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# A RARE UNUSUAL PRESENTATION OF GASTROSCHISIS-A CASE REPORT

Obstetrics & Gynaecology

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## ABSTRACT

Gastroschisis is a congenital defect in the anterior abdominal wall resulting in herniation of the abdominal viscera without any fetal membrane covering it. It usually occurs to the right of a normally inserted umbilical cord. The anomaly is associated with intrauterine growth retardation, stillbirth, and preterm delivery. We found a preserved specimen of a 20-week-old human fetus presenting with gastroschisis in the Departmental Museum. The fetus showed a hiatus on the left side in the infraumbilical portion of the anterior abdominal wall with evisceration of the liver, spleen, coils of the small intestine, and a segment of the large intestine. The autopsy of fetus shoes gastroschisis with splenic haemorrhage.

## KEYWORDS

stillbirth, intrauterine growth retardation, premature birth, congenital anomaly, anterior abdominal wall defect, gastroschisis

## INTRODUCTION:

Gastroschisis is a congenital defect in the anterior abdominal wall leading to the evisceration of abdominal viscera such as the small intestine, stomach, colon, or gonads. The protruding viscera are devoid of any covering and thus exposed to amniotic fluid and the external environment leading to injuries and infection. The incidence of gastroschisis has increased in recent times with a worldwide incidence of 1:3,000 to 1:10,000 births. Gastroschisis has a good prognosis and patient survival if cases are adequately managed .However, there is an augmented risk of intrauterine growth retardation, premature delivery, and fetal loss associated with gastroschisis, particularly in middle- to low-income countries. In middle- to low-income countries, the associated factors with high mortality are the absence of a prenatal diagnosis, prematurity, delayed surgery, parenteral nutrition, mechanical ventilation, lack of intensive care facilities, and delivery outside tertiary care centers [5]. In high-income countries, factors associated with increased mortality are low birth weight, prematurity, and the presence of complications such as bowel atresia, necrotizing enterocolitis, and other congenital malformations. The associated malformations (incidence: 5%-20%) include limb anomalies, renal defects, and cardiac abnormalities. Gastroschisis is usually detected during serial antenatal ultrasonography. Due to the increased mortality of affected fetuses, particularly in low socioeconomic status countries, it is imperative to focus on such cases. We have attempted to highlight one such case in the current case report.

## CASE REPORT:

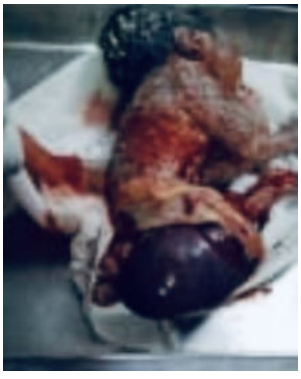

**Image 1**

A 21 years old, primigravida came to our OBG OPD With complaints of amennorrhea for 7months, presented with bleeding per vagina for past 1 hour, Patient is not booked and unimmunised. on clinical examination, Patient is conscious, oriented, afebrile, mild pallor, no pedal edema. vitals-BP-90/60mmg; PR- 120/min; Spo2-97% on room air. on per abdomen examination Uterus ~26 weeks ,mildly acting, not tense and non tender,fetal heart rate present. on prevaginal examination,Cervix uneffaced, os admits one finger,membrane intact, profuse bleeding per vaginum(++). In view of profuse bleeding per vagina and suspected abruption patient taken up for Emergency Lower segment caesarean section and delivered a still born anomalous baby of unidentified sex showing herniation of bowel through the defect in the umblical region not covered by membrane /sac-suggestive of gastroschisis shown in (IMAGE 1) .Final diagnosis made by an autopsy of fetus shows Gastroschisis with splenic haemorrhage.

## DISCUSSION:

The abdominal wall is formed by infolding of the cranial, caudal, and two lateral embryonic folds. As the abdominal wall is formed, the rapid growth of the intestinal tract leads to its migration outside the abdominal cavity through the umbilical ring and into the umbilical cord during the sixth week of gestation. By the 10th to 12th week, the abdominal wall is well formed and the intestine returns to the abdominal cavity in a stereotypical pattern that results in normal intestinal rotation and later fixation. The right paraumbilical area is an area at risk because it is supplied by the right umbilical vein and right omphalomesenteric artery until they involute. If this ordered development and involution is disturbed in degree or timing, then a body wall defect could result from the resulting body wall ischemia. The damage of the herniated intestinal loops is quite variable from the normal looking intestine to extremely damaged due to prolonged contact with the amniotic fluid, rich of gastrointestinal enzymes and waste products.

There is a possible embryological basis for gastroschisis. Gastroschisis is caused either by an occluded omphalomesenteric artery or early atrophy of the right umbilical vein before the fourth week of gestation. This results in infarction and subsequent rupture of the anterior abdominal wall, which causes protrusion of the abdominal viscera through the defect. Another theory, advocates that defective inclusion of the yolk sac in the fetal body stem leads to an additional opening in the anterior abdominal wall, and the viscera protrude outside the abdominal cavity.

Body mass index (BMI) and nutrient deficiencies in maternal dietary intake, low alpha-carotene, low total glutathione, and high nitrosamine intake during the trimester prior to conception have been associated with gastroschisis. This led to the hypothesis that younger age of mothers may lead to maternal fetal competition for nutrients with the result being maternal dietary inadequacy. Fetal growth failure in gastroschisis may be due to increased losses of protein from the exposed viscera, and inadequate supply of vital nutrients is an another factor.

Oligohydramnios is being present in up to 25% of cases. It is usually of moderate severity and associated with IUGR, fetal distress, and birth asphyxia. About 10% of gastroschisis babies have intestinal stenosis or atresia that results from vascular insufficiency in the bowel at the time of gastroschisis development or from later volvulus or compression of the mesenteric vascular pedicle by a narrowed abdominal wall ring. The differential diagnosis includes omphalocele, pentalogy of Cantrell, amniotic band syndrome, and limb–body wall complex and are easily excluded on the basis of associated skeletal and vertebral

anomalies. Gastroschisis and omphalocele are the two most common and important forms requiring differentiation.(TABLE 1)

**Table 1 Differences Between Gastroscisis And Omphalocele**

| GASTROSCHISIS | OMPHALOCELE |
|---|---|
| Location: right side | Location:centre |
| Content not covered by membranes | Presence of peritoneum amniotic membrane |
| No umblical cord | Umblical cord inserted in caudal area of hernial sac |
| Embryopathy | Fetopathy |
| Content: Intestine(100%),colon,bladder,gonads occasionaly | Content:intestine,liver(in most cases),spllen,colon,bladder(occasionally) |
| Rarely associated with other congenital anomalies(15%) | Frequently associated with other congenital anomalies(40-80%) |

One of the major techniques contributing to antenatal diagnosis of gastroschisis is to determine alpha-fetoprotein (AFP) and amniotic-fluid acetylcholinesterase levels in mother's blood. Fetal ultrasonography and/or magnetic resonance imaging brings significantly benefit to diagnosis. The fact that AFP level could also rise in fetal anomalies such as spina bifida should always be taken into consideration, thus it is recommended to determine the acetylcholinesterase/pseudocholinesterase ratio in amniotic fluid if encountered with these situations. Gastroschisis is usually detected during antenatal ultrasonography of expecting mothers around 18-20 weeks of gestation [2]. The associated risk factors are Caucasian race, Hispanic mother, young primigravida (<20 years), maternal malnutrition, prematurity of the fetus, low birth weight, exposure to nitrosamines, exposure to teratogens and agrochemicals during pregnancy, consumption of nonsteroidal anti-inflammatory drugs in the first trimester, smoking and alcohol, and illicit drug abuse.

As for babies born with anterior abdominal wall defect, the measures that must be taken and life support given after the birth immediately affect mortality and morbidity significantly. Following the birth of the baby, the segments of the intestines located outside of the abdomen should be covered by sterile wet gauze bandage or plastic film, thus baby should be kept warm and fluid, electrolyte replacement should be applied. Due to localization of the intestines outside the ventral cavity, these babies are in tendency to lose their body fluids and their body temperature tend to be lowered easily. In order to provide adequate urine output and maintain the acid-base equilibrium, fluid a few times more than the essential amount for an ordinary newborn could be necessary to replace into the babies with gastroschisis (150-200 ml/kg).

Treatment modalities such as primary closure, establishing a ventral hernia to be closed later, silastic silo are preferred recently. The prognosis is considered as good because of the frequency of having associated anomaly is low among the babies with gastroschisis. The mortality rate is known as between 5%-15%, mean 7.7%, provided respiratory circulatory insufficiency, sepsis or complications regarding total parenteral nutrition were not encountered. The survival rate is stated as 96% in cases with isolated gastroschisis treated in sufficient centers . Consequently, babies with gastroschisis represent an improved prognosis, parallel to advances in medicine in recent years. We are of the opinion that terminating these pregnancies is not a necessity. Therefore, diagnosing these babies in their prenatal early periods and transferring them to an experienced medical center containing multidisciplinary working facilities will contribute to the health of both mother and the baby.

## CONCLUSION:

gastroschisis and primary care services 17

## CONCLUSION

Gastroschisis is a congenital defect that, despite its low frequency, requires adequate knowledge not only from specialized personnel, but also from primary care physicians, taking into account that they are obliged to ensure an appropriate and timely referral of the patient to a higher complexity level to avoid complications.

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