

Perinatal management of gastroschisis

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Abstract

Gastroschisis is an abdominal wall defect, typically located to the right of the umbilical cord, requiring an early surgical treatment shortly after birth. Affected patients can be identified during intrauterine life with US and should be delivered in referral hospitals where a multidisciplinary approach can be provided, involving neonatologists, clinical geneticists, surgeons and other specialists. These patients require a complex management in Neonatal Intensive Care Unit (NICU) and a long term follow-up after discharge. Exceed the acute neonatal condition, gastroschisis has a good prognosis, if there are no overlapping complications, and it should be differentiated from omphalocele, burdened with worse prognosis, and other conditions in the wide spectrum of abdominal wall defects.

Keywords

Abdominal wall defect, prenatal diagnosis, newborn, malformation, intensive care, surgical treatment.

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Introduction

Gastroschisis (or laparoschisis) is an abdominal wall defect typically located to the right, rarely left, of the umbilical cord in which intestines, and occasionally other abdominal contents, herniate through the wound without lining membranes. Another rare form of gastroschisis described in the literature is the vanishing one (or closed). Gastroschisis has been also classified into simple and complex forms. The latter correspond to the cases where there are intestinal atresia, perforation, ischemia or necrosis, or loss of bowel occurred in utero.

In the wide spectrum of abdominal wall defects, gastroschisis should be distinguished from omphalocele. A first difference relates to the anatomy of the malformation: the first is a paraumbilical cleft, usually to the right of the umbilicus, while the second is a defect in the midline, with protrusion of part of the intestine into the base of the umbilical cord. In the omphalocele the herniated portion does not occur covered by skin, muscle or fascia, but only by a thin membrane consisting of amnion externally and peritoneum internally, with mesenchymal connective tissue between them. In gastroschisis, instead, the viscera displaced have no coating. Gastroschisis probably derives from a vascular defect, unlike omphalocele in which there is the persistence of the umbilical cord, in the region normally occupied by somatopleure. Recent hypotheses have focused on a vascular aberration of umbilical and omphalomesenteric veins, interfering with the development of the somatopleure at the junction with the body stalk or a solution of continuity that is formed later than the development of the abdominal wall. Alternatively, the cause of gastroschisis may be an early vascular accident involving the omphalomesenteric arteries [1, 2].

Gastroschisis has a better prognosis, with less perinatal mortality, compared to omphalocele (given its frequent association with karyotype abnormalities and genetic syndromes). However in infants with gastroschisis the herniated organs may present signs of compression, that carries an increased risk of obstruction, stenosis, perforation, meconium peritonitis, polyhydramnios and ischemic events. Epidemiological studies published in the last two decades, reveal an average prevalence of 1.76:10,000 born (0.4 to 3.01) [3-6].

The etiology of gastroschisis is not known, but is thought to be the result of a combination of genetic and non-genetic factors. Various hypotheses have been formulated. Support for a vascular mechanism

comes from the association with various vasoactive pharmacological compounds, such as a report of gastroschisis in dizygotic twins following excessive maternal alcohol ingestion in the first trimester (OR: 2.4) [7, 8]. Cigarette smoking (ORs: 1.2-2.1) and recreational drug abuse, known to cross the placenta and affect the umbilical circulation, have also been linked to gastroschisis and both induce vasoconstriction and have deleterious effects on the fetoplacental circulation [9-11]. Among the non genetic risk factors, drugs taken during pregnancy – such as aspirin, ibuprofen and acetaminophen (OR: 2.2) [12-16] – deserve attention. Another risk factor is represented by maternal genitourinary infections, before or during the first trimester of pregnancy [17]. The role of young maternal age is described in most studies: women aged between 14 and 19 have a higher risk of pregnancies with offspring affected by gastroschisis (OR: 7.2), compared to women aged 25-29 years [18]. In addition, combining maternal age and ethnicity, the data show a greater risk for white women between 20 and 24 years of age, especially of Hispanic nationality (OR: 1.5), as well as recent data from some British records indicate an higher incidence in England, with 2-3 cases for 10,000 births in total. The risk of recurrence is low (3-5%). Some studies attribute a role to father's age, indicating an increased risk among fathers aged between 20 and 24 years compared to those aged 25-29 years [19]. Regarding the genetic risk factors, there are only few cases in which chromosomal abnormalities have been detected.

Materials and methods

This retrospective study examined infants with gastroschisis born in the last decade at Palermo University Hospital. We collected anamnestic data, relating to the couple and the course of pregnancy, perinatal and neonatal features, paying attention to the pre- and post-surgical management.

The history has focused on the method of conception, the course of pregnancy and the presence of any risk factors. The family history has been directed toward search of previous cases of gastroschisis or hereditary diseases. It was investigated a correlation with the age of the parents and maternal intake of alcohol or smoking during pregnancy, as well as any maternal diseases or infections arising during pregnancy. Among the perinatal and neonatal parameters, moreover, it has been paid more attention to sex and karyotype, the mode of delivery, gestational age and birth weight.

The congenital defects have been described in the size of the breach, differentiating the herniated viscera, represented only by the intestine or also by stomach and bladder. In association with gastroschisis, in some infants there were other defects, some determining complications that influenced the clinical course. It has been assessed the timing and mode of surgery, distinguishing between cases with immediate closure and those with packaging of a silo. In addition we explored pre- and postoperative ventilatory support and nutrition, in terms of method and timing, which have influenced the average length of hospitalization.

Results

In the study period we assisted 5 infants with gastroschisis (pictures of patients 3 and 4 are shown in **Figures 1-3**). Prenatal ultrasounds allowed to identify, three patients with gastroschisis during intrauterine life, between the 18th and the 31st week of gestation. First of all, we evaluated the risk factors identified over the years by various authors. Parental age was on average 26 and 29 years, respectively in mothers and fathers, values that differ from those in the literature that report an increased risk of gastroschisis in women who conceive between 14 and 19 years (OR: 7.2) and for fathers aged between 20 and 24 years [18-19]. Regarding the possible association of the disease with smoking cigarettes, this habit was present only in one of the mothers of our patients. An important role is attributed to genitourinary infections, before or during the first trimester of pregnancy (OR: 5.0) [17], and we demonstrated a seroconversion for *T. gondii* in the first trimester in one pregnancy.

There was a predominance of females, with a ratio F:M of 1.5:1. At birth, we studied the karyotype of each newborn and there were no chromosomal abnormalities.

All neonates except one were born by caesarean section and in two cases there was meconium-stained amniotic fluid. The choice of timing was mainly based on the condition of the fetus, assessed by ultrasound examination.

At term birth is associated, in general, with a better neonatal outcome because of the possibility of earlier closure of the defect and less amount of time for the transition to oral feeding. Unfavorable prognostic signs at prenatal ultrasonography, which may represent an indication for preterm delivery (34-36 weeks of gestation) are: reduced



Figure 1. Patient 3 at birth. Massive herniation of the entire small bowel, stomach, bladder and spleen. Intense inflammatory component, with significant dilatation of the bowel loops, intensely erythematous and covered with fibrinous exudate. Chemical peritonitis has already led to the formation of adhesions between the loops.



Figure 2. Patient 4 at birth. Herniation of the small intestine. Bowel loops covered with fibrinous exudate and adherence, without significant dilatation.



Figure 3. Patient 3 in second day of life with a silo performed for gradual reduction of herniated viscera.

intestinal peristalsis, increase of thickening of the abdominal wall, reduced intestinal vasculature, oligohydramnios.

Our patients presented a mean gestational age of 36 weeks (range: 34.3-37.6) and an average birth weight of 2,200 g (range: 1,900-2,900). In all cases the wall defect was right located, confirming the extreme rarity of left locations [20] or even vanishing forms [21]. More than an infant presented at birth malformations associated with gastroschisis: intestinal malrotation (1/5), umbilical hernia (1/5), patent ductus arteriosus (1/5); intestinal aganglia (1/5). It has been shown that fetuses with abdominal wall defect are at risk of congenital heart disease and especially of persistent pulmonary hypertension, therefore echocardiography is indicated in pre-natal and postnatal follow-up [22]. Among the complications arising in patients with gastroschisis, the most frequent were: intestinal obstruction (2/5), biliary regurgitation (2/5), respiratory distress (2/5), acute renal failure (2/5), short bowel syndrome (1/5). In addition, a newborn developed an infection with coagulase-negative *Staphylococcus* (**Tab. 1**).

At birth, all patients showed a good adaptation to extrauterine life. All infants underwent the surgical correction of gastroschisis in the first 24-48 hours of life. Only in one newborn it was carried out the packaging of a silo (**Fig. 3**) and the delayed repair of the defect. Following surgery, the mean duration of total parenteral nutrition was 25 days (range: 10-52), while on average these infants were ventilated through an endotracheal tube for 12 days and required an average hospital stay of 62 days

(range: 24-98); the timing of nutritional support and ventilation were reduced to a minimum, taking into account the associated iatrogenic (infective, metabolic, ...) risks. Two patients died, for the other three surgery and neonatal intensive care were resolutive with a survival of 60%, while in the literature appears a survival of 90%.

Management of infants with gastroschisis

Gastroschisis is a rare malformation which requires timely surgical correction and neonatal intensive care. The medical team should be prepared before the birth of an infant with gastroschisis, therefore a prenatal diagnosis is very important for neonatal outcome. The possibility of a twin pregnancy increases the complexity of the perinatal management [23]. The diagnosis of gastroschisis can be made between the 18th and 20th week of gestation, suggested by the presence of irregularities of abdominal wall, hyperechoic loops, devoid of echogenic coating, protruding in lateral position respect to the normally inserted umbilical cord. Ultrasound examination can also detect IUGR and possible complications.

At birth, our first approach to the infant with gastroschisis is aimed at preventing heat and fluid loss, onset of infection, ischemic or traumatic injury to the intestine. Over the years there have been proposed various prognostic factors detectable at birth, which guide the post-natal care. The presence of respiratory failure seems to increase mortality. Our initial attitude in these cases is conservative

Table 1. Characteristics of gastroschisis and correlated diseases in five patients.

	Herniated viscera				Associated malformations	Complications
	EI	I	S	B		
Patient 1	-	+	-	-	-	infection by coagulase-negative <i>Staphylococcus</i>
Patient 2	+	-	-	-	aganglia	intestinal obstruction, acute renal failure, short bowel syndrome
Patient 3	+	-	+	+	-	biliary gastric regurgitation, intestinal obstruction, respiratory distress
Patient 4	-	+	-	-	intestinal malrotation, umbilical hernia	biliary gastric regurgitation
Patient 5	+	-	-	-	patent ductus arteriosus	renal failure, respiratory distress

EI: entire small intestine; I: some intestinal loops; S: stomach; B: bladder.

and, after obtaining an improvement in the clinical condition of the patient, we proceed with the surgery.

The aim of surgical treatment of gastroschisis is to restore the integrity of the anterior abdominal wall, reducing the herniated viscera within the abdominal cavity. The surgeon can proceed with the primary fascial closure of the defect or the allocation of the loops eviscerated in a container (silo) and reduction of the content over time. Schuster first described [24] the creation of a silo for patients in whom there was a lack of domain for the viscera (visceral abdominal disproportion), allowing a gradual return of the viscera in the abdomen and a natural dilatation of the abdominal wall in one or two days, without respiratory and hemodynamic instability usually determined by primary closure. It is important to monitor the intra-abdominal pressure already during surgery. The high intra-abdominal pressures may be responsible also for hemodynamic alterations, as in the case in which cause a compression of the inferior vena cava with a reduction in venous return to the right heart and heart failure [25]. After repair of congenital defects of the abdominal wall is often necessary to resort to the reconstruction of the umbilicus reaching results aesthetically pleasing.

The follow-up in the immediate postoperative involves cardiovascular and respiratory functions. Characteristic of newborns with gastroschisis is the “bell” aspect of the chest, due to the protrusion of the abdominal organs outside the abdominal cavity during pregnancy. Furthermore the difficulty in breathing or alteration between the abdominal muscle and diaphragmatic dynamic, makes it difficult to supply the patient, that requires a support of parenteral nutrition. Another characteristic of infants with gastroschisis is the intestinal failure, usually multifactorial, derived from motility disorders, associated intestinal anomalies and loss of intestinal length due to necrosis or surgical resection.

It is now recognized an important role in the early enteral feeding, as well as reducing the times of total parenteral nutrition, and therefore the associated risks, has a trophic action by stimulating the mucosa and promoting growth in length of the intestine. Therefore, early enteral feeding should be the common goal, especially among those neonates undergoing extended surgical resection that predisposes them to short bowel syndrome.

Other complications are those related to ventilation and nutrition. These can be the cause of infectious episodes, where endotracheal tubes

or catheters may be the site of entrance and proliferation of pathogens, bacteria or fungi [26-30]. Both treatments, if protracted, may also lead to organ dysfunction. In particular, prolonged mechanical ventilation can be responsible for the onset of bronchopulmonary dysplasia and oxygen dependence, and long-term parenteral nutrition can frequently cause liver failure and cholestasis.

The management of an infant with gastroschisis involves the cooperation of several specialists such as obstetrician, midwife, neonatologist, geneticist, sonographer, pediatric surgeon, and pediatric nurses. The abilities of integration and cooperation of these caregivers influence the clinical evolution and prognosis of the patient. The neonatologist assumes the role of patient care manager, dealing with various aspects of care in the neonatal intensive care unit, both in the preoperative and in the long and complex post-surgical hospital stay. In particular, the neonatologist manages assisted ventilation, total parenteral nutrition, prevention of infections and identifies and treats any associated diseases [31-34].

The management of patients with gastroschisis therefore requires a highly specialized hospital with a multispecialist team to address the different and complex problems of these young patients. An accurate long-term follow-up is needed to prevent frequent complications. Therefore the centralization of the patients to a referral hospital is crucial to increase and optimize clinical expertise.

Declaration of interest

The Authors declare that there is no conflict of interest.

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