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Original Research Paper

SURGERY

CLOSED GASTROSCHISIS: AN INFREQUENT ENTITY WITH POOR OUTCOME

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ABSTRACT

Intoduction Closed gastroschisis is rare congenital abdominal wall anomaly in which umbilical ring closed around viscera that can lead to loss of bowel length, short gut syndrome and carry high mortality.

Material and methods We have performed 8 year review (from 2010 to2018) of medical records of our cases. All information related to this patients were collected from medical records and analysed.

Results During this period we had operated total 6 cases of closed gastroschisis. All had entry and exit level atresia. All cases were operated after resuscitation. In all cases we had created divided stoma. All patients were provided multidisciplinary post-operative care but due to short bowel syndrome none of them survive.

Conclusion Closed gastroschisis is rare congenital anomaly that carry very poor prognosis in developing country.

KEYWORDS : Closed gastroschisis, Jejunal atresia, Short gut syndrome.

INTRODUCTION

Closed gastroschisis is congenital abdominal wall defect in which fascial ring closed around protruding viscera and it can manifest in various phenotypic way. [1] There may be entry or exit level atresia with subsequent complication (like short bowel syndrome).[1-3] Although survival rate in western world has improved very much due to availability of supportive care and antenatal detection, but in developing country like India still survival is very rare. We are presenting our experience with this entity.

Material and methods

After obtaining approval from the institutional review board at the Gandhi medical college, Bhopal a review was conducted of all infants born with gastroschisis between January 2010 and January 2018. The medical records were reviewed for patient demographics (maternal history, prenatal evaluations, gestational age at birth, birth weight, and sex) and details of gastroschisis and involved intestine, operative procedures and findings, nutritional support, ventilator requirements, hospital course, and overall outcomes (mortality, intra hospital complications, length of stay, outpatient follow-up).These data were collected and analysed.

Results

Between January 2010 and January 2018, we treated 6 cases of closed gastroschisis. In all this cases umbilical defect had closed before birth, causing strangulation of the herniated bowel. Ischemic damage was noted in the variably mummified, matted, and fibrosed remnant of extracorporeal midgut (figure one).

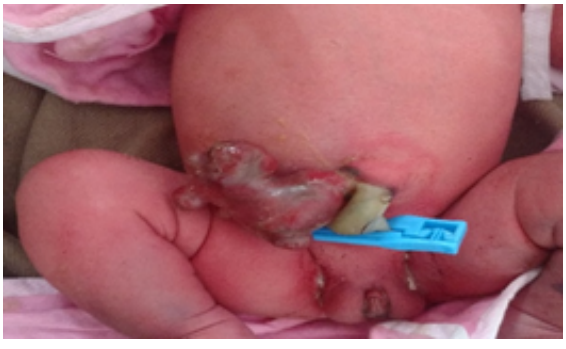

Figure one showing mummified, matted and fibrosed remnant.

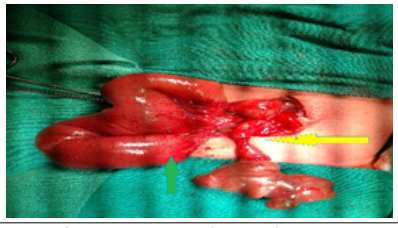

Figure two showing proximal jejunal (green arrow) and distal (yellow arrow) atresia

All of them were diagnosed postnatal. The maternal age ranged from 15 to 30 years (average, 22.5 years). The mother was primigravida in 80% of the cases. All of them were preterm delivered babies. Four infants were born by spontaneous vaginal delivery, and 2 infant was born by caesarean delivery because of fetal distress. Four of the patients were male and 2 were female. All of them were operated. Given the alarming appearance of the herniated bowel at birth, none were considered candidates for a bedside preformed silo. After resuscitation all were taken immediately to the operating room for exploratory laparotomy and careful inspection of the remnant midgut. In all cases the midgut has vanished and there was entry (jejunum) and exit (transverse colon) level atresia of bowel (figure two).

In all cases the length of small bowel from D-J (Duodeno jejunal) to entry atresia measured 10 to 15 cm(average 13.5 cm). In all cases divide stoma was fashioned. The findings of extreme short bowel and the implications of this diagnosis were discussed in multidisciplinary consultation with the family, neonatology, and gastroenterology. All of the family member denied aggressive care. Two of them died in hospital and rest were died at home.

Discussion

Closed gastroschisis is a rare congenital abdominal wall defect where the umbilical ring closes around the eviscerated bowel leading to extreme short gut.[1-5] It represent only 6% of all gastroschisis cases.[3-5] It is theorized that the abdominal wall defect can contract and even close in utero, which can lead to strangulation of the eviscerated bowel and the rare "vanishing gut syndrome." Another theory is that a volvulus occurs, which in turn leads to infarction and reabsorption of small bowel.The baby will be born with either a small (<1 cm) or no abdominal wall defect

remaining. The extrinsic bowel is then either reabsorbed or mummified. If this complication occurs, then the outcome is very poor with a mortality rate in excess of 70%.[2-7]

In all our cases there were entry and exit level atresia and the bowel in between this were mummified and useless. After excision of this mummified bowel only few centimetre of bowel left for survival.

Although multidisciplinary team had managed this cases but due to unavailability of resources and familial reluctance, all babies succumb due to short bowel syndrome and it`s related complications.

Conclusion

When gastroschisis is complicated by closure or contraction of the abdominal wall defect, resulting in Vanishing gut syndrome, morbidity and mortality is greatly increased. Treatment options are limited and highly complex. It is clear from our cases that even with surgical treatment, survival rates are still poor in resource poor country.

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