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CASE REPORT

Gastroschisis Spiral-like Closure with Umbilical Cord in a Limited-resource Setting: A Case Report

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ABSTRACT

Gastroschisis is one of the most frequent congenital surgical problems in fetuses and neonates, with a continuously increasing incidence worldwide. In limited-resource settings, the management of this condition is difficult, and the mortality rates are much higher than those observed in high-income countries. We report the case of a newborn with gastroschisis and viscero-abdominal disproportion, submitted to abdominal wall closure through utilization of umbilical cord tissue. This technique has not already been applied in low-resource countries, where it can show advantages such as lower risk of abdominal compartment syndrome and local infection, high availability, and low cost.

Key words: Gastroschisis; Intra-abdominal pressure; Limited-resource setting; Umbilical cord

INTRODUCTION

Abdominal wall defects, mainly represented by gastroschisis and omphalocele, are the most frequent congenital surgical problems in fetuses and neonates [1]. Gastroschisis is a full-thickness defect of the abdominal wall, typically on the right side of a normally inserted umbilical cord, resulting in protrusion of the abdominal organs into the amniotic cavity without a covering membrane [1].

During the past decades, the incidence of gastroschisis has steadily increased worldwide reaching values from 2 to 5:10000 live births [2]. The risk factors for gastroschisis can be subdivided into sociodemographic, mainly young maternal age and low socioeconomic status, and teratogenic ones, such as maternal smoking, alcohol, cocaine, aspirin, ibuprofen, and some vasoconstricting drugs.

Gastroschisis can be easily diagnosed antenatally during routine ultrasound monitoring, with a detection rate of 90% within the second trimester of pregnancy in developed countries. Prenatal diagnosis allows planning of early referral to a tertiary multidisciplinary center with maternal-fetal medicine, genetic counseling, neonatology, and pediatric surgery. No clear evidence supports elective preterm delivery or cesarean section in the absence of obstetric indications [1].

The gold standard surgery for newborns with gastroschisis is still to be identified, and the choice of the strategy usually depends on the degree of viscero-abdominal disproportion. The main options include primary closure and preformed silo placement with gradual visceral reduction before abdominal wall closure [1].

The leading complications of gastroschisis and its surgical treatment can be classified into: Gastrointestinal, related to intestinal damage and effects of prolonged parenteral nutrition; respiratory, caused by impairment of respiratory mechanics since intrauterine life and sudden increase of intra-abdominal pressure after closure of the abdominal wall defect; and infectious, due to loss of anatomical defense mechanisms, use of prosthesis, and central venous lines [3]. These complications account for gastroschisis-related mortality and long-term morbidity. In high-resource countries, the overall post-natal survival estimates of newborns with gastroschisis reach approximately 90–95%, but complex cases present worse outcomes [1].

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In limited-resource countries, although the quantity and quality data are lacking, the gastroschisis mortality rate seems to be much higher [4].

We report the case of a newborn presenting simple gastroschisis, with viscero-abdominal disproportion, who was submitted to closure of the abdominal wall defect through a new technique of utilization of the umbilical cord in a limited-resource setting.

CASE REPORT

A male term neonate, with low birthweight, home delivery, was taken to the Central Hospital of Beira for congenital malformation. The initial examination revealed gastroschisis, with protrusion of stomach, small bowel, cecum, and appendix, associated with viscero-abdominal disproportion. The viscera were wrapped in common non-sterile bandages. At presentation, newborn was in critical condition with severe hypothermia (33°C), desaturation, and dehydration. He was stabilized through heating, oxygen administration, fluid resuscitation, and first-line antibiotic therapy. About 20 h later, he was participated to surgical closure of the abdominal wall defect by means of umbilical cord (that had been left long), under general anesthesia wherein a longitudinal opening of the umbilical cord, dissection, and ligature of the umbilical vessels with reduction of the protruding viscera into the abdominal cavity was done. The suture of the umbilical tissue around the abdominal wall defect followed by spiral-like arrangement of the cord and final closure was done (Figure 1). No intraoperative complication was detected. At the end of the intervention, the intravesical pressure increased to 20 cm H_2O, while 24 h later, it decreased to 17 cm H_2O. In the early post-operative phase, the newborn succumbed to death owing to severe sepsis and hyperglycemia.

DISCUSSION

Congenital anomalies, and in particular gastroschisis, represent a significant contributor to the avertable global burden of disease profile [4]. The disparity in survival rates of neonates with gastroschisis between high-income and low-income countries is evident. In Sub-Saharan Africa, mortality rates of newborns with gastroschisis range from 35% to 100% and septicemia represents one of the most common complications and causes of death in limited-resource settings [4,5].

In low-income countries, the management of neonates with gastroschisis is difficult due to poor prenatal diagnosis, late referral to tertiary centers through unappropriate transport system, shortage of neonatal intensive care facilities, dearth of trained surgeons, support personnel and surgical equipment, and higher risk of sepsis [6].

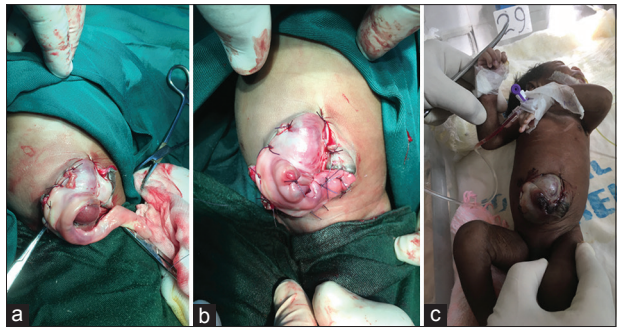

Figure 1: The operative technique and its result. (a) Spiral-like arrangement of the umbilical cord around the abdominal wall defect. (b) Final result of the intervention. (c) Appearance of the abdominal wall 24 h after the operation

The surgical strategy plays a central role in the management of newborns with gastroschisis, but the gold standard intervention has not been identified and contrasting data have been reported. In fact, a review showed no significant difference in outcome between the use of preformed silos and other types of surgical treatment [7], while other authors suggested potential major benefit of primary closure over silo placement followed by delayed closure [8]. Considering the risk of abdominal compartment syndrome due to the presence of viscero-abdominal disproportion, the primary closure is not always possible and it represents a critical problem, especially in low-resource settings, where surgeons usually adopt custom silos [6]. Gastroschisis spiral-like closure with umbilical cord is an affordable alternative. In case of successful reepithelialization, this strategy represents a primary closure; otherwise, the infant could require a second surgical step for definitive closure of the abdominal wall defect.

Different techniques of umbilical cord utilization for closure of gastroschisis have previously been reported [9,10], but to the best of our knowledge, this is the first application in a limited-resource setting. It could represent a good practice as the collocation of the umbilical tissue around the abdominal wall defect in a spiral-like way can allow a primary closure of the defect avoiding excessive increase of intra-adominal pressure and therefore reducing the risk of abdominal compartment syndrome and respiratory complications. This aspect is particularly important in low-resource centers, where ventilatory support usually is not available.

Another advantage of this procedure is represented by the use of autologous tissue, with reduced risk of local infection, high availability, and low cost. These characteristics make this surgical strategy affordable in limited-resource settings. To apply this technique, it is essential to educate the obstetricians of primary and secondary health-care services to early send newborns with gastroschisis to the referral center and to leave the umbilical cord as long as possible.

Finally, although the surgery procedure was successfully performed, with an acceptable intra-abdominal pressure, our patient died because of sepsis, one of the most common complications of gastroschisis in limited-resource settings.

In conclusion, in patients with gastroschisis, especially in case of viscero-abdominal disproportion, the use of the umbilical cord for the repair of the abdominal wall defect is an option that is easily applicable and affordable even in limited-resource settings. To have the chance to perform this technique and to obtain an acceptable survival rate, without waste of resources, it is crucial to create an integrated and multidisciplinary approach and to train all the involved health workers, in particular obstetricians who should refer patients with gastroschisis early and leave the umbilical cord as long as possible.

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