Uncommon surgical emergencies in neonatology

Emergenze chirurgiche rare in epoca neonatale

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Abstract

Objective. Over the past decade, multiple factors have changed the pattern of neonatal surgical emergencies. An increase in prenatal screenings and the development of neonatal tertiary care centres have changed the clinical approach to these kids.
Materials and methods. Between 1995 to 2011 were retrospectively reviewed 34 patients with diagnosis of uncommon rare neonatal surgical emergencies at our institute. We analyzed: sex, gestational age, weight at birth, primary pathology, prenatal diagnosis, associated anomalies, age and weight at surgery, clinical presentation, start of oral feeding and hospitalization. The follow-up was performed at 6,12, 24 and 36 months.
Results. There were 21 male and 13 female. The gestational age ranged between 28 and 36 weeks. The weight at birth ranged between 700 and 1400 grams. Oral feeding was started between 4th and 10th postoperative day. The average hospitalization was about 70.47 days. To date, all patients have finished the follow-up. They are healthy.
Conclusion. The outcome of the patients with uncommon surgical emergencies is different based on the etiology. Overall survival is generally good but is influenced by the associated anomalies.

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Introduction

Over the past decade, multiple factors have changed the pattern of neonatal surgical emergencies. An increase in prenatal screenings and the development of neonatal tertiary care centres have changed the clinical approach to these kinds of emergencies. In fact, most conditions leading to a gastrointestinal (GI) emergencies, are uncommon and treatment in specialist centres enables concentration of appropriate resources and expertise. Co-morbidity is common, particularly in the preterm or low birth weight infant.[1]
Symptoms of GI emergencies may be subtle, including irritability or feeding intolerance, or they may be more apparent, with vomiting (bilious or non bilious), abdominal distension, and shock. Vomiting in the neonatal period should always prompt the consideration of a pathologic process. It may be difficult to differentiate a life-threatening cause from a mild viral gastroenteritis or even severe gastroesophageal reflux. The initial symptoms may be nonspecific and the history may not be helpful in a neonate who has not developed a normal pattern. Early diagnosis, availability of diagnostic service and prompt surgical intervention with optimal pre- and post-operative care are necessary to increase survival of newborns with such problems.[2]
Table 1 reports a list of principal causes of neonatal surgical emergencies of the GI tract, divided into common and uncommon, performed according to the current literature.
We report our experience with these pathologies.

Materials and Methods

This is a retrospective study of 34 patients admitted to our tertiary care institute (Pediatric Surgery Unit of Siena) between 1995 and 2011 with diagnosis of uncommon rare neonatal surgical emergencies. We did not consider the common emergencies. We analyzed: sex, gestational age, weight at birth, primary pathology, prenatal diagnosis, associated anomalies, age and weight at surgery, postnatal diagnosis, clinical presentation, start of oral feeding and hospi-

talization. The follow-up was performed at 6, 12, 24 and 36 months after surgery. It provided a clinical evaluation and a questionnaire to the parents about growth, weight, type and frequency of alvus, presence of aerophagia, meteorism and/or rectal bleeding, type of feeding, presence of vomit, excessive belching, and/or hematemesis.

Results

There were a total of 34 patients. 21 (61,8%) were male and 13 (38,2%) female. The gestational age ranged between 28 and 36 weeks. The weight at the birth ranged between 700 and 1400 grams.

The diseases diagnosed were: 6 (17,6%) spontaneous gastric perforation (5 male, 1 female), 8 (23,6%) spontaneous intestinal perforation (5 male, 3 female), 2 (5,9%) gastric duplication and acute stomach volvulus (1 male, 1 female), 5 (14,7%) duodenal duplication (3 male, 2 female), 1 (2,9%) preduodenal portal vein (1 male), 4 (11,8%) cystic duplication of the bowel with volvulus (3 male, 1 female), 2 (5,9%) intestinal pseudo-obstruction (1 male, 1 female), 3 (8,8%) neonatal appendicitis (1 male, 2 female), 2 (5,9%) Littre's hernia (1 male, 1 female), 1 (2,9%) colon duplication (1 female) (Tab. II).

The prenatal diagnosis was made within 31 gestational weeks, in 7 cases (20,6%) before 20 weeks and included gastric duplication (1 case), bowel duplications (2 cases), and duodenal duplications (4 cases).

Seventeen patients (50%) had associated diseases. Volvulus, caused by a duplication, occurred in 6 (17,6%) neonates (2 with gastric duplications and 4 with bowel duplication); botallo arterial duct in 8 (23,6%) infants; interatrial septal abnormalities in 2 patients (5,9%); gallbladder malformation in 1 neonate (2,9%).

The patients were operated between birth and 25 days of the life and the weight ranged between 750 and 1800 grams. 4 babies (1 gastric perforation, 3 intestinal perforation) were operated in neonatal intensive care unit (NICU) because of their severe cardiac and respiratory conditions.

As we can observe in table II, we assembled the patients in three groups, according to clinical presentation: 17 (50%) with gastrointestinal obstruction, 14 (41,2%) with gastrointestinal perforation and 3 (8,8%) with neonatal appendicitis.

All 17 (50%) children with bowel obstruction underwent emergency surgery. The diagnosis was performed with clinical evaluation and imaging studies. Vomiting and distension were the most common presentations. Chest and abdominal x-ray showed marked distension of the bowel. Contrast studies were necessary clarify the anatomy and the site of obstruction in 11 cases (32,4%).

Ten of 17 children (29,4%) (2 gastric duplication and acute stomach volvulus, 4 duodenal duplication, 4 cystic duplication of the bowel with volvulus) were operated at birth. 3 (8,8%) of 17 (1 duodenal duplication, 1 colon duplication and 1 preduodenal portal vein) were operated at 3 days of age. 4 (11,8 %) of 17, 2 with Littre's hernias and 2 with intestinal pseudo-obstruction, underwent surgery in the first 20 days of life.

The treatment was different based on the primary pathology. The gastric duplication with volvulus were in the greater curvature and we performed derotation, excision together of a wedge of stomach with the cyst and closure of the gap with a double layer of horizontal sutures. In 3 cases of duodenal duplication we excised it, in 2 cases we performed an incision of the cystic wall and cysto-duodenal anastomosis. In 1 of these cases cholecystectomy was performed, because of the gallbladder malformation. In the cases with bowel duplication with volvulus derotation, resection and anostomosis end-to-end were made. In the case of preduodenal portal vein, duodenoduodenostomy as described by Kimura was performed. In the patients with Littre's hernias the resection of a Meckel's diverticulum with adjacent small bowel segment was carried out, with subsequent anastomosis end-to-end and closure and division of the sac. The 2 neonates with intestinal pseudo-obstruction underwent different surgery. A 6 months child was admitted to our Department for multiple episodes of intestinal subocclusions. She had an ileostomy performed in another Hospital. The history of the girl showed a colostomy at birth, ileostomy at fifteen days of life and pull through for Hirschprung at 4 months. We performed a diagnosis of myogenal intestinal pseudo-obstruction after bowel biopsies and we closed the ileostomy. The other child underwent bowel biopsies and the diagnosis was neurological intestinal pseudobsruction. Now, both of them are on "therapy parenteral nutrition". There were no intraoperative and postoperative complications.

Fourteen (41,2%) patients were operated for gastro-intestinal perforation. All patient had abdominal distension, respiratory distress, cyanosis and hypoxemia. An abdominal x-ray confirmed the presence of massive pneumoperitoneum. Laparotomy revealed 8 small bowel perforations and 6 gastric perforations, all along the greater curvature of the stomach, 4 over the anterior wall and 2 over the posterior wall. The gastric perforation, was repaired in two layers. There were no intraoperative complications. One (2,9%) of these babies died in 12th day after surgery for severe postoperative septicaemia and cardiac anomalies associated.

In the patients with bowel perforations we performed temporary ileostomy previous resection of perforated tract. The intestinal continuity was restored within 3 months after surgery. No intraoperative complications were found. There were 2 postoperative complications. In one patient, the ileostomy was revised because of prolapse (40th postoperative day), in the other one, the ileostomy was early closed (45th postoperative day) because of the intussusception of proximal ileostomy.

Three (8.8 %) babies had appedicitis. 2 of these newborn showed a rectal bleeding, therefore they underwent laparotomy because of a suspected volvulus. An acutely inflamed appendix was found intraoperatively, leading to appendectomy. Histological investigation confirmed acute ulcero-phlegmonous appendicitis. The last patient, instead, was referred to our center in 22nd days of life,

Table 1

NEONATAL SURGICAL EMERGENCIES OF GI TRACT. THE TABLES SHOWS THE PRINCIPAL CAUSES OF NEONATAL EMERGENCIES, SPECIFYING THE COMMON AND UNCOMMON ONES.

Occlusion		
	Common	**Uncommon**
	Malrotation (duodenal obstruction, volvulus, internal hernia)	Gastric duplications and acute stomach volvulus
	Duodenal atresia, stenosis or annular pancreas	Congenital pyloric atresia or web
	Jejunal atresia or stenosis	Duodenal duplication
	Ileal atresia or stenosis	Preduodenal portal vein
	Simple meconium ileus	Cystic duplication of the bowel with volvulus
	Meconium ileus with perforation	Colon atresia
	Meconium plug syndrome	Colon duplication with volvulus
	Hirschsprung's disease	Functional Intestinal obstruction
	Drug-induced ileus	Intestinal Psuedo-Obstruction
	Hypertrophic pyloric stenos	Neuronal Intestinal Dysplasia
	Meckel's diverticulum	
Perforation		
	Common	**Uncommon**
	Necrotising enterocolitis	Spontaneous intestinal perforation
	Meckel's diverticulum	
Miscellaneus		
	Common	**Uncommon**
	Hemangiomas, lymphangiomas and vascular	Neonatal appendicitis
	malformations of GI	Littre's hernia

Table 2

UNCOMMON CAUSES OF NEONATAL SURGICAL EMERGENCIES OF GI TRACT, TREATED IN THE DEPARTMENT OF PEDIATRIC SURGERY OF SIENA.

Occlusion		
	Uncommon	**Numbers of cases**
	Duodenal duplication	5 cases
	Preduodenal portal vein	1 case
	Cystic duplication of the bowel with volvulus	4 cases
	Intestinal Psuedo-Obstruction	2 case
	Gastric duplications and acute stomach volvulus	2 cases
	Colon duplication	1 case
Perforation		
	Uncommon	**Numbers of cases**
	Spontaneus gastric perforation	6 cases
	Spontaneous intestinal perforation	8 cases
Miscellaneus		
	Uncommon	**Numbers of cases**
	Neonatal appendicitis	3 cases
	Littre's hernia	2 cases

because of anorexia, vomit and abdominal distension. He has been treated with antibiotic and fluids therapy. The abdominal x-ray showed free air in the peritoneal cavity, dilated ileal loops, some air–fluid levels in the lower abdomen but no evidence of pneumatosis intestinalis. A diagnosis of peritonitis was made, maybe secondary to neonatal necrotizing enterocolitis (NNEC) and the emergency laparotomy showed perforated appendicitis. There were no obvious signs of NNEC, Hirschsprung's disease or other obvious anomalies in the intestine. An appendicectomy was performed and full-thickness biopsies taken from multiple sites from the large bowel with normal histological report. There were no intra and postoperative complications.

In all patients the oral feeding was started between 4th and 10th postoperative day.

The average hospitalization was about 70.47 days (range 15 days-6 months).

About the follow-up, we evaluated clinically all the patients at 6, 12, 24 and 36 months after surgery. To date, all patients have finished the follow-up. So, we can conclude that all of them have a good growth, a good weaning and nobody has problems with the oral feeding (vomit, drooling, hematemesis or belching). The two patients with TPN are very well. 4 (11,8%) patients (intestinal volvulus) are constipated, 3 (8,8 %) have meteorism. Contrast x-ray was done in the patients who had bowel anastomosis and it showed a normal transit.

Discussion

It is very difficult to find a clear definition of "rare" pathology especially in neonatal age. Because of that we decided to divide our patients based on the type of clinical features related with the presence of "uncommon" or "common" aetiology. Then we focused our attention on the group's uncommon pathology. From January 1995 to December 2011 36.000 newborns were borned on our district; 31 were admitted to our Department for rare surgical emergencies caused by "uncommon" aetiology. In our series the clinical picture of obstruction or perforation had a similar distribution. Male to female ratio was 3:1. Birth weight was always under 1500 grams. All patients with cystic abdominal mass had a prenatal diagnosis. All babies underwent to surgery early. There mortality's rate was 3.2%. The complication's rate was 6.4% and it involved just patients with intestinal perforation.

Intestinal obstruction is the most common surgical emergency of the newborn. The incidence of neonatal intestinal obstruction is approximately 1 case for every 500-1000 live births. Approximately 50% of these neonates have intestinal atresia or stenosis. Duodenal atresia and jejunal atresia occur in approximately equal numbers, although some authors report that jejunolileal atresia is the more common.[3] Congenital pyloric atresia (CPA) is a very rare condition that was first described by James Calder in 1734.[4] Commonly, CPA occurs as an isolated lesion, which has an excellent prognosis, but it can also be seen in association with other malformations, which can have a negative impact on the final outcome.[5] Colon atresia is an extremely rare conditions.[6]

Intestinal duplications are rare congenital malformations that could cause intestinal obstruction as mass effect or causing volvulus. Duplications of the alimentary tract occur in 1 of 4500 births. Gastric duplications constitute 8% of these or roughly 17 of every 1,000,000 births. A bowel volvulus caused by the presence of a cystic intestinal duplication can lead to an intestinal perforation. The different locations and sizes of these duplications require a specific diagnostic and surgical approach. Early diagnosis and treatment of uncomplicated intestinal duplications by means of prenatal sonographic screening and laparoscopic-assisted resection, respectively, are desirable in this congenital malformation, in order to prevent the risk of occlusion/perforation.[7,8]

Prenatal ultrasonography can readily identify proximal obstructing lesions that can produce proximal bowel dilation with hyperperistalsis and the classic "double bubble" appearance of duodenal atresia. Distal intestinal obstructions are less likely to cause polyhydramnios but on occasion dilated loops of bowel may be identified as anechoic masses.

Over the maternal polyhydramnios, four clinical findings suggest intestinal obstruction in the neonate: excessive gastric aspirant, abdominal distention, bilious vomiting, and failure to pass meconium. The presence or absence of each of these clinical findings depends largely upon the level of gastrointestinal obstruction. Early recognition of intestinal obstruction is imperative if the complications of respiratory compromise and sepsis are to be avoided.[2]

Gastrointestinal perforation is a catastrophic condition in neonates, especially in premature neonates.

Spontaneous perforations of the gastrointestinal tract occur in the stomach, duodenum, small intestine or colon. The problem is encountered only infrequently; so, the exact incidence is unknown. Male infants are affected more often than females (approx. 4:1).[9]

Gastric perforation among neonates is a rare but frequently fatal condition of uncertain etiology. It is associated with high mortality, particularly in premature infants. There is also a trend towards higher mortality in lower-birth-weight infants.[10]

Almost all spontaneous perforations of the GI tract are considered to be the result of ischemic necrosis. The perforation is the end result of "selective circulatory ischemia," a defense mechanism of the neonate to hypoxia, physiologic stress, and shock. In response to physiologic stress (hypoxia, hypovolemia, etc.), blood is selectively shunted away from mesenteric vessels to the more vital heart and brain. Local mesenteric ischemia can progress to microvascular thrombosis and subsequent gastrointestinal wall necrosis and perforation. Although ischemia is likely the underlying problem, other factors including bacterial colonization, hyperosmolar feeds, and an immature neonatal immune system may also contribute.[9]

Specifically, in the gastric tract, gastric wall ischemia, hypothalamic mechanisms, excessive gastric acid-pepsin ulceration and congenital muscular wall defects could account for spontaneus gastric perforation, even if many authors believe that this spontaneus rupture is due to an acute rise in the intragastric pressure at the time of parturition.[10]

Most infants can immediately present signs of perforation. In fact, in case of spontaneous gastrointestinal perforation infants present within the first week of life (usually 4-5 days) with an abrupt onset of abdominal distention and associated tachycardia, hypovolemia, and poor systemic perfusion. With severe pneumoperitoneum, respiratory function is compromised requiring urgent intubation. Typically, the abdomen is markedly distended and tympanitic to percussion. Pneumoperitoneum is usually present in these infants. The clinical course of neonates with spontaneous gastrointestinal perforation may mimic those of NEC or other diseases associated with perforation.[9,11,12]

Acute neonatal appendicitis is a rare condition associated with significant morbidity and mortality. The severity of this disease is caused by its tendency to occur more frequently in premature infants, an increased perforation rate with rapid progression to peritonitis, and delay in diagnosis and intervention. The presentation of neonatal appendicitis can be identical to necrotizing enterocolitis, leading to misdiagnosis.[13]

Littre's Hernia was originally defined by Rinke in 1841 as "the presence of a Meckel's diverticulum in any hernial sac". It is generally difficult to differentiate from other types of hernia, until complications arise. It is a rare and accidental finding at any age, but absolutely exceptional in neonates. The potential surgical risk is linked to the presence of a Meckel' diverticulum.[14]

The initial treatment of any suspected neonatal obstruction includes placement of a nasogastric tube to decompress the stomach and

to prevent vomiting/aspiration. Fluid and electrolyte replacement should be quickly undertaken to resuscitate the infant and restore circulating blood volume in anticipation of the potential need for surgical intervention. Most obstructive lesions in neonates will require surgical therapy and surgery should not be delayed once volume resuscitation is adequate. If an intestinal anastomosis is anticipated peri-operative antibiotics are indicated.[3]

In the presence of clinical and radiological signs of perforation, treatment commences as soon as possible, simultaneous to the diagnostic workup. Rapid deterioration is anticipated and prevented with aggressive fluid resuscitation, intravenous antibiotics, correction of acid-base disturbances, and nasogastric decompression. Intubation and ventilatory support is required in infants with respiratory distress. Aspiration of the massively distended pneumoperitoneum can be helpful in infants with severe life-threatening respiratory compromise. Surgical exploration is indicated. The site of perforation is identified although in up to 10% of cases the perforation site has sealed spontaneously and cannot be identified. Surgical treatment is dictated by the infants physiologic condition and the findings at laparotomy (i.e., site of perforation, tissue condition, soilage, etc) and include primary repair, resection with external diversion, resection with anastamosis, drainage, etc. Obstruction distal to the site of perforation is excluded whenever possible.[15] The outcomes from neonatal intestinal obstruction or perforation vary with the etiology of the disease. Overall survival is generally good but often is influenced by the associated anomalies of each condition.[2]

A multi-disciplinary team of surgeons, anaesthetists, neonatologists, radiologists, cardiologists, obstetricians, nurses, physiotherapists and other health professionals experienced in dealing with extremely small infants will provide the best outcome.

The prognosis is adversely affected by prematurity, the presence of other anomalies, and a delay in diagnosis.[15]

Conclusions

According to the literature and after the review of our casuistry, we can affirm that it's very difficult to qualify a "rare" disease, especially surgically. However it's possible to find some uncommon causes that can produce a surgical emergency. The majority of these disease occurs as neonatal acute abdomen. The outcome of these patients is different based on the etiology even if the overall survival is generally good. So, in conclusions, the aim of our study is to confirm that it is necessary to evaluate also these diseases in case of neonatal acute abdomen. Also, an early diagnosis and surgery can ensure the good survival even if it is influenced by the associated anomalies.

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