



Case Report
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Left-Sided Gastroschisis: Case Report and Some Anatomic and Technical Considerations



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Abstract

Gastroschisis, a congenital periumbilical abdominal wall defect/malformation, allows intrauterine herniation of intestines at the right side of the umbilical cord. Its pathogenesis is still blurred as no tenable explanations or specific causes are found that can elucidate its increasing incidence presently in place among teenager and very young pregnant women. Left-gastroschisis is a rare variant of the commoner right-sided gastroschisis. It is sporadic in its prevalence and has as well an etiology unknown. When left-sided gastroschisis is present, one should be aware that an increased number of associated extra-intestinal abnormalities are likely. Our case has HIV exposure as a potential factor that could have had a role in the ontogenesis of the defect.

Keywords: Left-sided gastroschesis; HIV exposure; Fascia alba split technique

Introduction

Gastroschisis is a well known clinical entity of a congenital periumbilical abdominal wall defect/malformation through which intestinal herniation occurs during the foetal life [1-3]. Its incidence is increasing worldwide [4-6]. Intrauterine exposure of the eviscerated intestines to amniotic fluid makes them oedematous, chronically inflamed and covered by yellow-greenish peels as end-result of amniotic deposition over them of fluid pigments and other body waste. This is the obvious clinical picture of the infants at birth. Other hidden characteristics are the obligatory association of intestinal malrotation and the universally reported female predominance [7,8].

Left-sided gastroschisis, though uncommon, is suitably well described and reported in literature [9-15]. However the exact incidence of such a peculiar form of gastroschisis is unknown in South Africa. Its rarity when coming to South African reports may risk to confine this type of rare congenital anomaly into an anatomical curiosity more than an autonomous abdominal wall defect/malformation within the broader subject of gastroschisis. The HIV exposure of our patient seems purely serendipitous while exposure to other health conditions is well established risk for gastroschisis [16-18].

Case Report

A normal vaginal delivery of a young pregnant woman attending the antenatal surveillance programme at a peripheral clinic of our referral territory was marred by the discovery that her newly delivered baby-girl had a large gastroschisis. The transfer to neonatal intensive care unit (NICU) of Grey's Hospital had been speedily organized. After the haemodynamic stabilization the baby was taken to theatre for correction of the defect and repositioning of the oedematous bowel into the abdominal cavity.

The intraabdominal reduction of the eviscerated and oedematous intestines was facilitated by our technique of generously splitting, subcutaneously, the fascia alba toward the xiphisternum and the pubic bone. Intestines were accommodated without difficulty. The patient tolerated well the procedure and was extubated at the end of the operation. She remained on full parenteral nutrition (PN) for a few days. After the opening of the intestines she was gradually introduced to breast milk and was able to be on full breast feeding after a week from the operation day. Shortly thereafter she was discharged home. The follow-up was uncomplicated and the patient transferred to the

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Paediatric outpatient Clinic for the routine control of growth and vaccination programme. We were unable to find extra intestinal malformations and any causal link between HIV exposure and gastroschisis (Figure 1).



Figure 1: Showing the defect on the left with the umbilical cord to the right.

Discussion

Gastroschisis with its increasing trend of occurrence in very young pregnant women represents a challenge for clinicians as well as for researchers. Both groups have not yet come out with definitive answers about aetiology and pathogenesis of this congenital defect/malformation. Many are the embryologic hypotheses put forward but none has reached a solid universal consensus [19-21]. No certain risk factors have been identified which make weak the numerous published epidemiological surveys [22]. Sex ratio in all studies have always confirmed female predominance that is even more pronounced for the patients affected by the left-sided gastroschisis [7,8].

Though the abdominal wall defect is usually small, still the entire intestine and stomach can herniate through. Despite such a massive intestinal herniation there is no a preferred mode of delivery infants with gastroschisis. None has a prognosticator value and as a consequence caesarian section should not necessarily be seen as the only way of delivering infants with prenatal diagnosis of gastroschisis [23-25]. Gut oedema present in the herniated intestines after birth is the combined result of their chronic oedema from the continuous amniotic bath during the intrauterine time and of a potentially impaired portal venous return that after birth may be worsened. Their deterioration may become very rapidly sinister in the postnatal period if none cares for securing an appropriate venous drainage to the herniated bowel.

The venous axis must be kept straight to avoid venous kinking and the onset of an acute venous obstruction that would irreversibly worsen the state of intestines by aggravating their oedema and eventually by compromising their viability. A silo may represent a temporary solution to this pressing problem before taking the patient to theatre for bowel reduction into the abdominal cavity and for the final repair of the abdominal wall defect. However, grossly dilated gut containing inspissated meconium represents more a quandary than an ideal condition

to work with. Extensive bowel manipulation may indeed worsen its oedema and make serosal tears possible. Intraabdominal hypertension is the risky consequence of an oedematous gut that is brought back into an abdominal cavity in which its wall has never been stretched during the intrauterine life by the presence of growing intestines, its inside anatomical space is limited and is "unprepared" to accommodate organs with a volume well over its capacity.

The familiar intraoperative abdominal stretching is most of the times unable to increase significantly the compliance of the abdominal wall to make the abdomen more receptive. The fascial layers are inelastic and their expanse limited, unless stretched to a point of their laceration. The habitual operative maneuvering of abdominal wall stretching then cannot always be successful [26,27]. We have introduced in our approach to gastroschisis the breach of the hernial ring and a generous split of the fascia alba toward xiphisternum and pubic bone before the intraabdominal intestinal reduction. The skin above the fascial layer is much more malleable and the intestine has no great difficulty to be housed back into the abdominal cavity. This is what we have observed since the introduction of our technical approach.

This patient was not an exception, her intraabdominal pressure remained low as it was demonstrated by a low airway pressure during the operative time and her extubation at the end of the operation. Though the acquired knowledge of left-sided gastroschisis is considerable, its aetiology remains obscure and pathogenesis controversial as there are no answers for its incidence [28-32]. The economic burden of gastroschisis on countries with high natality rates is conspicuous and the measures to prevent it concealed [33-43].

Conclusion

Left-sided gastroschesis is a rare neonatal condition and the role of maternal HIV infection is not well known. Many techniques are used to manage this condition. At Grey's Hospital, we use a generous split of the fascia Alba toward the xiphisternum and the pubic bone to breach the hernia ring.

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