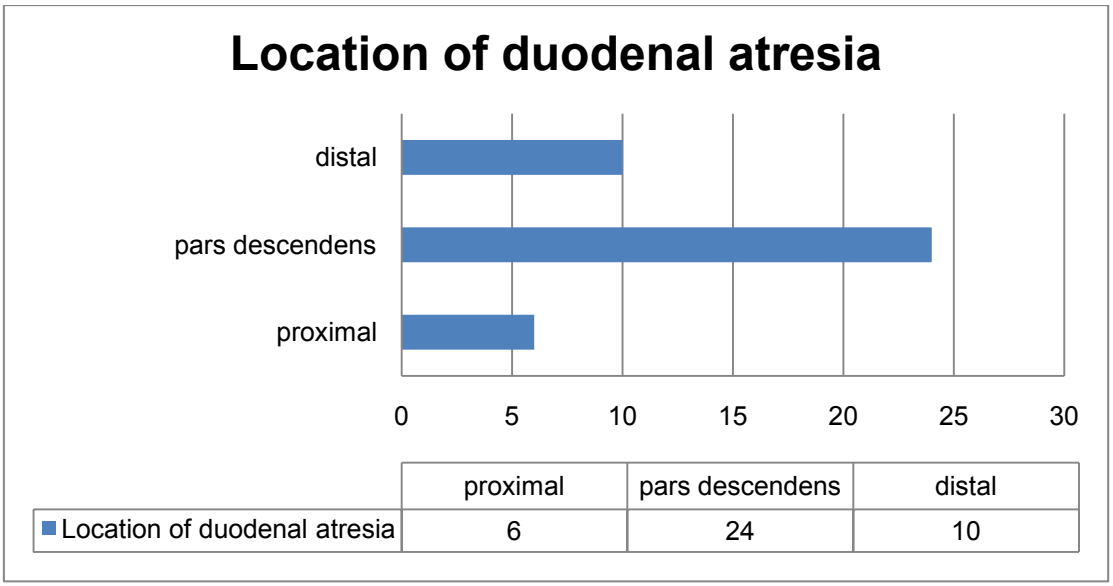


Figure 21: Location of duodenal atresia

3.1.4.2. Jejunal atresia

Figure 20 illustrates the incidence of the location of jejunal atresia. In twelve cases proximal atresia was the most frequent form (48.00%), followed by middle and multiple forms with six cases (24.00%) and the distal form with four cases (16.00%).

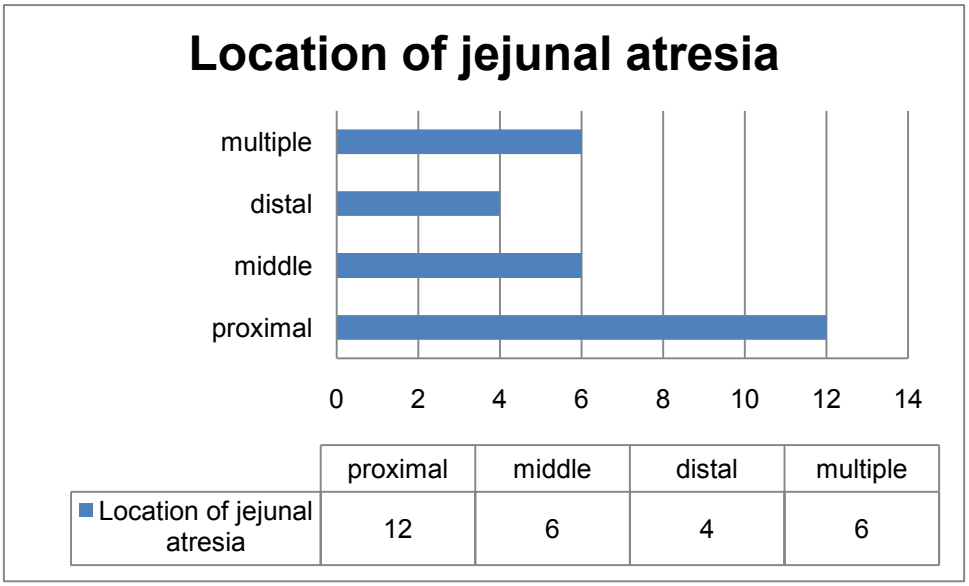


Figure 22: Location of jejunal atresia

3.1.4.3. Ileal atresia

As shown in figure 21, proximal atresia was found in five infants with ileal atresia (20.00%), a middle atresia in three infants (12.00%), a distal atresia in twelve infants (48.00%) and multiple atresias in 5 infants (20.00%).

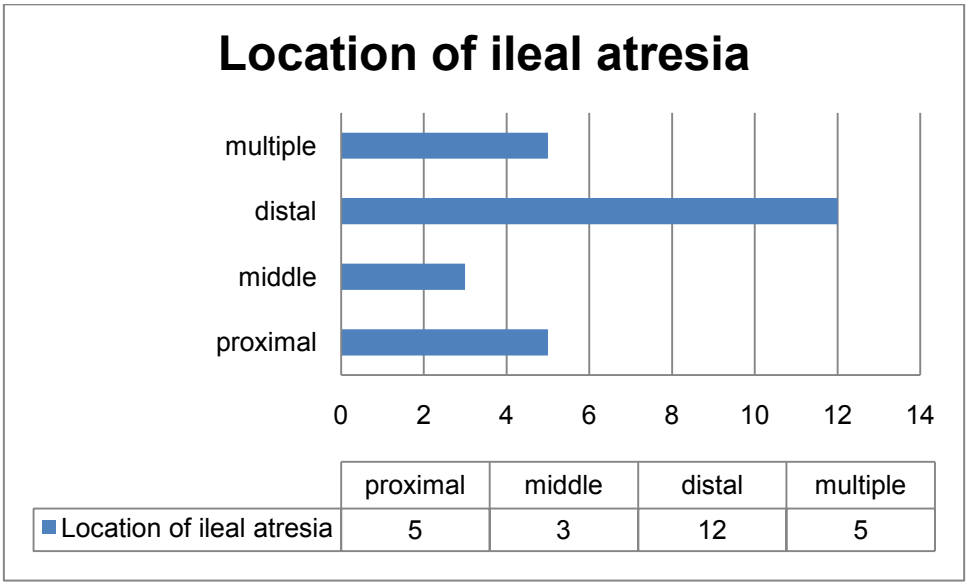


Figure 23: Location of ileal atresia

3.1.5. Type of atresia

3.1.5.1. Duodenal atresia

In 21 of 40 cases of duodenal atresia had a type 1 atresia (52.5%), a type 2 atresia occured two times (5%) and a type 3 atresia in 1 case (2.5%). A stenosis caused by pancreas annulare was present in 16 cases (40%).

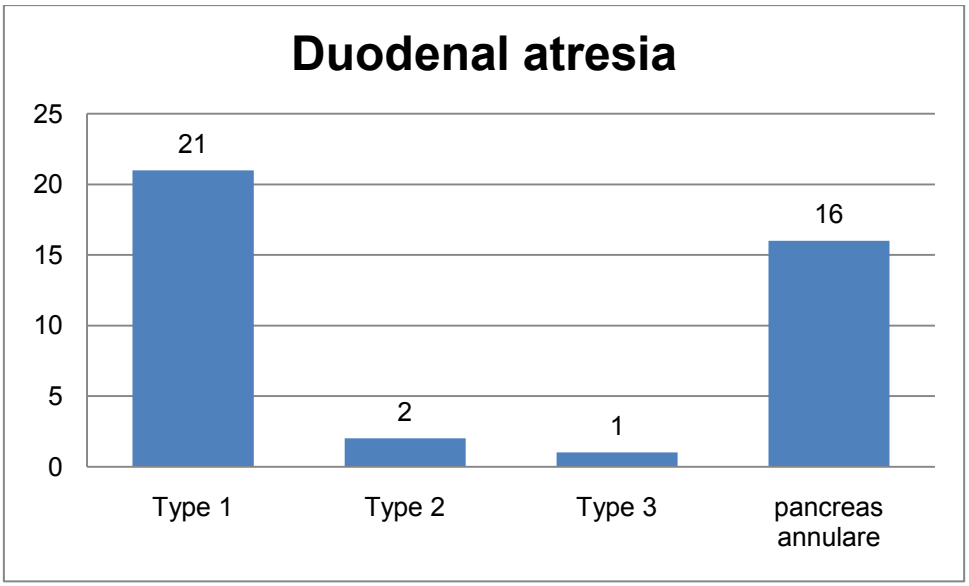


Figure 24: Type of duodenal atresia

3.1.5.2. Jejunal atresia

A type 1 atresia was found in seven of 25 infants (28.00%), a type 2 atresia in two infants (8.00%), a type 3a atresia and a type 3b atresia in five infants (20.00%) and a type 4 atresia in six infants (24.00%).

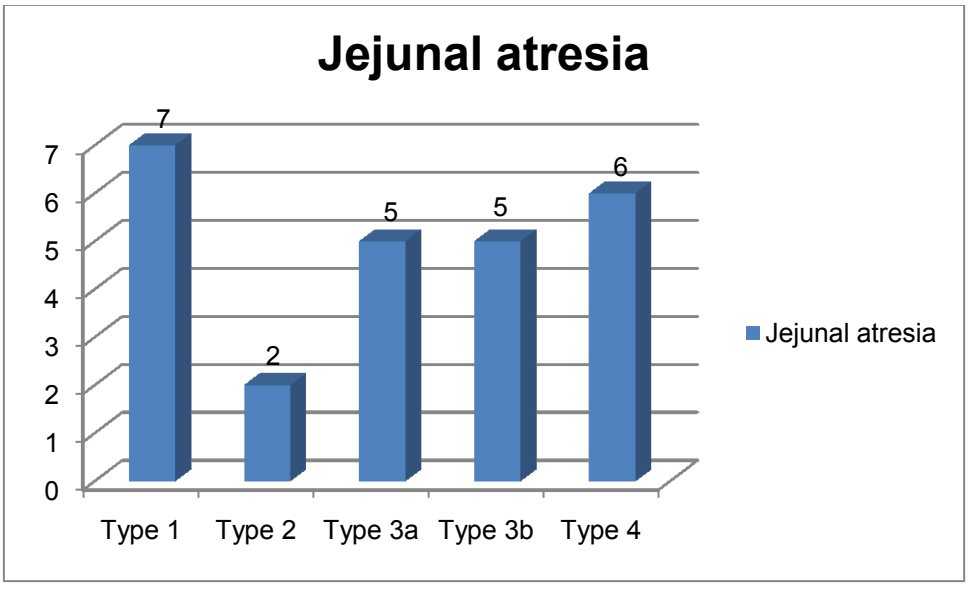


Figure 25: Type of jejunal atresia

3.1.5.3. Ileal atresia

As demonstrated in figure 24, of the 25 ileal atresias eight were a type 1 atresia (32.00%), two a type 2 atresia (8.00%), six a type 3a atresia (24.00%), four a type 3b atresia (16.00%) and five were a type 4 atresia (20.00%).

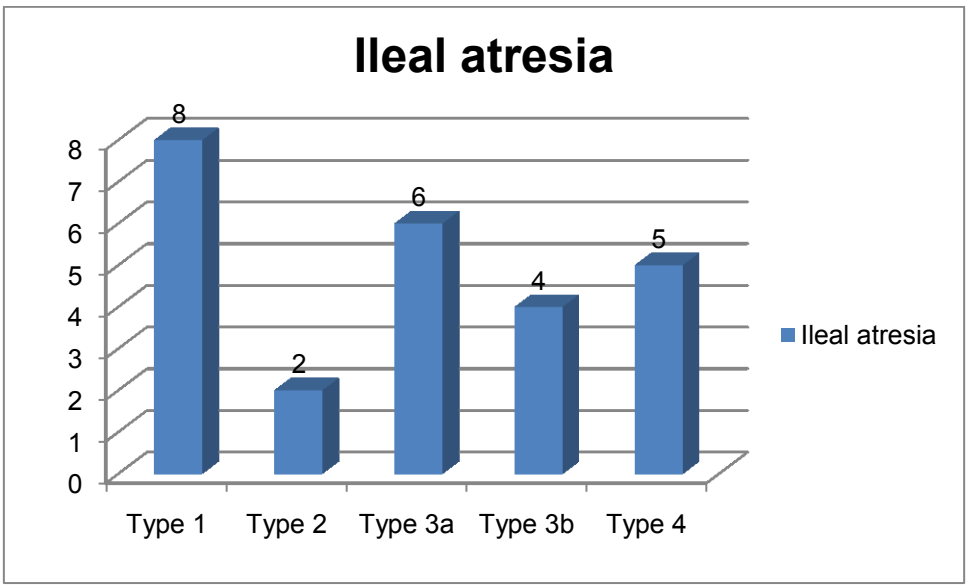


Figure 26: Type of ileal atresia

3.1.6. Associated anomalies

71% of all treated infants with small bowel atresia in the study have associated anomalies.

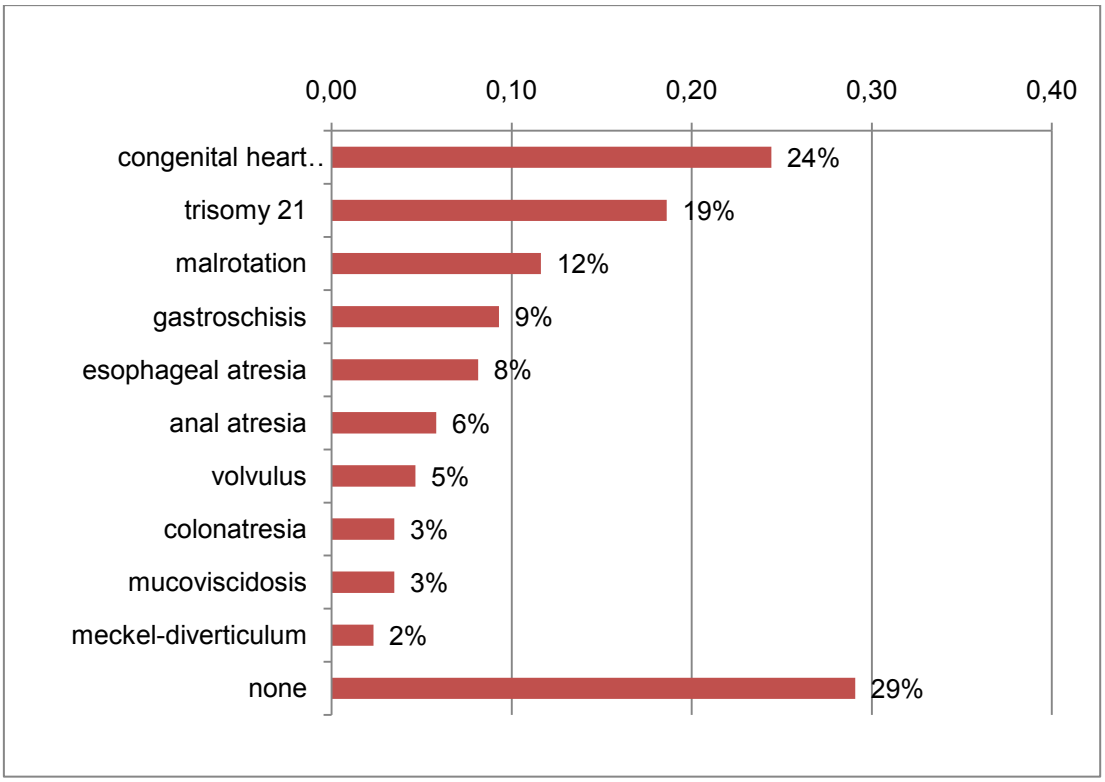


Figure 27: Associated anomalies 1

Congenital heart disease was found at 24%, trisomy 21 at 19%, malrotation at 12%, gastroschisis at 9%, esophageal atresia at 8%, anal atresia at 6%, volvulus at 5%, colonatresia at 3%, mucoviscidosis at 3% and finally Meckel-diverticulum at 2%.

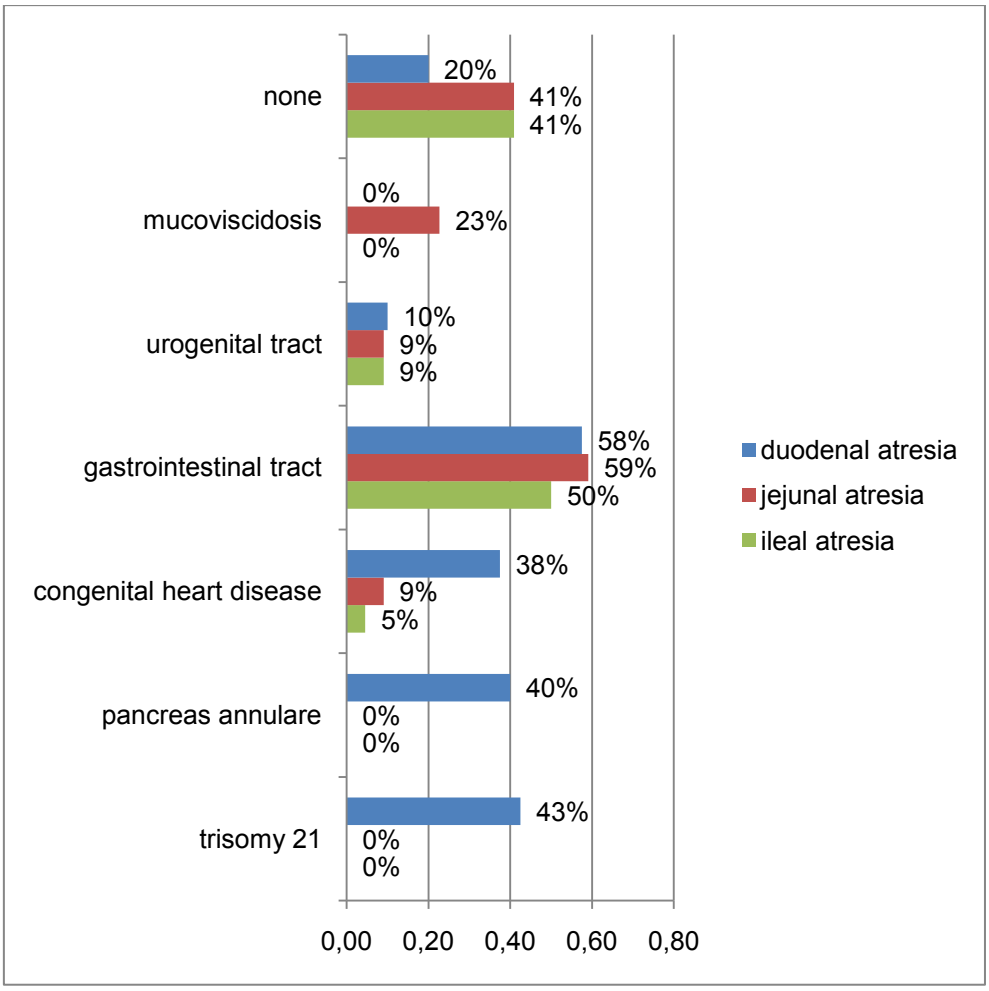


Figure 28: Associated anomalies 2

Retrospectively, 43% of all infants with duodenal atresia had trisomy 21, 40 % pancreas annulare, 38% congenital heart disease, 58% other gastrointestinal anomalies, 10% urogenital anomalies and just 20% have no further anomalies.

Of the babies with duodenal atresia associated with gastrointestinal anomalies, in 7 cases a malrotation was found, in 4 cases an esophageal atresia or a volvulus, in 2 cases a gastroschis or a Meckel-diverticulum and one time an analatresia, a situs inversus or a Morbus Hirschsprung was a secondary finding.

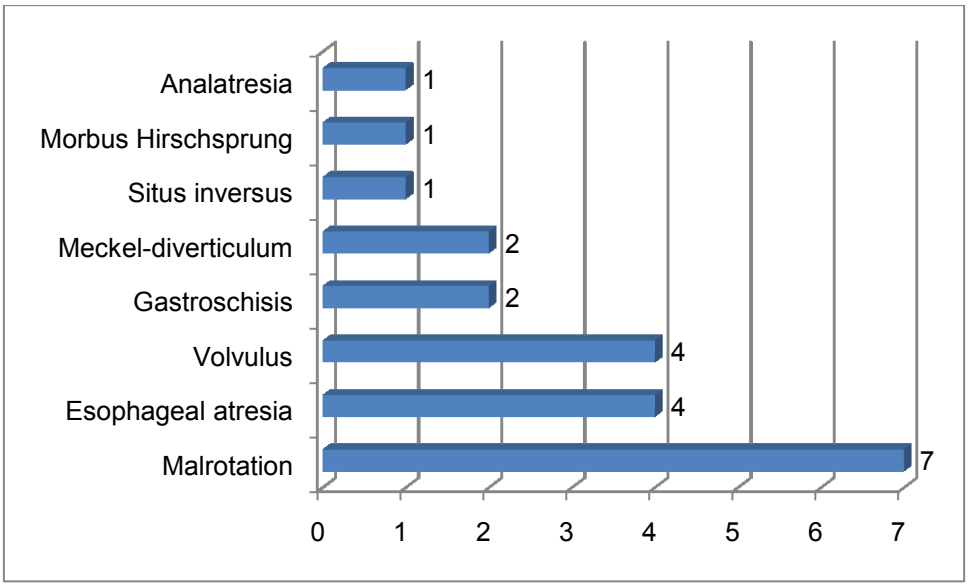


Figure 29: Gastrointestinal anomalies associated with duodenal atresia

Children with jejunal atresia had congenital heart disease in 9%, other gastrointestinal anomalies in 59%, urogenital anomalies in 9%, mucoviscidosis in 23% and no other anomalies in 41%.

Associated analatresia occurred three times, malrotation, esophageal atresia, volvulus and gastroschisis were present in each case one time and one newborn with a jejunal atresia had an associated colonatresia.

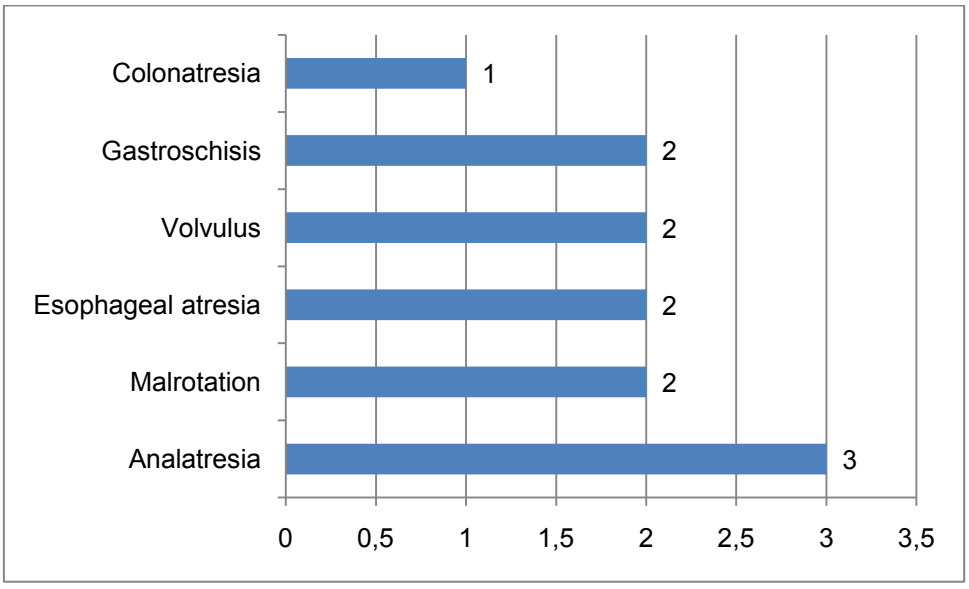


Figure 30: Gastrointestinal anomalies associated with jejunal atresia

And last, infants with ileal atresia showed 5% congenital heart disease, 50% other gastrointestinal anomalies, 9% urogenital anomalies and 41% no other anomalies.

4 of 22 treated babys had an associated gastroschisis, two a malrotation or a colonatresia and one baby had an associated esophageal atresia.

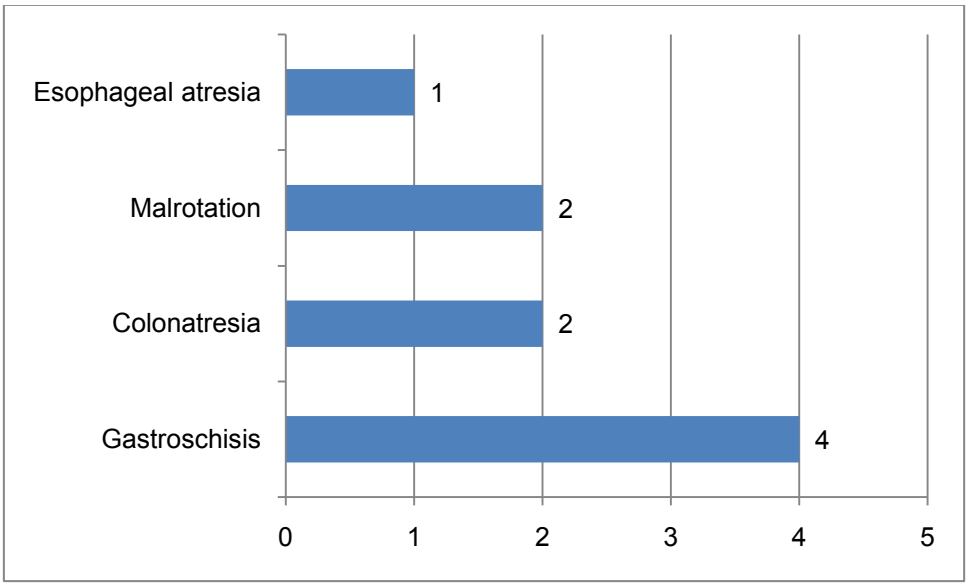


Figure 31: Gastrointestinal anomalies associates with ileal atresia

Considering the relationship between duodenal atresia and trisomy 21, the incidence of both increased correlatively during the last 35 years.

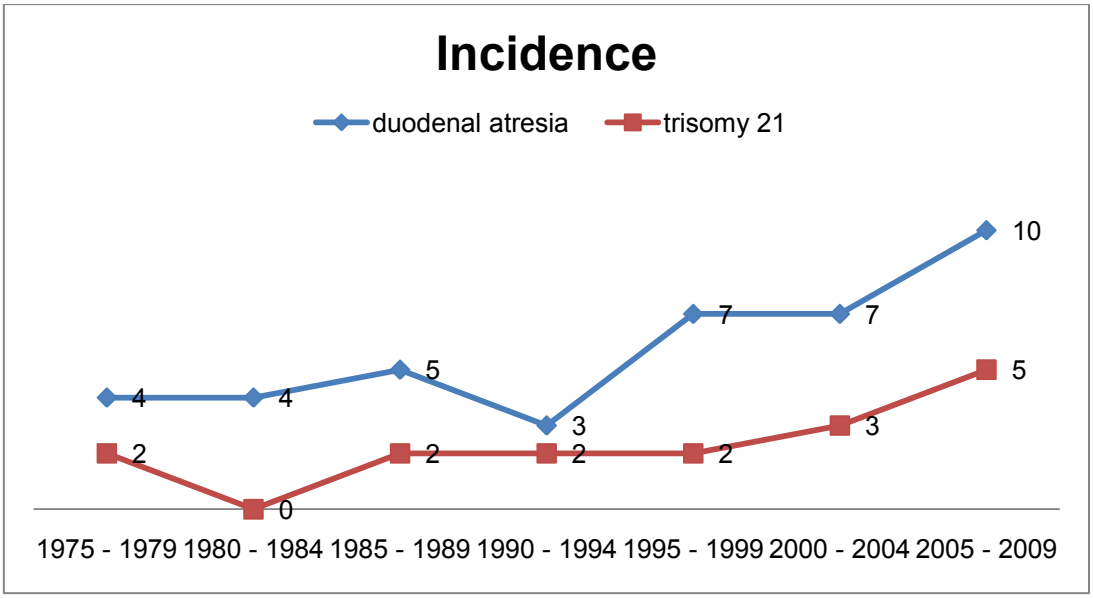


Figure 32: Incidence of duodenal atresia and trisomy 21

3.2. Prenatal period

3.2.1. Prenatal diagnosis

Prenatal care allows nowadays detecting of the majority of congenital anomalies including small bowel atresia. The following figure lists important risk circumstances which indicate or even induce this kind of anomaly.

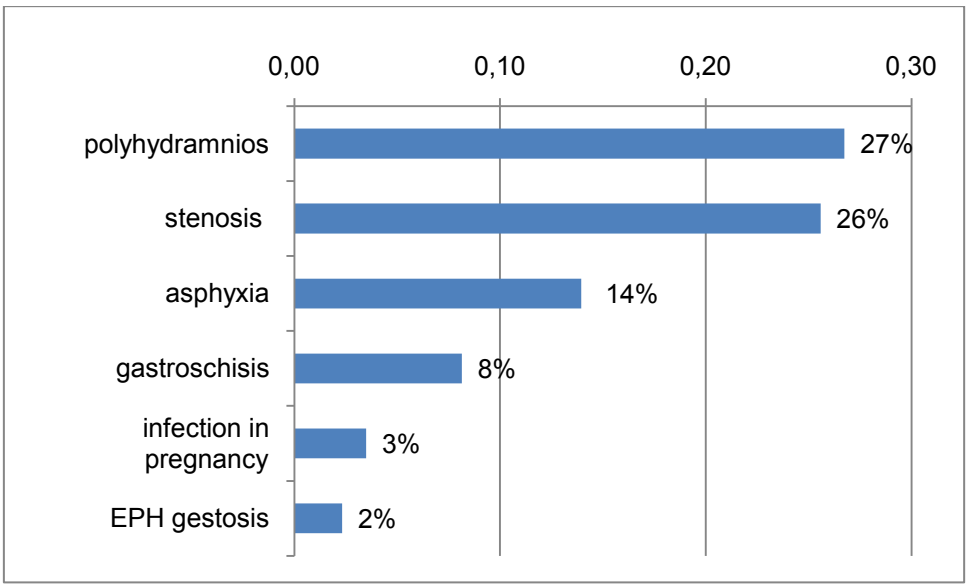


Figure 33: Prenatal diagnosis 1

Prenatal in 40% of all treated babies at Department of Pediatric and Adolescent Surgery at Medical University of Graz either polyhydramnios or stenosis were found in the ultrasound. The incidence of polyhydramnios was 27% and the incidence of stenosis 26%. Furthermore, asphyxia was diagnosed at 14%, gastroschisis at 8%, infection in pregnancy at 3% and EPH gestosis at 2%.

Figure 32 specifies important risk circumstances according to atresias.

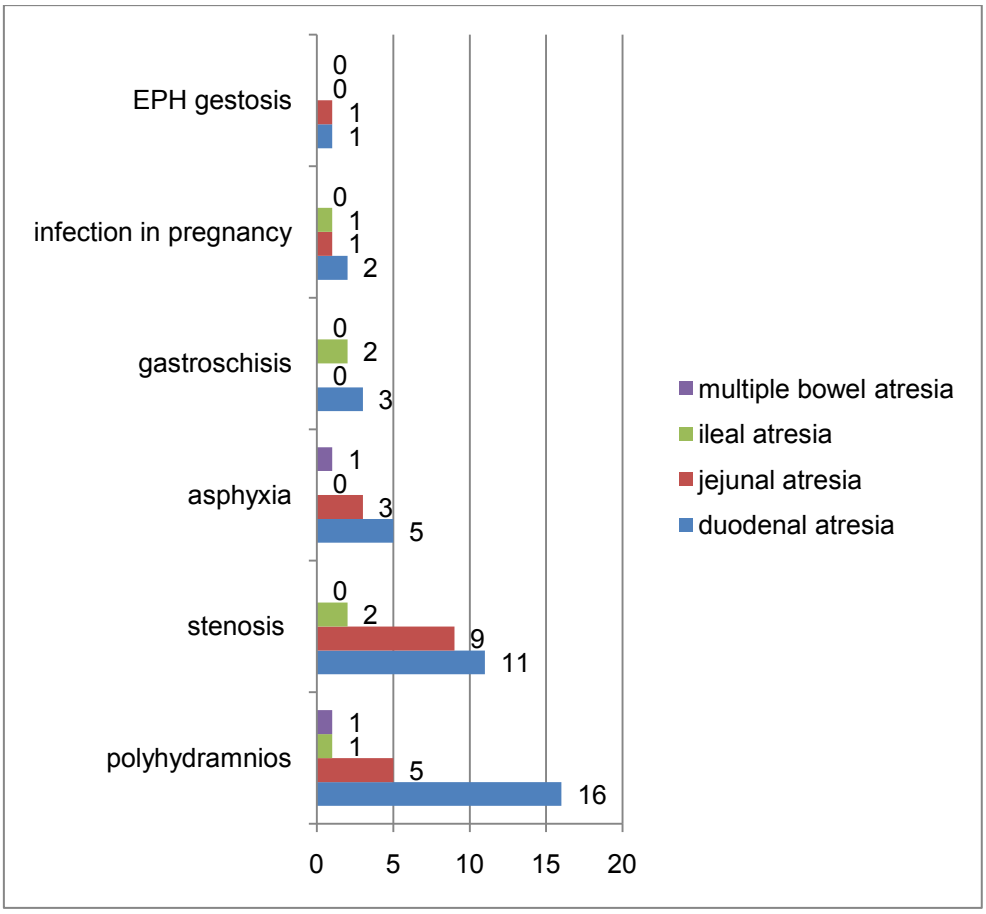


Figure 34: Prenatal diagnosis 2

In cases of duodenal atresia in 16 of 40 babies polyhydramnios was diagnosed (40%), in eleven babies duodenal stenosis (27.5%), in five babies asphyxia in the CTG (12.5%), in three babies gastroschisis (7.5%), in two babies infection in pregnancy (5%) and one baby's mother had EPH gestosis (2.5%).

In five out of 22 newborns with jejunal atresia polyhydramnios was diagnosed (22.7%), in nine infants jejunal stenosis (40.9%), in three infants asphyxia in the CTG (13.6%), in one infant infection in pregnancy (4.5%) and one infant's mother had EPH gestosis (4.5%).

Furthermore in two children of 22 with ileal atresia polyhydramnios, ileal stenosis and gastroschisis was present (9.1%), in one newborn polyhydramnios was found and one baby suffered from an infection during pregnancy (4.5%).

Finally in one infant of three with multiple small bowel atresias polyhydramnios and peripartal asphyxia was detected (33.3%).

3.3. Perinatal period

3.3.1. Amniotic fluid

Amniotic fluid consists of 99% water, lanugo hair, epithelium, uric acid, vitamins and finally infantile urine. Its normal color is clear up to light yellowish. But if the color of the amniotic fluid is green (=”missfärbig”), it indicates a stressful situation, for example an intrauterine asphyxia, with meconium discharge into the amniotic fluid.[6]

In the study 16 of 86 newborns were born with a green amniotic fluid (18.60%) and 70 of 86 newborns with a clear amniotic fluid (81.40%).

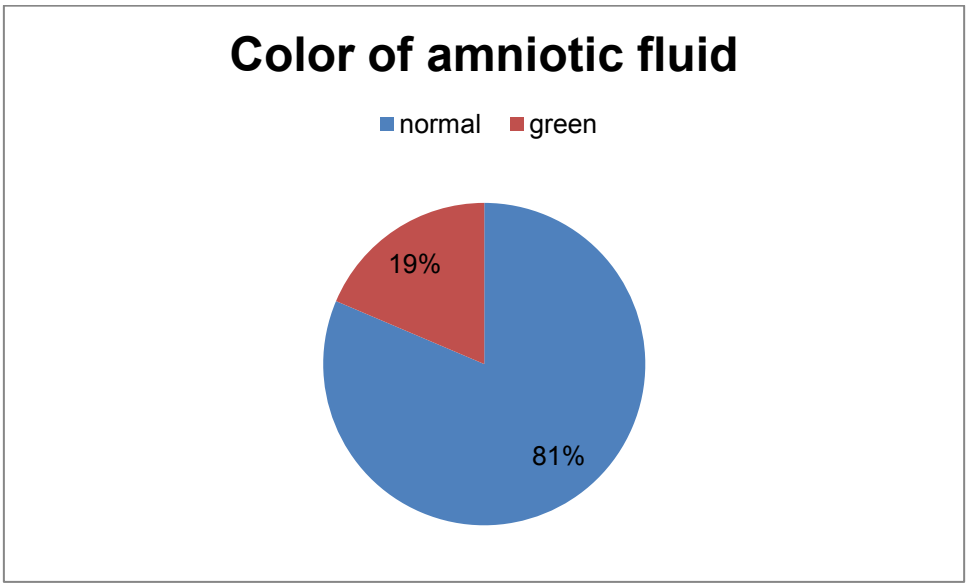


Figure 35: Color of amniotic fluid

Figure 34 shows the distribution of these 16 green amniotic fluids relating to nature of the newborns’ atresias. Five of 40 duodenal atresias had a green amniotic fluid (12.50%), seven out of 22 jejunal atresias (31.82%), three out of 22 ileal atresias (13.64%) and one of three multiple small bowel atresias (33.33%).

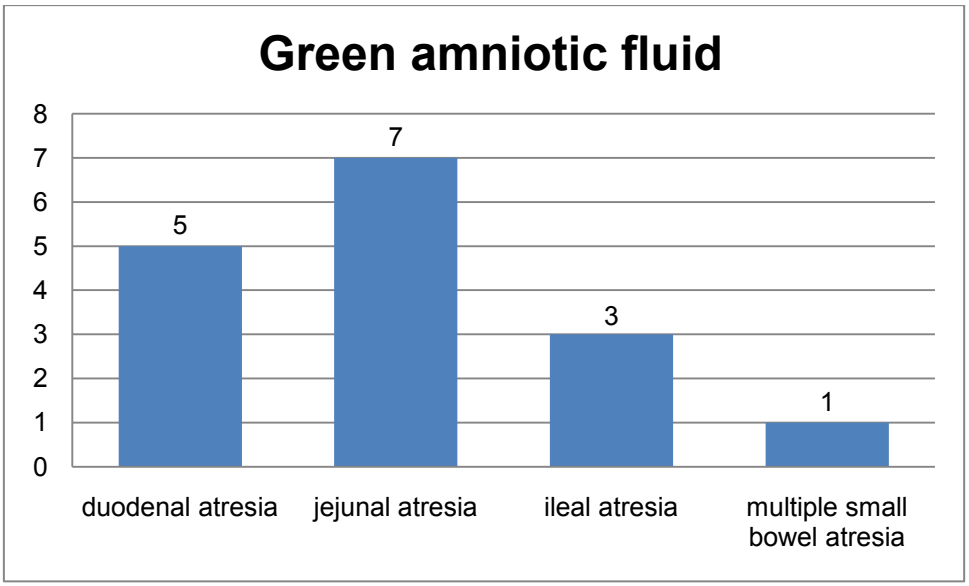


Figure 36: Distribution of green amniotic fluid

3.3.2. APGAR index

After birth, the examination of the newborn is conducted according to the schema of Virginia Apgar. The determination of the score is performed one, five and ten minutes after transection of the umbilical cord. The APGAR index consists of five criteria and for each 0-2 points are given. The summation of all five criteria result in APGAR index.[6]

Points	0	1	2
Skin color	Blue or white	Trunk auroral, extremities blue	auroral
Respiration	None	Gasping or abnormal	Normal, strong, screaming
Heart rate	None	< 100/min	> 100/min
Tonicity	Atonic	Inactive movements	Spontaneous movements
Reflex at suction	None	Grimacing	Cough, sneeze

Table 5: APGAR index

A score of 9 to 10 is ideal, 7 to 8 is within normal range, 4 to 6 indicates moderate distress and 0 to 3 serious distress, which needs resuscitation of the baby.[6]

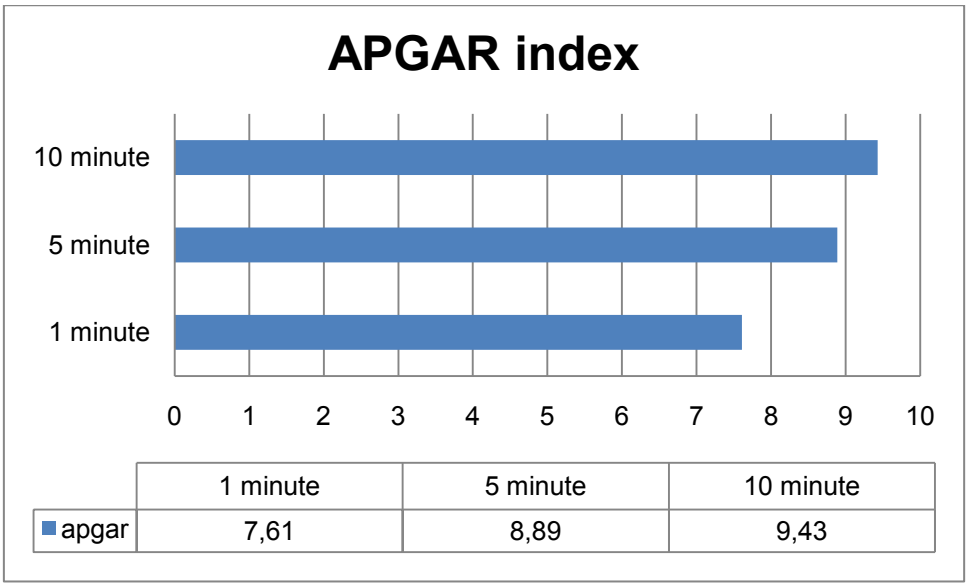


Figure 37: APGAR index

As shown in figure 35, the mean APGAR score of all newborns was 7.61 in the first minute (median 8), 8.89 after five minutes (median 9) and 9.43 after ten minutes (median 9).

3.3.3. pH-value of alantoic vein

The pH-value of the alantoic vein is a parameter to evaluate the status of the newborn. Its standard value is between 7.1 and 7.35. The newborns in this study had a mean pH value of 7.26 ± 0.14 and a median of 7.28.

3.4. Perioperative period

3.4.1. Type of operation

The choice of the operative strategy depends from the type of atresia and length of the bowel. In the 86 cases of small bowel atresia 73 end-to-end anastomosis or rather web excisions (89%), three end-to-side anastomosis (3.7%) and one side-

to-side anastomosis (1.2%) were used. Four infants were operated on in other surgical centers.

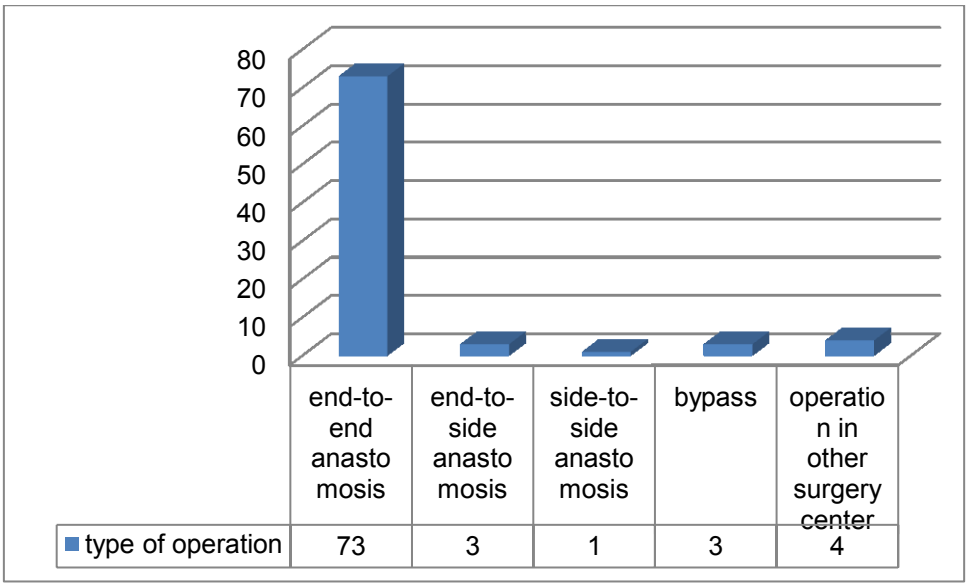


Figure 38: Type of operation

3.4.2. Date of operation

Date of operation can influence the postoperative outcome, thus the study has also investigated these dates. The mean date of operation was 2.67 days (median 2 days) and the following figure shows its distribution during the first week of life.

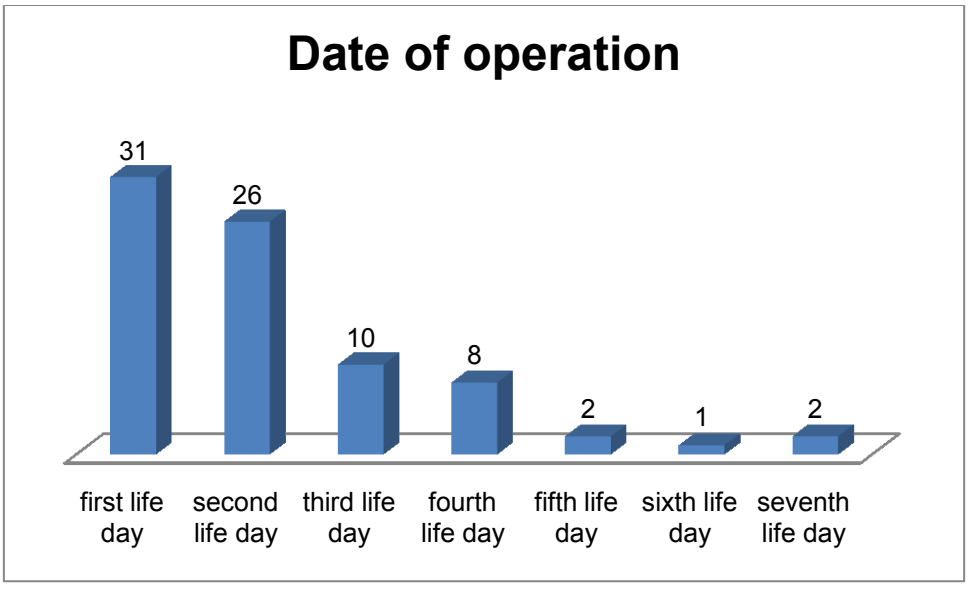


Figure 39: Date of operation

Eccept for three children, all other babies were treated during their first week of life. 31 children were operated on their first day of life, 26 on their second day of life, 10 on their third day of life, 8 on their fourth day of life, 2 either on their fifth or on their seventh day of life and one child was treated on his sixth day of life.

3.4.3. Reoperation

Altogether 25 out of 86 infants with small bowel atresias had to be reoperated (29%). Two infants suffered from an anastomotic insufficiency (2.3%), 17 infants had an anastomotic stenosis (19.8%) and six infants developed a mechanical ileus due to adhesions (6.9%).

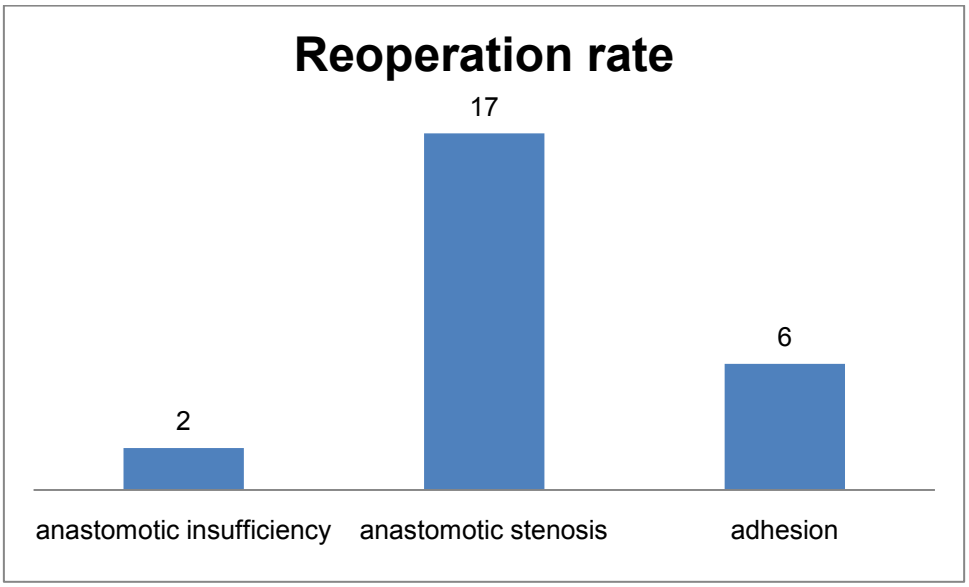


Figure 40: Reoperation rate 1

The mean gestational weight of reoperated newborns counts 2.44 kg.

Divided in particular atresia the following statement can be made.

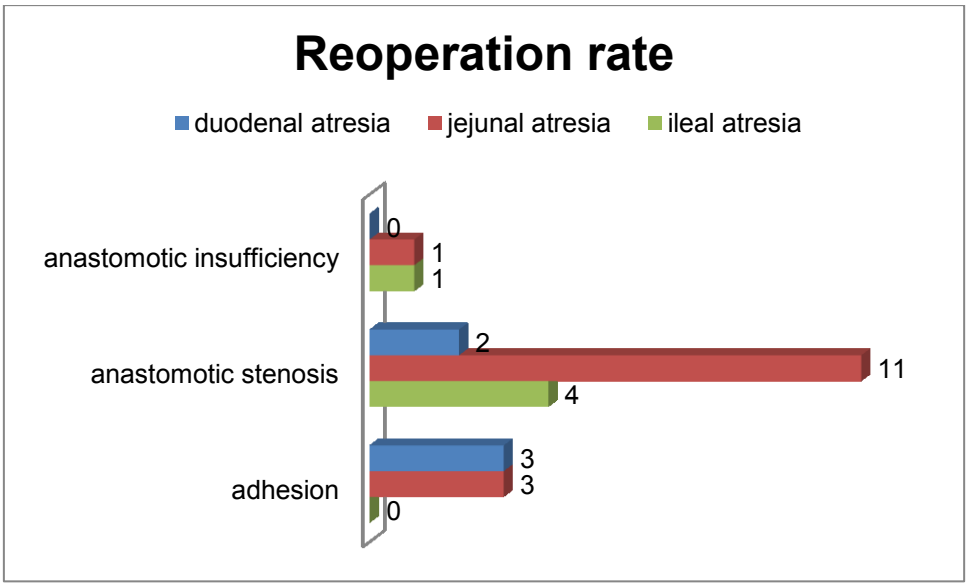


Figure 41: Reoperation rate 2

Two babies with duodenal atresia were treated because of anastomotic stenosis (5%) and three due to adhesion ileus (7.5%). Babies with jejunal atresia were treated due to an anastomotic insufficiency in one case (4.5%), eleven cases due to an anastomotic stenosis (50%) and in three cases due to adhesion ileus (13.6%). Babies with ileal atresia were treated because of anastomotic insufficiency in one case(4.5%) and in four cases due to an anastomotic stenosis (18.2%).

And finally the reoperation rate in regard to the last 35 years appears is shown in figure 40.

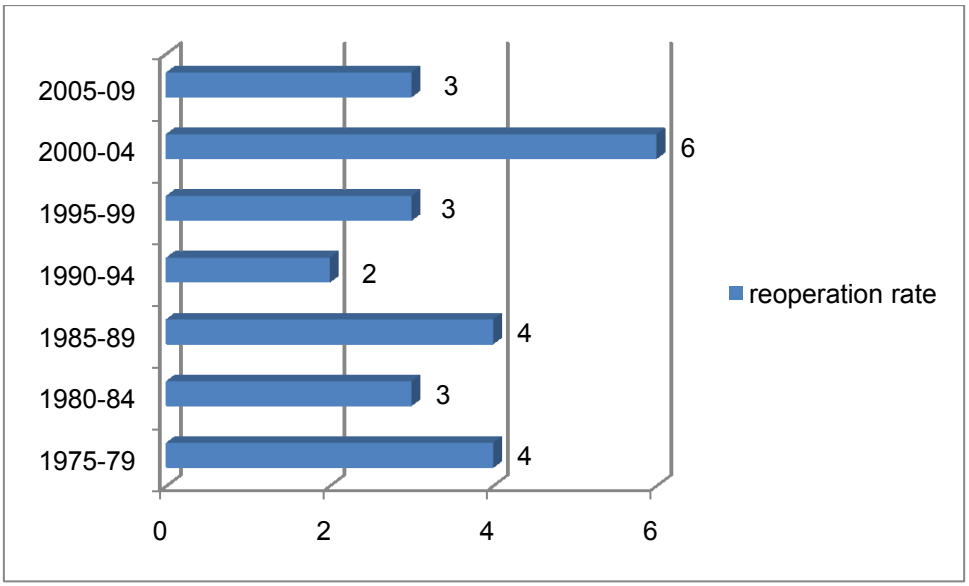


Figure 42: Reoperation rate 3

3.5. Postoperative period

3.5.1. Duration of stay in hospital

The mean duration of stay in hospital is shown in the following figure. It were 99.8 days ± 61.0 from 1975 to 1979, 80.3 days ± 67.3 from 1980 to 1984, 102.8 days ± 73.2 from 1985 to 1989, 29.5 days ± 13.8 from 1990 to 1994, 72.3 days ± 63.8 from 1995 to 1999, 52.0 days ± 44.0 from 2000 to 2004 and 54.1 days ± 31.8 from 2005 to 2009. The trend line shows a clear decline over the last 35 years.

All in all was the median of the duration of stay in hospital 55 days, the average 72 days ± 59.9.

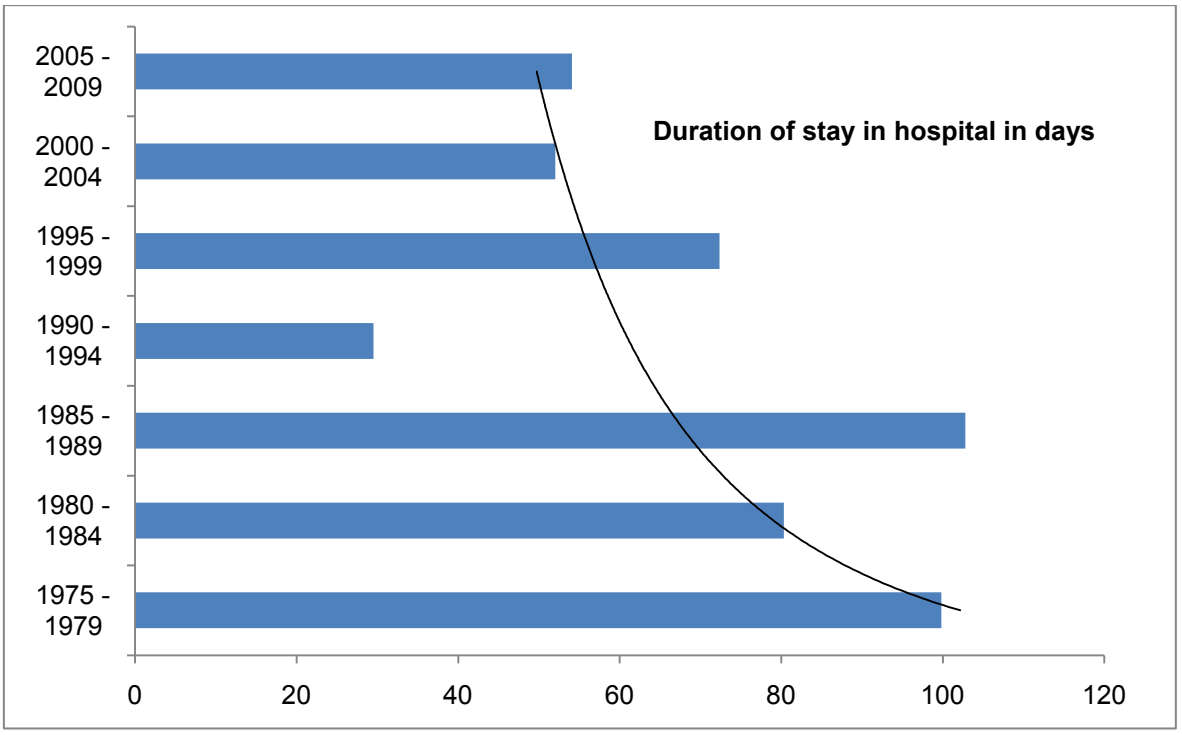


Figure 43: Duration of stay in hospital 1

The following figure illustrates the mean duration of stay during the last 35 years.

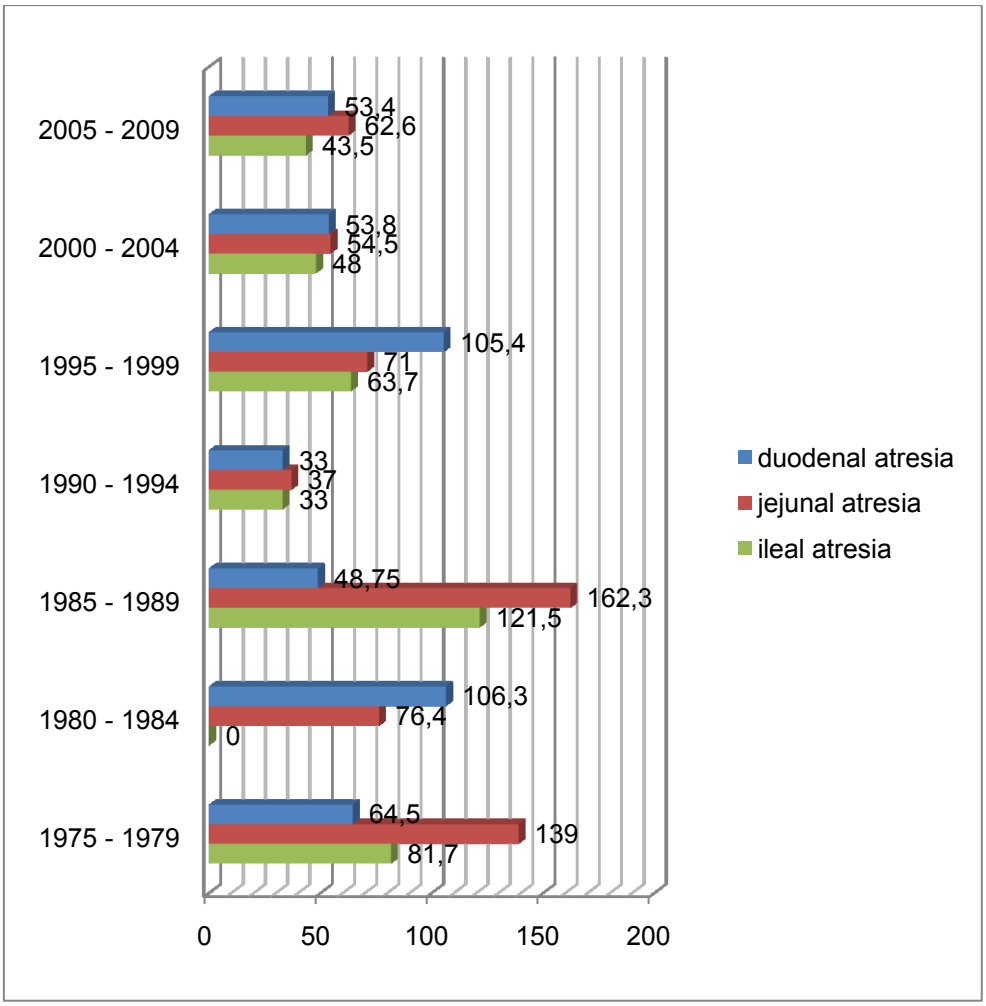


Figure 44: Duration of stay in hospital 2

From 1975 to 1979 patients with duodenal atresia were 64.5 days in hospital, with jejunal 139 days and with ileal 81.7 days, from 1980 to 1984 with duodenal atresia 106.3 days and with jejunal 76.4 days, from 1985 to 1989 with duodenal atresia 48.8 days, with jejunal 162.3 and with ileal 121.5 days, from 1990 to 1994 with duodenal atresia 33 days, with jejunal 37 and with ileal 33 days , from 1995 to 1999 with duodenal atresia 105.4 days, with jejunal atresia 71 and with ileal 63.7 days, from 2000 to 2004 with duodenal atresia 53.8 days, with jejunal 54.5 and with ileal 48 days and from 2005 to 2009 with duodenal atresia 53.4 days, with jejunal 62.6 and with ileal 43.5 days.

3.5.2. Duration of stay in intensive care unit (ICU)

Retrospectively, figure 43 illustrates the mean duration of stay in ICU of studied babys with small bowel atresia. From 1975 to 1979 it were at 79.50 days, from 1980 to 1984 at 80.25 days, from 1985 to 1989 at 78.56 days, from 1990 to 1994 at 21.83 days, from 1995 to 1999 at 56.17 days, from 2000 to 2004 at 43.58 days and from 2005 to 2009 at 45.58 days. Again the trend line shows a decrease of stay in the ICU over the last 35 years.

Altogether the median of the duration of stay in intensive care accounted 34 days and the average 58 days.

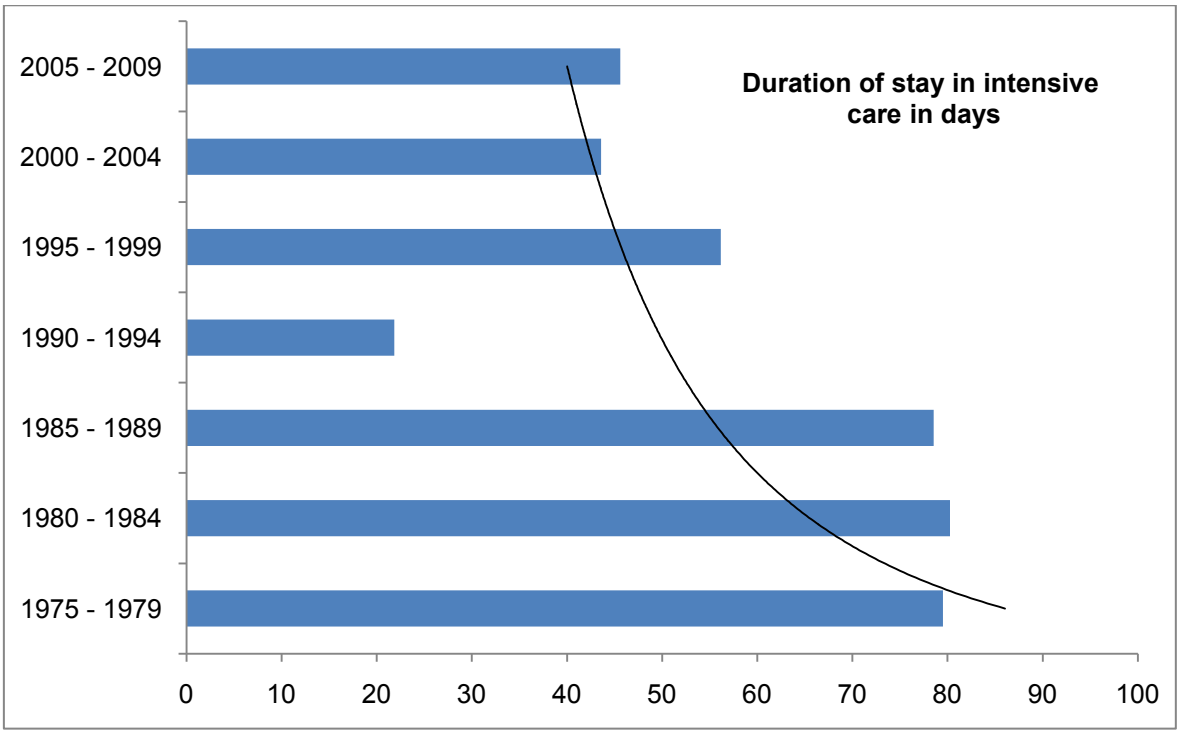


Figure 45: Duration of stay in ICU

3.5.3. Duration of ventilation

The following figure shows the mean duration of ventilation after surgery. From 1975 to 1979 it accounted 2.00 days, from 1980 to 1984 2.63 days, from 1985 to 1989 3.50 days, from 1990 to 1994 5.22 days, from 1995 to 1999 3.17 days, from

2000 to 2004 2.69 days and from 2005 to 2009 3.14 days. Finally the median duration of ventilation was 2 days and the average 3 days.

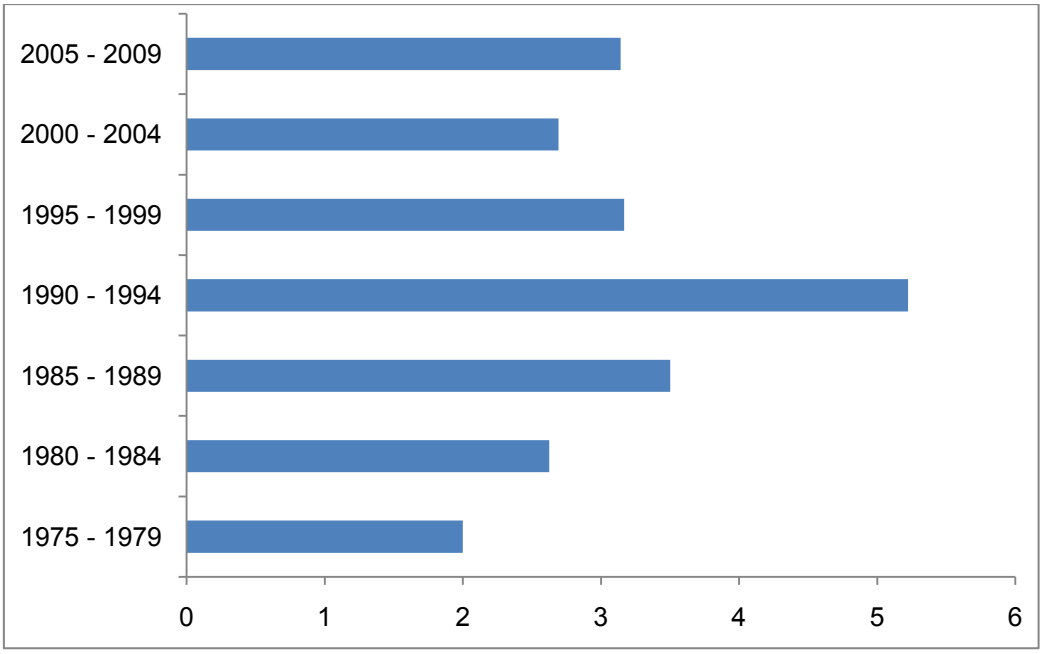


Figure 46: Duration of ventilation

3.5.4. Oral feeding

Oral feeding is essential for a newborn, which requires 120 calories/day. Therefore the study explored the mean onset of oral feeding with 13.00 days (median 13) and the mean date of total oral feeding with 54.00 days (median 54).

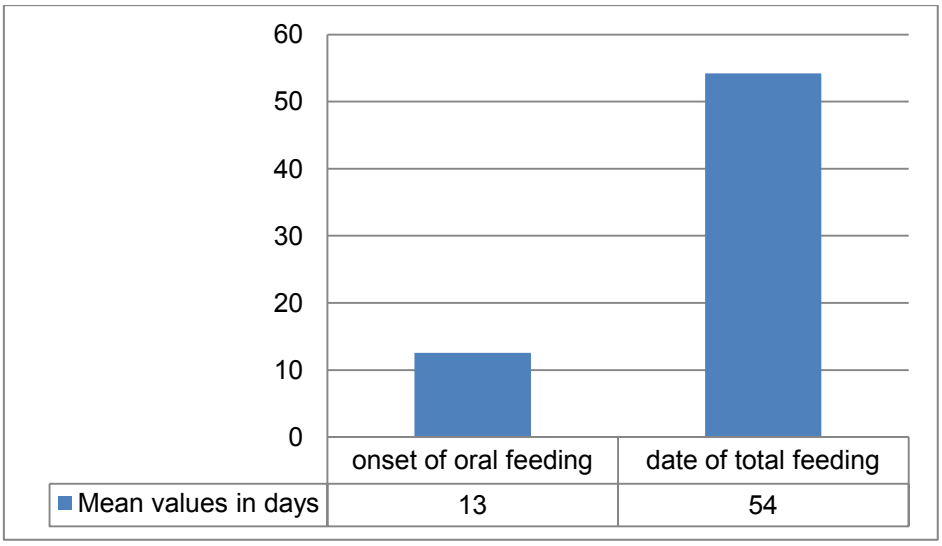


Figure 47: Oral feeding 1

The figure shows a significant development versus earlier onset of oral feeding and total oral nutrition in the study periods.

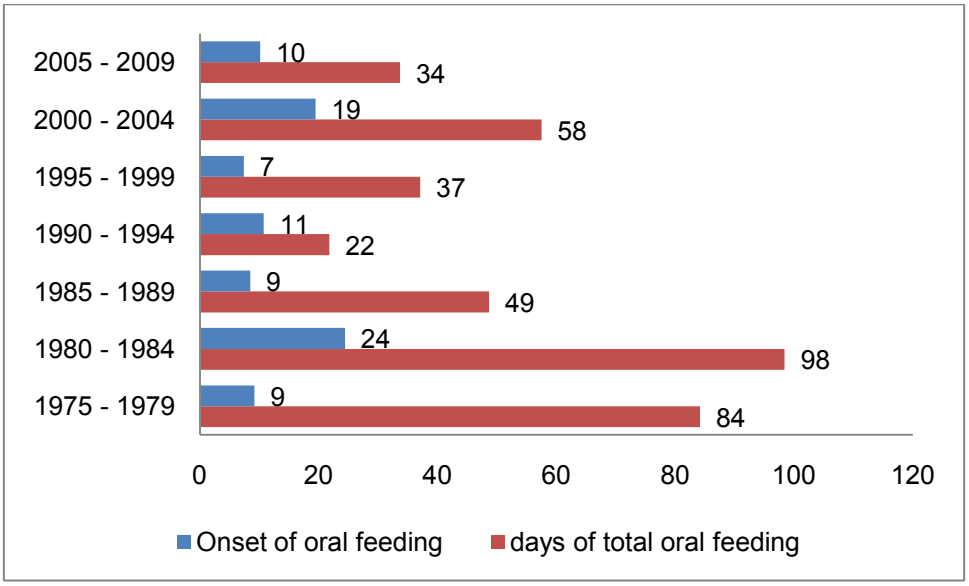


Figure 48: Oral feeding 2

3.6. Complications

3.6.1. Early complications

The following figure shows the patients with anastomotic stenosis (19.8%), ileus/subileus symptomatology and sepsis each (16.3%) and anastomotic stenosis, short bowel syndrome and burst abdomen each (2.3%).

Ten babies with ileus/subileus symptomatology had to be reoperated.

The newborns with early complications had on average 2.49 kg (mean gestational weight of all treated babies is 2.49 kg) and were born with a gestational age of 34.47 weeks (mean gestational age of all treated babies is 36.38 weeks).

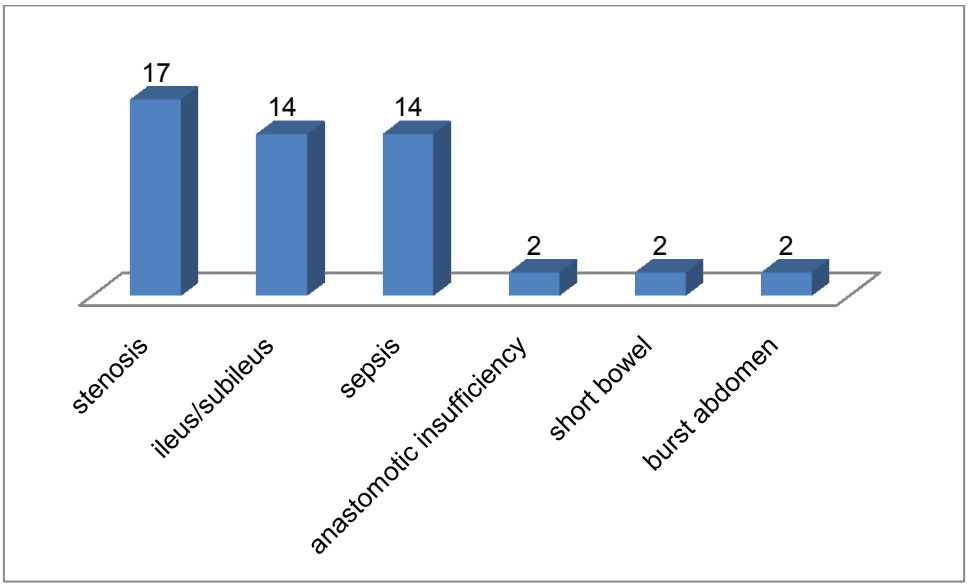


Figure 49: Early complications 1

In premature infants (gestational age < 37. gestational week) stenosis was diagnosed in 15.1%, ileus/subileus symptomatology in 9.3%, sepsis in 11.6%, and anastomotic stenosis, short bowel syndrome and burst abdomen each with 2.3%.

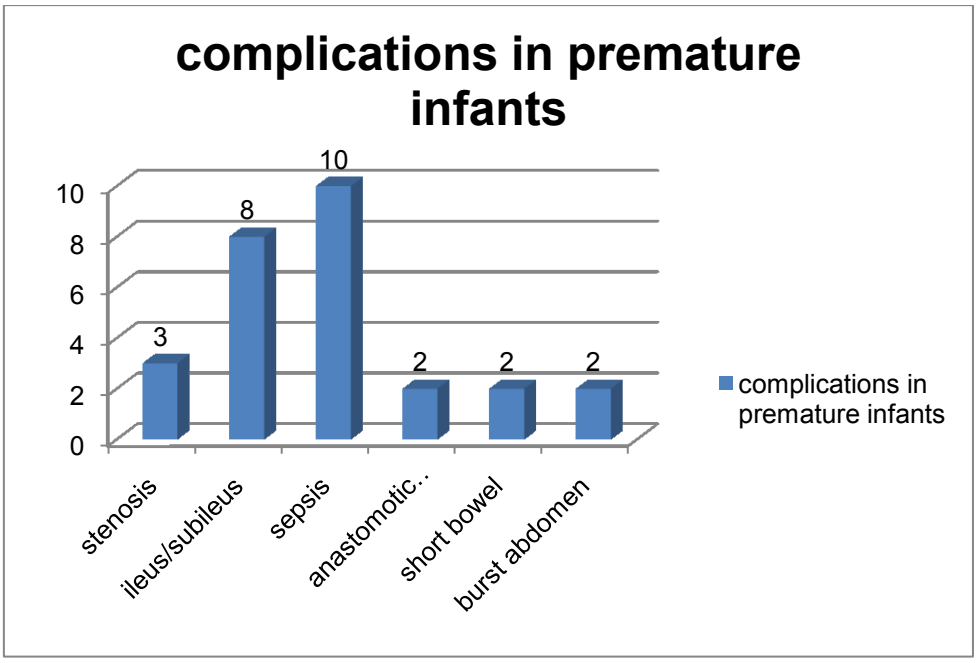


Figure 50 Early complications in premature infants

In duodenal the ileus/subileus and sepsis rate was 7.5% and the short bowel rate 2.5%. While in jejunal atresias the stenosis rate was 23%, the ileus/subileus rate 50%, the sepsis rate 27%, the anastomotic insuffiency and short bowel rate 9%. And finally in ileal atresias the stenosis, ileus/subileus and sepsis rate was 14% and the anastomotic insufficiency and short bowel rate was 4.5%.

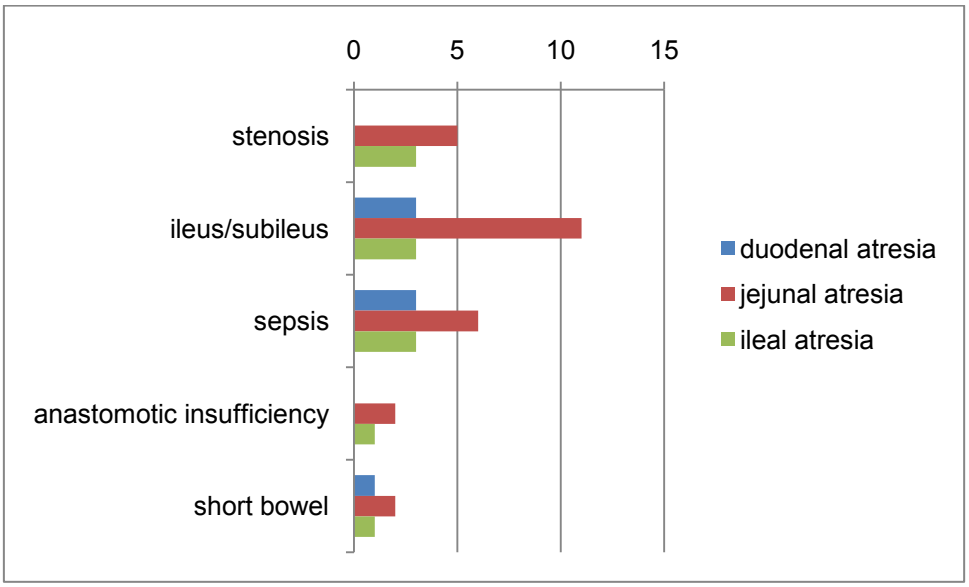


Figure 51: Early complications 2

3.6.2. Late complications

The somatogramm is a table with standard values of weight and seize according to age. Insufficient growth and dystrophy can be detected with the somatogramm.

In our study infants with small bowel atresia were examined after 3, 6, 9, 12, 18 and 24 months. Seize persisted till the 24th month on the 25th percentile and weight till 18th month on the 10th percentile but increased at 24 months to the 25th percentile.

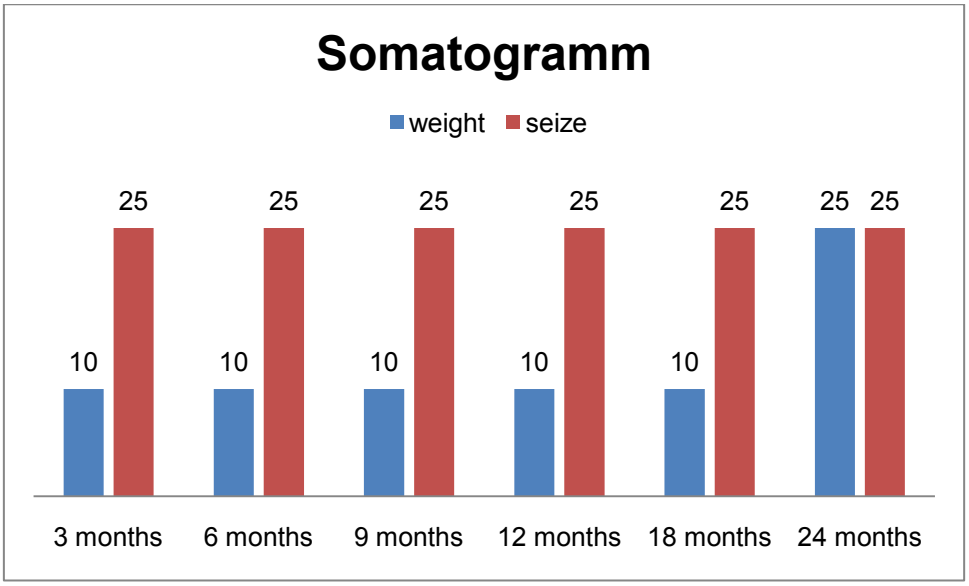


Figure 52: Somatogramm

3.6.2.1. Outpatient visits

36 of the 86 treated infants (41.9%) came on an average of 9.25 times in the outpatient clinic of the Department with symptoms of

- Recurrent vomiting
- Stomach ache/colic
- Obstipation

or needed an additional feeding by gastric tube.

Of these 36 patients 19 had a duodenal atresia (47.5%), nine had a jejunal atresia (41%), six had an ileal atresia (27%) and two had multiple atresias (67%).

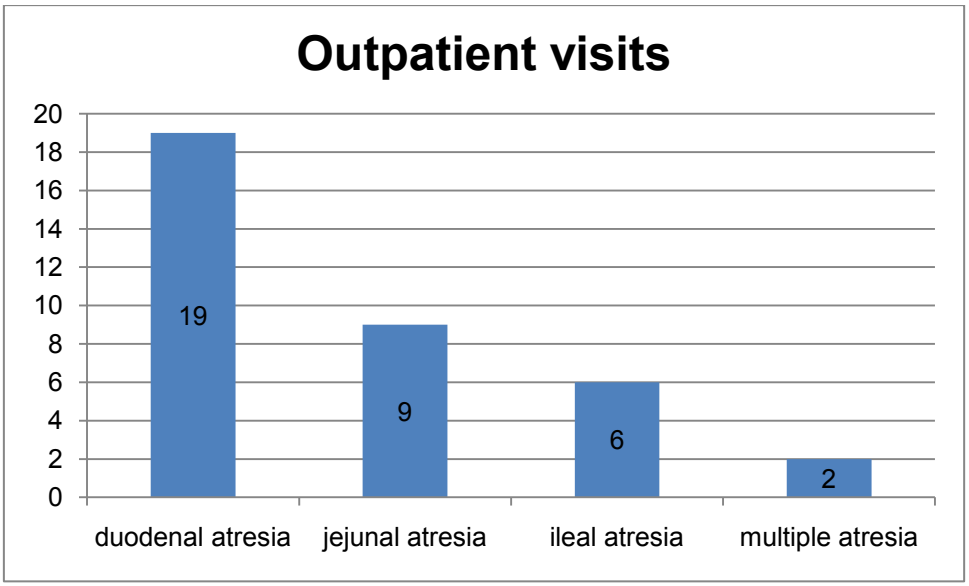


Figure 53: Number of outpatient visits

Next figure shows the distribution of outpatient visits in relation to the gestational age. Except newborns with 3 months, all other babies were rather mature than premature.

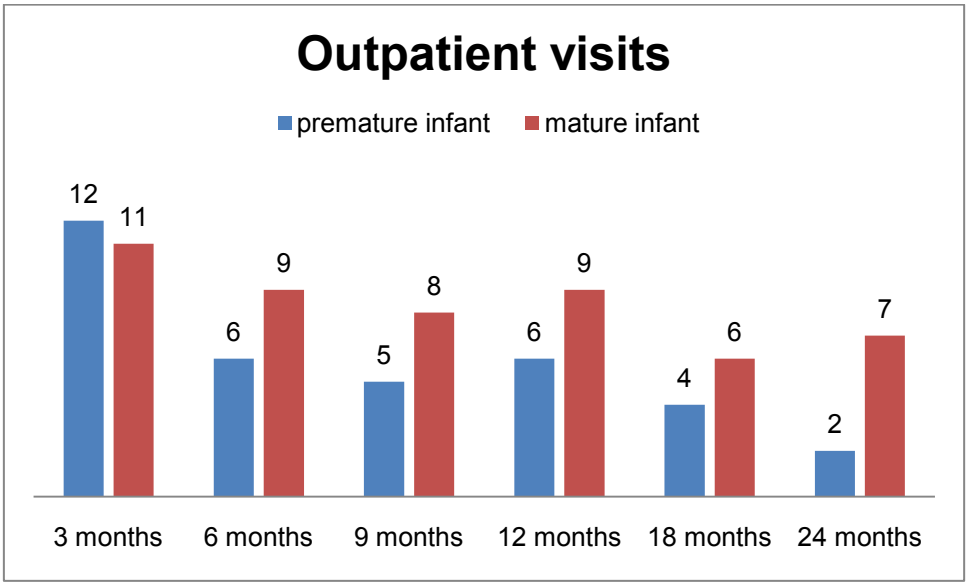
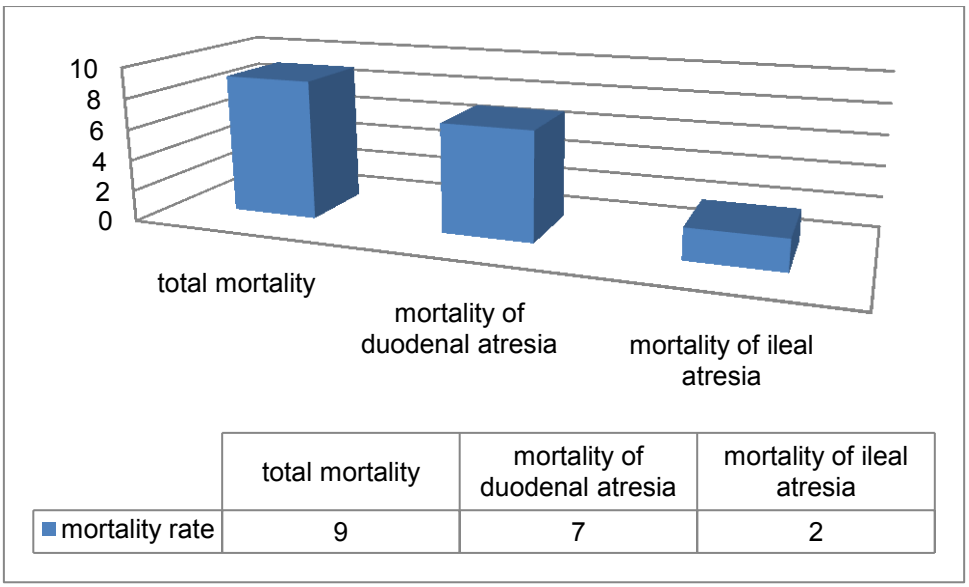


Figure 54: Distribution of outpatient visits in relation to the gestational age

3.6.3. Mortality

The absolute mortality rate accounts 9 of 86 infants (10.5%). Of these, seven infants had a duodenal atresia (17.5%) and two infants had a ileal atresia (9.1%). Six of them were boys and three were girls.



	total mortality	mortality of duodenal atresia	mortality of ileal atresia
mortality rate	9	7	2

Figure 55: Mortality rate

7 of these 9 children had associated anomalies; two of them had trisomy 21, two gastroschisis, two esophageal atresia, 2 malrotation, one short bowel syndrome and one liver and kidney failure.

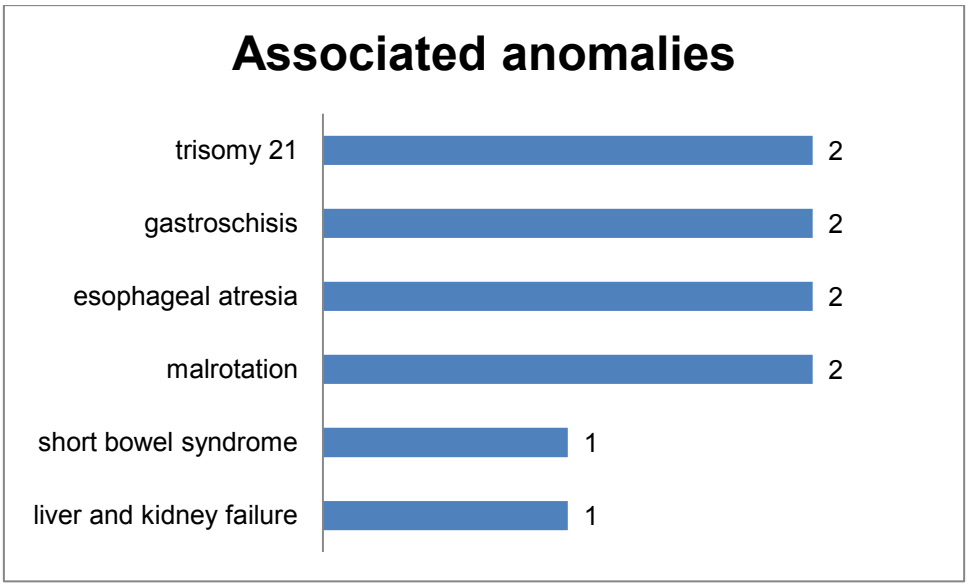


Figure 56: List of associated anomalies in dead infants

Four infants died during the first week, three died during the first month and two died during the first six month.

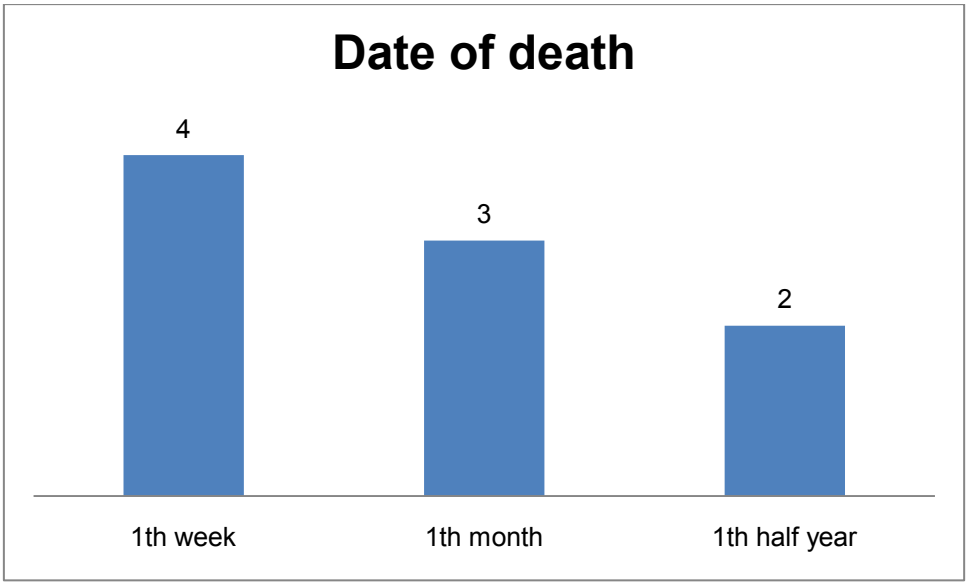


Figure 57: Date of death

On average all nine babies had a gestational age of 35.6 weeks and a gestational weight of 2061 g. 6 of the nine were premature and 5 of the nine weighed under 2000 g.

Moreover five of these nine infants were operated on their first day of life, three on their second day of life and one infant was treated on its sixth day of life.

Except one child, in all children polyhydramnios, gastroschisis or bowel stenosis was detected by ultrasound and their mothers had an infection during pregnancy or complication during the birth.

The death causes were sepsis, inoperable congenital heart disease and short bowel syndrome with liver insufficiency.

For the last 35 years, the mortality rate remained constant with two dead infants from 1975 to 1979, one infant from 1980 to 1984, 1985 to 1989, from 1990 to 1994 and 2005 to 2009 and three infants from 1995 to 1999.

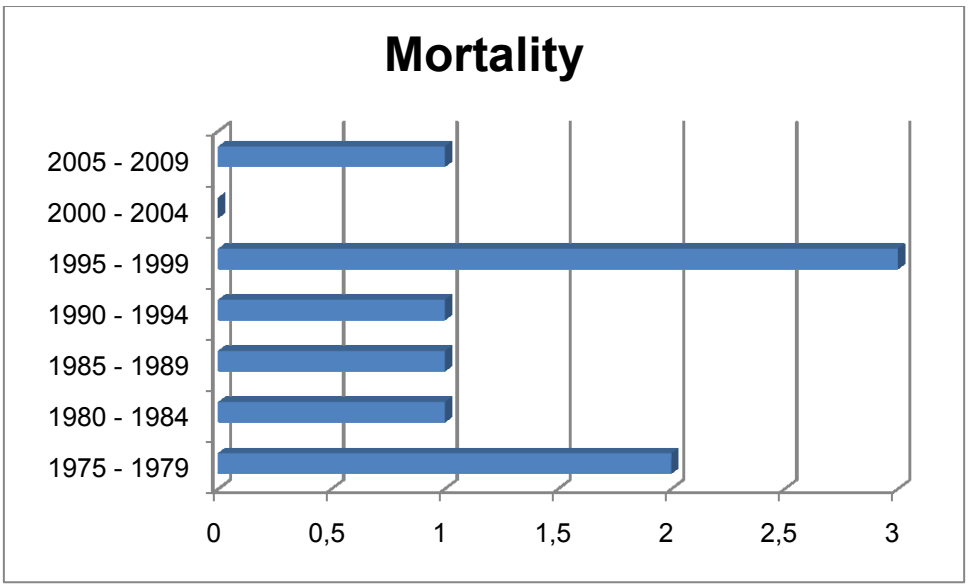


Figure 58: Mortality rate during the last 35 years

The following figure shows key data of each dead baby:

	1	2	3	4	5	6	7	8	9
Year of birth	1979	1979	1981	1989	1991	1996	1997	1998	2005
Weight at birth (g)	3200	1570	2900	980	1890	1720	1400	2070	2822
Gestational age (weeks)	40	34	40	35	35	32	32	35	38
Location of atresia	duodenal	duodenal	duodenal	ileal	ileal	duodenal	duodenal	duodenal	duodenal
Assoc. anomalies	Complex cardiac defect			Esophage-al atresia, PDB	Gastroschi-sis, lung hypoplasia	Trisomy 21	Gastroschi-sis, short bowel syndrome	Short bowel syndrome	Trisomy 21, esophageal atresia
Complications	Twisted cord, asphyxia	Sepsis, cardio vascular collapse	Sepsis, liver and kidney failure	IRDS	Pneumo-thorax	Amniotic infection syndrome	Sepsis, peritonitis, intracere-bral hemorra-ghe	Cholestasis, liver failure	pneumonia
1st operation (day of life)	6	3	2	2	1	2	1	1	1
Day of death (day of life)	22	17	36	4	4	7	8	164	135

Table 6: Key data of each baby

4. Discussion

This study shows that the incidence of small bowel atresia increased steadily during the last 15 years. While the number of cases with jejunal and ileal atresias remained constant during the last 35 years, the number of duodenal atresias increased during the last 20 years from 3 cases from 1990 to 1994 to 7 cases in a period of 1995-1999 and 2000-2004 to reach the highest number with 10 cases from 2005 to 2009. A possible explanation for such a development could be the new awareness in prenatal diagnosis, improved perinatal management and neonatal intensive care, which led to the survival of children with a lot of comorbidity and associated anomalies.

Compared to other studies our result of incidence with duodenal atresia : jejunoileal atresia = 1 : 1.15 was similar to values of 1.05 : 1 in [7], 1 : 1.2 in [8] and 1.08 : 1 in [9].

In the course of this study we noticed a prevalence of the location of duodenal atresia in pars descendens (60%), which can be explained by the higher rate of ceses with pancreas annulare (40%). If one deducts the cases with pancreas annulare from the cases with an atresia in the pars descendens 8 cases remain, a number close to the cases with distal atresia (10) and proximal atresia (6). In addition, a higher rate of proximal jejunal atresias occured while distal, middle and multiple locations with 4, 6 and 6 cases are rare. Finally a higher rate of distal ileal atresias was registered, although the proximal, middle and multiple locations were again analogical.

In regard to the different types of small bowel atresia studies and references report many variations. This could be due to a different use of classification of each surgeon. On the one hand almost all references agree about the rate of stenosis caused by pancreas annulare. For example, there are 39% in [10] and 33% in [9]. These values are confirmed by the 40% in our study. On the other hand there are controversial data about type 1, 2 and 3 atresias of the duodenum. In the study we counted the rate of type 1 atresia up to 52.5%, while in [10] the rate was 19.5% and in [5] it was 92%. The rate of type 2 and 3 was in our study 7.5% versus 41.6% in [10] and 8% in [5].

The 10.5 % mortality rate (9 of 86 infants) is distributed to 7 infants(17.5%) with duodenal atresia and 2 infants (9.1%) with ileal atresia. These numbers are similar to [9] with 20% and [7] with 7%. There were no surgical complications, which led to death. Six of them were premature and five weighed under 2000g. Furthermore seven of these children had following associated anomalies: trisomy 21, gastroschisis, esophageal atresia, malrotation, short bowel syndrome, congenital heart disease and liver and kidney failure. Four infants died during the first week, three died during the first month and two died during the first 6 months. Moreover five of these nine infants were operated on their first day of life, three on their second day of life and one infant was treated on its sixth day of life, for this reason a delayed in diagnosis can be excluded. Except one child, in all newborn either a prenatal polyhydramnios or a gastroschisis or a bowel stenosis was recorded or their mothers suffered from an infection during pregnancy or an periportal asphyxia. The final cause of death in 4 infants was sepsis. Further causes are inoperable congenital heart disease and short bowel syndrome. With regards to the last 35 years, the mortality rate remained constant with a median of 2 dead infants in a decade.

In our study the most frequent associated anomalies were of gastrointestinal nature, which had more than the half of the children. Further impressive data were, that infants with duodenal atresia had at 80% associated anomalies including 43% trisomy 21, 40% pancreas annulare and 38% congenital heart disease. Moreover in infants with jejunal atresia mucoviscidosis was found in 23%. These above data correlate with other studies (9,10).

Considering the relationship between duodenal atresia and trisomy 21, the incidence of both increased correlatively during the last 35 years, these results are identical to [12], in which an overall increased association of duodenal atresia with trisomy 21 is shown.

In 40% of all newborns polyhydramnios and stenosis were found in the prenatal ultrasound, however these children did not have a better prognosis compared to the other newborns. In cases of duodenal atresia in 16 of the 40 babies polyhydramnios was diagnosed (40%), while in five infants of 22 with jejunal

atresia (22.7%) and in just one child of 22 with ileal atresia a polyhydramnios was diagnosed (4.5%). These numbers are similar as in other studies (12,13).

With regard to sex ratio the study showed male : female ratio of 1.15 :1. The sex ratio of jejunoileal atresias with male : female = 1.3 :1 is higher.

In regard to gestational age we found avarage gestational age of 36.38 weeks, whereat 43% were premature infants and 20% weighed under 2000g. Furthermore the study shows decreasing gestational age. The causes of this development could be new awareness in prenatal diagnosis with the consequence of a higher rate of cesarean deliveries.

In 19% of our patients with small bowel atresia the amniotic fluid was green as a sign of stressful situation. Remarkably, the rate in jejunal atresia patients was 32%. However the APGAR index and the pH-value of alantoic vein showed normal data.

73 end-to-end anastomosis or rather web excisions (89%), three end-to-side anastomosis (3.7%) and one side-to-side anastomosis (1.2%) were performed. These data are similar as in other reports (8,14). Eccept of three babies, all other neonates were treated during their first week of life. The median of the mean date of operation accounts two days.

Altogether 29% of infants with small bowel atresias had to be reoperated. Of these children 2.3% were treated due to an anastomotic insufficiency, 19.8% of infants due to an anastomotic stenosis and 6.9% of infants due to adhesions. Remarkably, in 50% infants with jejunal atresia had to be treated due to an anastomotic stenosis in eleven cases. The mean gestational weight of these reoperated babies is 2.44 kg, that’s nearly as much as the average of gestational weight of all treated babies (2.49 kg).

Moreover in our study complications were analyzed at 19.8% anastomotic stenosis and 16.3% had ileus/subileus or sepsis. In jejunal atresias the stenosis rate was 23%, the ileus/subileus rate 50%, the sepsis rate 27%, the anastomotic insuffiency and short bowel rate 9%. Ten babies of 14 with ileus/subileus symtomatology had to be reoperated. The mean gestational weight of the reoperated newborns counts 2.49 kg, that’s exactly the same average value in our study and the mean

gestational age of the reoperated babies is 34.47 weeks. According to that, the reoperated babies were in average two weeks younger than all treated newborns in the study with 36.38 weeks.

While the duration of stay in hospital and intensive care decreased during the last 35 year from 100 or rather 80 days to 54 or rather 46 days, the duration of ventilation kept constant. This is explainable by improved neonatal intensive care and surgical techniques. Furthermore there's no correlation between the location of the atresia and the duration of stay in hospital and in intensive care and ventilation.

In terms of oral feeding we found out that the mean onset of oral feeding remained constant during the last three decades, but the mean date of total oral feeding declined dramatically from 84 days to 34 days. That's a difference of 50 days.

In somatogramm all examined children (42%) had a constant low weight (10) and low seize percentile (25). These children came during their first two years of life on average 9 times in ambulance with symptomatology of gastrointestinal nature. And of these 36 patients 19 had a duodenal atresia (47.5%), nine had a jejunal atresia (41%), six had an ileal atresia (27%) and two had multiple atresias (67%).

The basic limitation is the retrospective character of this study with some missing data. Furthermore the partially incomplete patients' documentation was another limitedly factor.

5. Conclusion

In our study we analyzed 86 cases of small bowel atresia including duodenal atresia, jejunal atresia and ileal atresia.

Incidence of small bowel atresia increased steadily during the last 15 years, especially the incidence of duodenal atresia and its associated trisomy 21.

With regard to sex ratio the study showed male : female ratio of 1.15 :1. The sex ratio of jejunoileal atresias with male : female = 1.3 :1 is higher.

The avarage gestational age was 36.38 weeks, whereat 43% were premature infants and 20% weighed under 2000g. In summery that's a decreasing development during the last 35 years.

In 89 % end-to-end anastomosis or rather web excisions were performed and except of three babies, all other neonates were treated during their first week of life, the median accounts two days.

Altogether 29% of infants with small bowel atresias had to be reoperated. Remarkably, that the mean gestational weight of reoperated babies isn't dependent on reoperation rate, indeed the newborns were in average two weeks younger than all treated babies in the study.

The mortality rate during the last 35 years remained constant with a median of two dead infants in a decade. Overall mortality is up to premature birth, associated anomalies and location of atresia and isn't dependent on delayed diagnosis, prenatal diagnosis and trisomy 21. The leading cause of death was sepsis, congenital heart disease and short bowel syndrome.

In our study the most frequent associated anomalies were of gastrointestinal nature, which had more than the half of the children. Newborns with duodenal atresia had at 80% associated anomalies including 43% trisomy 21, 40% pancreas annulare and 38% congenital heart disease.

There is no correlation between postoperative outcome and location of atresia.

The duration of stay in hospital and intensive care decreased during the last 35 years to the half and the mean date of total oral feeding declined dramatically from 84 days to 34 days.

Infants with jejunal atresia have earlier complications than infants with other small bowel atresias, while infants with duodenal and multiple atresias have more long-term complications, which are higher in general than described in literature. More than one third children with congenital small bowel atresias have during their first two years of life gastrointestinal problems in form of ileus/subileus symptomatology and low weight percentile.

Today overall survival rate of newborns with small bowel atresias is about 90%.

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