

# Gastroschisis management and outcome in Guntur Medical College Hospital – A Retrospective study

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## ABSTRACT

**BACKGROUND:** Gastroschisis is a congenital paramedian full-thickness abdominal wall fusion defect with the visceration of abdominal contents, mostly contains small and large intestines without sac. Defect is usually to the right side of the umbilical cord. The bowel is non-rotated, without secondary fixation to the posterior abdominal wall. No genetic association reported

**MATERIALS AND METHODS:** During the study period, ninety-six patients were treated; studied as two groups, Simple gastroschisis and complex gastroschisis depending upon presence or absence of intestinal perforation, necrosis, stenosis, malrotation and volvulus and intestinal

atresia. Simple gastroschisis cases 74 (77.08%), complex gastroschisis cases 22 cases (22.91%) studied. Primary reduction of eviscerated bowel and fascial layer closure of abdominal wall was selected as initial surgical procedure in patients with stable general condition and smaller size of the eviscerated bowel mass which can be accommodated in abdominal cavity. Primary surgical procedure done in 46 patients, Silo construction and later repair done in 28 patients in simple gastroschisis group. Complex gastroschisis group 22 cases, (12 intestinal atresia cases, three perforations, necrosis of small bowel in three cases, volvulus with impending gangrene due to narrow stalk with small defect in three cases, one patient total gangrene of midgut).

**RESULTS:** Total number of cases were 96 from 2015 to 2021. Simple gastroschisis group 46 primary surgical repairs and 28 silos, Complex gastroschisis group surgical repair in 11 cases, remaining 11 cases both surgical and silo used. Silo was not used as exclusive method of management in our institute due to local prevailing conditions. Simple gastroschisis group 74 cases, 46 patients underwent primary surgical repair, 41 patients (89.13%) survived, Five deaths (10.86%), 28 patients underwent silo construction, 19 patients survived (67.85%), Nine deaths (32.14%). Over all simple gastroschisis group 60 patients survived (81.08%), 14 deaths (18.91%) recorded. Complex gastroschisis group 22 patients, 13 (59.09%) patients survived, 9 deaths (40.90%). Total patients survived in both groups 73, out of 96 patients (76.04%) with overall deaths 23 (23.95%).

**CONCLUSION:** Retrospective study of gastroschisis patients admitted in our hospital from 2015 to 2021. Total number of patients were 96; Simple gastroschisis 74 cases, 46 primary surgical repairs 41 (89.13%) survived, 28 silos with 19 patients survived (67.85%). Complex gastroschisis group 22 cases, 13 (59.09%) patients survived. Total patients survived 73 (76.04%) out of 96 patients with overall deaths 23 (23.95%). Most of the deaths occurred due to sepsis and prematurity with low birth weight. With limited facilities, Simple gastroschisis group survival good 81.08% and overall survival rate of 76.04% could be achieved in the department.

## KEYWORDS

Gastroschisis, Omphalocele, Abdominal Wall Defect, silo

## INTRODUCTION.

Gastroschisis is a congenital paramedian full-thickness abdominal wall fusion defect with the evisceration of abdominal contents, mostly contains small and large intestines without sac. Defect is usually to the right side of the umbilical cord. (1) The incidence of gastroschisis is around 1.4 per 10,000 pregnancies and is rarely associated with other congenital abnormalities. The bowel is non-rotated, without secondary fixation to the posterior abdominal wall. No genetic association reported

Embryology gastroschisis - After fertilization, cell division produces a hollow sphere, divided by a bilaminar plate with the epiblast and hypoblast, which are close to the amnion and yolk sac cavities

respectively. The hypoblast develops into the placenta, epiblast cells form the embryo. The bilaminar disc is divided axially by the primitive streak, apex of primitive streak is the primitive node. Primitive streak gives neural tube (central nervous system). Bilaminar disc becomes a disk with three germ cell layers to develop into para-axial (peripheral nervous system), intermediate (gonads and kidneys), and lateral mesoderm, which further divides into splanchnic (gastrointestinal [GI] tract along with endoderm) and somatic (body wall) with epiderm (2).

At sixth week, rapid increase in size of the liver and intestines, causes herniation of the midgut into the amniotic cavity. At 10th week, return of the midgut into abdomen due increase in size of cavity. Rotation and fixation of duodenum and ascending colon (the proximal and distal midgut) occurs upon their return. Ingrowth of mesoderm forms the urorectal septum and disintegration of cloacal membrane leads to urogenital and GI tracts outlets. Lateral folds, cephalic and caudal folds fused, causing constriction of the umbilical ring(3). Gastroschisis, may be caused by weakness in the body wall caused by failed ingrowth of mesoderm, or impaired midline fusion. Folic acid deficiency, hypoxia, and salicylates may be factors. Elevation of maternal serum alpha-fetoprotein (MSAFP) is seen in omphalocele and gastroschisis. An elevated AFP warrants ultrasonographic examination. Polyhydramnios in gastroschisis indicates associated intestinal atresia. Antenatal detected gastroschisis(4), delivery should be planned at tertiary care center. Gastroschisis mostly occurs lean, young and low socioeconomic women. Gastroschisis incidence is on increasing trend over the decades of study. Gastroschisis Classified into "simple" versus "complex" (macroscopic intestinal abnormalities) may be a reliable predictor of outcome. [5]

Sepsis the primary cause of death in more than half of these patients. Pulmonary hypoplasia and respiratory failure are two important predictors of mortality. Hypothermia, Dehydration, Hypoglycemia, In utero growth restriction(IUGR). Oligohydramnios, Fetal distress and birth asphyxia, Injury to the intestines while delivery and transport along with prematurity and low birth weight also negatively influences outcome. Improvements in antibiotics respiratory care, total parenteral nutrition, transport, anesthesia and surgery increased the survival rates up to 90%. Long-term morbidity is related to pseudo-intestinal obstruction (intestinal dysmotility), short gut, malabsorption due to mucosal injury) and gastroesophageal reflux disease, poor wound closures, ventral hernias which may require surgical repair. Improvement in parenteral and enteral nutrition, new venous access devices, catheter sepsis prevention and early treatment, surgical innovative procedures, and aggressive management of bacterial overgrowth in stagnant loops of intestine improve the outcomes.

The vertical defect is around 3 to 5 cm in size with normal umbilical attachment. There are usually the only small and large bowel protruding outside the abdominal wall. The spleen and liver protrude rarely. Other major organ system malformations so far rarely seen with gastroschisis. Intarction or atresia of the herniated bowel can be seen as complications. Proposed Mechanism are 1. omphalomesenteric artery block in utero, 2. Premature atrophy right umbilical vein with mesenchymal

damage.3. Vascular insult of the right lateral fold. Potentials suspected

Teratogens Associated with Gastroschisis are 1. Organic chemicals/solvents, 2. Cyclo-oxygenase inhibitors (Aspirin, ibuprofen), 3. Decongestants, 4. Acetaminophen, 5. Oral contraceptives, 6. Maternal smoking, 7. Alcohol, 8. Illicit drugs, (e.g. cocaine, amphetamine), and 9. X-ray irradiation in early pregnancy. The abdominal wall is developed from the fusion of four embryonic folds: cephalic, caudal, right and left lateral folds of the mesoderm origin. Gastroschisis is an embryologic abnormal development of the abdominal wall. These folds meet in the Centre of the embryo to form the abdominal wall and umbilicus. Each mesodermal fold creates a different part of the abdominal wall. The cephalic fold develops as the upper abdomen wall, the caudal fold develops as the lower abdomen wall and the lateral folds transform into both sides of the abdominal wall. Gastroschisis can be diagnosed antenatally as early as 12 to 16 weeks of gestation. Gastroschisis is an abdominal wall defect lateral to the umbilical cord, whereas omphalocele is a defect in the umbilicus where the intestines are enclosed within the umbilical cord coverings. Umbilical cord is normal in Gastroschisis. Two defects must be differentiated (6) because there is a higher incidence of major congenital/chromosomal anomalies associated with Omphalocele. Chromosomal anomalies (7) are very rare (less than 1%) in gastroschisis like normal population. Gastroschisis typically presents with a small, poorly developed abdominal cavity. Nonrotation occurs almost universally, because the intestines are outside. Cryptorchidism is a very common association up to 30 percent, Intestinal atresia's 5-10%. Mode of Delivery whether vaginal or caesarean delivery, does not have significant differences in gastroschisis outcome (8). Preoperative management of the gastroschisis patient includes factors that influence the outcome negatively: hypothermia, dehydration, gastric distention and intestinal vascular compromise, infection, hyaline membrane disease. Stabilization of the patient before the surgical repair is most important for optimum outcome. The maintenance of intravenous fluid requirement for these newborns with gastroschisis is increased 2 to 2.2 fold due to the excessive losses through the exposed bowel. Gastrointestinal decompression was done in right lateral decubitus position to enhance venous blood return from the gut. Primary closure of the abdominal defect is the aim. If primary closure cannot be done, the alternative is a staged Silo repair (9). Airway and intra-abdominal pressures kept less than 25 and 20 mmHg respectively, to prevent adverse hemodynamic consequences to other organs and tissues as well as to prevent abdominal compartment syndrome. Primary repair includes stretching of the abdominal wall, irrigating with warm saline and evacuation of meconium from bowel, evacuating the contents of the stomach, and enlarging the defect by creating a ventral hernia. Abdominal compartment syndrome (10) may result after the tight repair, leading to venous compression with compromised renal blood flow and the glomerular filtration rate resulting in decreased urine output. Urinary catheter placed; 120 to 150 mL/kg/d of isotonic solution is needed. Paralytic ileus is frequently seen in the postoperative period and may persist for several weeks. Nasogastric tube drainage has to be replaced. CPAP and Ventilatory support may be required. Enteral feeds and TPN may be considered as early as possible. Early Enteral feeding with human milk decreases time to discharge in infants following gastroschisis

repair. Postoperative TPN the minimal daily requirements are 100 kcal/kg/day, 3 to 4 g/kg/day intravenous lipids, 3 g/kg/day protein, and dextrose to maintain euglycemia. Early postsurgical management of the infant with gastroschisis includes monitoring of vital signs, fluid and electrolyte balance maintenance and pain management, cardiovascular and respiratory stabilization. After the repair, intra-abdominal pressure monitoring and observe for venous compression. A urinary catheter may be useful to relieve bladder distention and to allow for a more accurate assessment of urine output. Electrolyte balance to be maintained. A large-bore nasogastric/orogastric tube intermittent suction is needed to prevent gastrointestinal distention caused by hypoperistalsis. A dynamic ileus or hypoperistalsis is frequently seen in the postoperative period and may persist for several weeks. The early gastrointestinal drainage is characteristically green because of the stasis of biliary and pancreatic secretions in the immediate postoperative period. Once gut motility improves, the drainage becomes clear in appearance. Nasogastric/orogastric tube loses has to be replaced to maintain homeostasis. Because of the postoperative increased intra-abdominal pressure, close monitoring of respiratory status is essential for the first 48 to 72 hours. Respiratory support may be required to optimize oxygenation and ventilation. High abdominal pressure may interfere with optimal expansion of the diaphragm and venous return, impeding both ventilation and oxygenation. Some infants may benefit from Continuous Positive Airway Pressure (CPAP) or mechanical ventilation to maximize lung expansion, lung volume and oxygenation. Central Venous Pressure (CVP) must be continuously monitored during CPAP, because of its hindering effect on the CVP. Some infants may not tolerate CPAP, because of increased abdominal distention due to increased airflow to the gastrointestinal track. A properly functioning nasogastric/orogastric tube will minimize this risk. Outcomes are affected by cholestasis secondary to long-term TPN, malrotation, midgut volvulus, hypoperistalsis, Gastroesophageal Reflux (GER) and aspiration pneumonia.

The most common complications resulting in increased morbidity and mortality are intestinal atresia/stenosis, sepsis and Necrotizing Enterocolitis (NEC). Intestinal atresia is considered a poor prognostic factor in infants with gastroschisis with mortality ranging from 40% to 60%. Intestinal atresia is seen in approximately 5% to 25% of newborns with gastroschisis.

Infection is another complication associated with gastroschisis defects. Sepsis was responsible for 70% of all mortality. NEC is a common complication that occurs in approximately 15- 20% of all infants with gastroschisis. Possible late complications such as strictures, bowel adhesive obstruction and GER may be seen. Infants were classified as "simple" defect without atresia, volvulus. Infants classified as complex (11) included those infants with atresia, stenosis, volvulus "complex" gastrointestinal problems. The survival rate of low-risk infants was more than 90% compared with 60% for the high-risk infants. The most common long-term complications noted were problems related to the abdominal scar (30%) and functional gastrointestinal problems such as GER (50%).

## MATERIALS AND METHODS

Retrospective study of neonates admitted with diagnosis of Gastroschisis; presented with eviscerated edematous bowel, sepsis, respiratory distress, hypothermia, hypovolemic shock. Ninety six Gastroschisis patients were admitted during the span of seven years (January 2015 to December 2021), male patients—54 (56.25%), female patients—42 (43.75%). Pediatric surgery department in government general hospital, Guntur Medical College, Guntur is mid-level referral hospital for six districts, approximately 15 million population. All the patients presented to us within 48 hours. General observation is that most of these patients belongs to low socioeconomic group. Most of these patients come to government hospital only due to financial reasons. Those patients who presented with severe sepsis with very poor general condition and expired before any surgical intervention were excluded from study. Nine patients died before any surgical intervention done.

## METHODS

During study period ninety-six patients were treated; studied as two groups, Simple gastroschisis and complex gastroschisis depending upon presence or absence of intestinal perforation, necrosis, stenosis, malrotation and volvulus and intestinal atresia. Simple gastroschisis cases 74 (77.08%), complex gastroschisis cases 22 (22.91%) studied. Primary reduction of eviscerated bowel and fascial layer closure of abdominal wall was selected as initial surgical procedure in patients with stable general condition and smaller size of the eviscerated bowl mass which can be accommodated in abdominal cavity. Primary surgical procedure done in 46 patients, Silo construction and later repair done in 28 patients in simple gastroschisis group in which primary surgical procedure cannot be done due to poor general condition and high volume edematous thick bowel. Complex gastroschisis group 22 cases, (Twelve intestinal atresia cases, three perforations, necrosis of small bowel in three cases, volvulus with impending gangrene due to narrow stalk with small defect in three cases, one patient total gangrene of midgut). Procedure done in Complex gastroschisis. 1) Exclusively surgical repair only in 11 cases 2) Initial surgical procedure followed by silo in 7 cases 3) Initial silo followed by surgical procedure in 4 cases (missed the atresia initially). Operative technique was release of bands and gentle adhesion lysis and decompression of the bowel and reduction of gut through the existing abdominal wall defect with or without widening.

Sex	Total cases	Simple gastroschisis	Complex gastroschisis
Male	54 (56.25%)	42 (56.75%)	12 (54.54%)
Female	42 (43.75%)	32 (43.24%)	10 (45.45%)
Total	96	74	22

Table 1- Distribution of cases

Type of gastroschisis	Primary surgical repair	Silo followed by surgery	survival	deaths
Simple 74 cases-- primary surgical repair	46	-	41(89.13%)	5
Simple gastroschisis SILO followed by surgery		28	19 (67.85%)	9
Complex 22 cases-- primary surgical repair	11	-	8 (72.72%)	3
Complex gastroschisis, SILO followed by surgery		11	5 (45.45%)	6
Total cases 96	57 (59.37%)	39 (40.62%)	73 (76.04%)	23 (23.95%)

Table 2: Type of Procedure and survivals.

Primary surgical repair in simple gastroschisis 41 patients survived (89.13%). Silo followed by surgery in simple gastroschisis 19 patients survived (67.85%). Primary surgical repair cases in complex gastroschisis 8 patients survived (72.72%). Silo followed by surgery in complex gastroschisis 5 patients survived (45.45%). Simple gastroschisis group total survival 60 patients (81.08%), deaths 14 (18.91%). Complex gastroschisis group total survival 13 patients (59.09%) death 9 (40.90%).

#### Management:

Most of the babies with gastroschisis presented to us with dehydration and hypothermia. Many of these patients eviscerated bowel is covered with cotton pads, old cloths, gauze bandage and some are with meconium staining bowel. After securing reliable IV access and orogastric tube or nasogastric tube passed to decompress the stomach, IV fluids bolus 20 ml per kg was given. After stabilizing hemodynamically and hypothermia corrected, the unsterile looking dressings were removed. Removed the flakes and cotton fibers from surface of the bowel. Thorough wash was given with warm normal saline. Examined the size of the defect, status of the bowel wall whether pliable, soft or thick, edematous, stiff. We look for any intestinal atresia, gangrene bowel, impending strangulation, twisting or volvulus, necrosis of bowel, bowel and mesentery tears. Colonic irrigations were done with warm normal saline to evacuate meconium in the colon along

with milking of meconium in midgut with sterile precautions. We covered the newborn once they reach us, by sterile dressing with sterile plastic bag then assessment was made whether to do primary closure with reduction of bowel or Silo construction followed by surgical repair. Stabilization of the patients from hypothermia, hypovolemic shock was done. Cleaning of the eviscerated bowel with warm normal saline. Broad spectrum antibiotics given. Injection ceftriaxone 100 mg per kilogram, Inj. amikacin 15 mg Inj. Metronidazole 2 mL per kilogram, injection vitamin K one mg stat, ranitidine 5 mg given. NGT inserted and decompressed the stomach and small bowel. Rectal wash with warm normal saline to evacuate meconium. Preoperative evaluation for associated anomalies and basic surgical workup done including hemogram, blood grouping and typing. Throat suction done and oxygen was given by mask administered. Intravenous fluids Isolyte P given 150 ml per kg. Corrected electrolyte imbalances. Kept under warmers. Monitored vital signs

<b>Weight Range</b>	<b>No of patients</b>	<b>male</b>	<b>female</b>
<b>1kg to 2.5 kg</b>			
<b>Less than 1.5 kg</b>	<b>23</b>	<b>14</b>	<b>9</b>
<b>1.5 kg to 2.0 kg</b>	<b>60</b>	<b>32</b>	<b>28</b>
<b>2.0 kg to 2.5 kg</b>	<b>13</b>	<b>8</b>	<b>5</b>
	<b>96</b>	<b>54</b>	<b>42</b>

Table 3- weight and sex details. (12)

All patients presented within 48 hours

Antenatal detection was noted in 16 patients, male 9 patients, female 7 patients.

Gestational age	No of patients	Male	Female
Less than 28 weeks	6	3	3
28-32 weeks	34	18	16
32-36 weeks	<b>51</b>	31	20
More than 36 weeks	<b>5</b>	2	3
Total	<b>96</b>	54	42

Table- 4 Gestational agedetails (13).

Data collected regarding mode of delivery shows 72 cesarian sections, 24 vaginal deliveries. One patient had family history of gastroschisis in sibling who was successfully operated and surviving without any problems

### **Surgical Management**

Primary reduction of eviscerated bowel and repair of abdominal wall was selected as initial surgical management in 46 patients.

Silo construction was selected as initial procedure in 28 patients.

Silo was selected when primary repair was not feasible because of size of the edematous bowel and poor general condition in simple gastroschisis group.

### **Primary Reduction and Repair**

Under general anesthesia, bowel was cleaned with warm saline and flakes over bowel were cleared. Bands released and adhesion lysis was done. Decompression of the bowel and reduction of gut through the existing abdominal wall defect without widening if bowel is less edematous and pliable. If bowel is grossly edematous and stiff, extending the incision along midline Linea Alba was the primary modality of management; 3-0 vicryl and 3-0 prolene was used for repair. Total of 57 patients underwent primary reduction and repair (46 simple gastroschisis, 11 complex gastroschisis).

### **SILO Reconstruction**

39 patients underwent Silo reconstruction as primary reduction and repair was not possible due to large volume of edematous bowel and small abdominal cavity (28 simple gastroschisis, 11

complex gastroschisis). Silo was constructed with Prolene mesh on the outside and Urobag inside or double mesh (vicryl-prolene mesh). Mesh was anchored to the abdominal wall with 3-0 prolene. Every alternate day, reduction of the size of Silo was done in NICU and primary muscular closure or skin closure was done when size of abdominal cavity was adequate.

Hospital stay in simple Gastroschisis ranging from 12 days to 48 days, mean hospital stay was 24.6 days. Complex Gastroschisis cases hospital stay ranging from 18 days to 94 days, mean hospital stay was 48.6 day. Trial feeding started 6<sup>th</sup> day onwards with parenteral nutritional support with amino acids and fatty acids solutions (Astymin 3 and intralipid 20%). umbilical vein was used for infusion. Ventilatory support was given in simple Gastroschisis (24 patients) ranging from 2 days to 34 days, mean support 5.4 days, in Complex Gastroschisis Ventilatory support (18 patients) ranges from 5 days to 42 days, mean 16.2 days support. No CPAP support given in our institute.

## Discussion

Gastroschisis is a full-thickness abdominal wall defect on right paraumbilical region without a covering membrane. Gastroschisis rarely associated with malformations (14) outside of the gastrointestinal tract. Gastrointestinal problems (stenosis, intestinal atresia, perforation, necrosis, volvulus, malrotation) may present in up to 25% of cases. Maternal serum alpha-fetoprotein (AFP) is increased in gastroschisis. Prenatal ultrasound shows free-floating bowel loops and paraumbilical abdominal wall defect. Mode of delivery has no significant effect on outcome of gastroschisis. Trial of labor rather than cesarean birth can be tried for, except the liver is significantly herniated. Rarely intrauterine growth restriction (IUGR) may be associated. Infants with gastroschisis have the most favorable prognosis and with excellent long-term outcomes when compared to other abdominal wall defects. Overall survival rates are 80 to 95 percent in gastroschisis neonates born. Complex gastroschisis have more severe gastrointestinal, respiratory, and infectious complications in the neonatal period with high in-hospital mortality, bowel obstruction, short bowel syndrome, necrotizing enterocolitis, need parenteral nutrition, and tube feedings on discharge. Complex gastroschisis also likely to have a longer hospitalization. Most newborns with gastroschisis have an intestinal rotational anomaly. The incidence of volvulus is higher in omphalocele than gastroschisis. Intestinal adhesions and adhesive bowel obstruction are more common in gastroschisis. Undescended testis is associated with 15% to 30% of gastroschisis cases with orchiopexy in the first year of life. Cosmetic umbilicoplasty or umbilical reconstruction surgery can be considered for psychosocial stress if umbilicus not present. Neurodevelopment, learning issues, and overall health-related quality of life to be within a normal range in these children. Preterm delivery is common with gastroschisis up to 28% compared to normal 6%. Complications can occur with total parenteral nutrition (TPN) and line sepsis, necrotizing enterocolitis (NEC), and abdominal wound infections. If associated with intestinal complications such as atresia, ischemia, perforation, or development of necrotizing enterocolitis consider as complex gastroschisis and have a higher mortality rate, may need multiple operative interventions, and prolonged hospitalization, high rates of sepsis, and

prolonged cholestasis and intestinal transplantation due to intestinal failure. Approximately 10% of newborns with gastroschisis have associated atresia, mostly jejunal or ileal, will have significantly worse outcomes. The timing of surgical management of the atresia depends on the state of the bowel wall inflammation to hold sutures well. Early versus late operations (before or after 21 days of life) for intestinal atresia, no significant difference in outcomes and potential for early feeding between the two groups noted. Team work of specialists, including an obstetrician, pediatric surgeon and neonatologist essential for optimal outcome. The best time for delivery may be near term. Controversies in management include primary surgical closure or sutureless closure, mode of delivery, timing of delivery, silo versus surgical closure, time of treating intestinal atresia. The outcomes of gastroschisis depend on the birth weight, prematurity, comorbidities, and the state of the bowel (atresia, vanishing gastroschisis, vascular compromise of bowel).

**CONCLUSION:** Retrospective study of gastroschisis patients admitted in our hospital from 2015 to 2021. Total number of patients were 96; Simple gastroschisis 74 cases, 46 primary surgical repairs 41 (89.13%) survived, 28 silos with 19 patients survived (67.85%). Complex gastroschisis group 22 cases, 13 (59.09%) patients survived (15). Total patients survived 73 (76.04%) out of 96 patients with overall deaths 23 (23.95%). Most of the deaths occurred due to sepsis and prematurity with low birth weight. With limited facilities, Simple gastroschisis group survival good 81.08% and overall survival rate of 76.04% could be achieved in the department.

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Clinical presentation of children with gastroschisis and small for gestational age

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