# Left-Sided Congenital Abdominal Wall Defect and Intestinal Malformation-A Rare Case

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***Abstract***:
*A case report of a female newborn with a rare congenital abdominal wall defect associated with intestinal malformation. To our knowledge, only a few case reports of left sided congenital abdominal wall defect with this type of anomalies have been reported in the world literatures and this is the first in Fiji. This case brought numerous challenges to the team in terms of defining the pathology, role of undertaking surgery, providing supportive and nutritional therapy to a neonate and the ethical dilemma with the management in a developing country.*

***Key words:*** *congenital, abdominal wall, left defect, short gut syndrome*

***Abbreviations:*** *BWT-birth weight, SGS-short gut syndrome, TPN-total parenteral nutrition*

## Case Presentation

A female newborn baby (BWT-1.58kg) was delivered by emergency caesarian section to a 23 year old Indo-Fijian at 36 weeks of gestation. The mother had a normal first trimester foetal scan but repeated scan in the third trimester detected polyhydramnios with ventral abdominal wall defect. At birth, a non-dysmorphic child with normal respiratory and cardiovascular systems and who passed urine soon after delivery. She was confirmed to have a rare left-sided abdominal wall defect. This abdominal wall defect was located on the left lumbar region about 3cm from the umbilical cord.

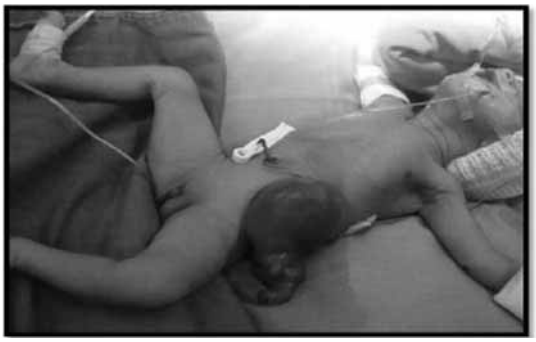

**Figure 1: Showing the eviscerated bowel loops and stomach on the left side of the abdomen away from the umbilicus**

The defect was a rounded and measured 5 x 4cm in diameter, lateral to the rectus abdominis with eviscerated stomach and bowel (Figure 1) without any peritoneal or amniotic cover. The eviscerated bowel was quite oedematous and thickened with a tapered end which was draining bile and saliva. The middle segment of the evisceration had what looked like a duplication of the small bowel or appendix. There were no other associated anomalies seen pre-operatively. Her anus was patent in the normal anatomical position with normal anal tone. Pre-operative echocardiogram and a renal ultrasound scan were normal and no further contrast study was done.

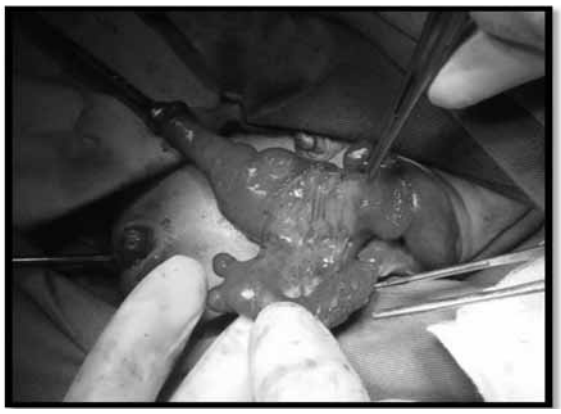

**Figure 2: Showing the stomach (top right hand side marked by the forcep) being separated from the colon (inferior loop being held). Also had duplicated appendix on the caecum**

Intra-operatively (figure 2), most of the stomach and bowels were encased within the eviscerated mass including part of the reproductive organs. The proximal small bowel extending from the stomach to the tapered atretic opening was 9cm. The duplication of the appendix was confirmed with the length of large bowel from the caecum measured 12cm. This was associated with a second distal atresia from the rectum to the anus which measured 6cm. In total, only 27cm of bowel length was identified in this neonate. This is Short-gut syndrome (SGS) which is incompatible with survival without parental nutrition.

The abdominal contents were reduced with formation of a proximal stoma from the proximal atresia. The abdominal defect was then closed primarily with minimal tension. She was ventilated for 8 days post operatively and oral gastric tube feeds was commenced on Day 6. After 21 days in the hospital, her mother took her home and she later succumbed on Day 24 of life.

## Discussion

Fiji has a population of about 880,000 people and has approximately 20,000 deliveries per year. Paediatric surgical service in our institution at the Colonial War Memorial Hospital (CWMH) in Suva encounters great difficulties and challenges due to the lack of other supporting service. With limited resources such as the lack of a paediatric nutritional service, having Total Parenteral Nutrition (TPN) is a major hurdle in the management of abdominal wall defects with or without SGS.

Congenital abdominal wall defects in general have an incidence of approximately 2.15cases/10 000 live births.[1] The most common defects are midline defects such as gastroschisis (usually right-sided) and exomphalos (omphalocoele) having a prevalence of approximately 1.35/ 10 000 and 0.77/ 10 000 live births respectively.[1] With left sided abdominal wall defects, left sided-gastroschisis are more common and these have been

reported in 16 cases in the world literatures.[4,5,6] These left-sided gastroschisis are periumbilical defect, similar to a right-sided gastroschisis. Aetiology for these left sided gastroschisis have been postulated as a result of either an involution of the left umbilical vein,[2] or malpositioning of yolk stalk to left of the midline in wall fold failure.3 Associated intestinal anomalies include malrotation, atresia and SGS. As high as 40% of cases would association with extra-intestinal congenital abnormalities.[4]

This large abdominal wall defect seen in our patient cannot be described as a left-sided gastroschisis. It was rounded in shape and it was lateral to the rectus abdominis. The abdominal wall defect was distant from the umbilicus and these are few in the literature.[5] A Medline and Pubmed internet search only revealed 3 other case reports of similar left-sided abdominal wall defect which are distant from the umbilicus. This is a separate congenital pathology and it is important to differentiate these from a left-sided gastroschisis.

The possible embryology of this rare anomaly remains unknown[6]. The principles of management on the other hand should be similar as to how we manage gastroschisis with its associated anomalies. Our case underwent a Laparotomy to assess the content of the eviscerated mass through the left-sided abdominal wall defect. Multiple small bowel atresia's with SGS was noted. In our institution, having a SGS is not compatible with life and is partly due to the unavailability of a better nutritional service where TPN could be provided. In our case, she later developed complications of under nutrition, septicaemia and died at Day 24 of life.

Outcomes in any gastroschisis are determined by many factors such as the exteriorized bowel and the mortality rate should be around 8%[7] or less in developed countries. This is supported by the improvement in the surgical care, improvement in the neonatal intensive care, better neonatal support and better nutritional service with the availability of TPN. These TPN has allowed many children with SGS to survive whom would have died as a result a result of poor enteral absorption and malnutrition. In our institution, none of the children born with SGS has survived.

## Conclusion

Left-sided gastroschisis is an uncommon abdominal wall defect. This case of a Left-sided abdominal wall defect cannot be classified as a gastroschisis. This is extremely rare and it's difficult to name this type of defect. This is our sole experience with a congenital left-sided abdominal wall defect that not only was a diagnostic dilemma but also challenging to treat and manage. Such complex congenital cases with SGS in our institution have poor outcomes despite the best of facilities and expertise available.

*"Silence is organized knowledge. Wisdom is organized life."*

*Immanuel Kant*