PST | PEDIATRIC SURGERY IN TROPICS

WAUGH SYNDROME: AN INTERESTING CLUSTER OF PRESENTATIONS AT A TERTIARY INSTITUTION

Eliyah Aaron Sischy[1], Matlhatse Phuti Maluleke[1], Nooreen Amod[2], Alireza Dehghan-Dehnavi[2], Elliot Motloung[1]

[1]Department of Pediatric Surgery, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University, Molotlegi Street, Ga-Rankuwa Zone 1, Gauteng, South Africa, 0208

[2]Department of Diagnostic Radiology, School of Clinical Medicine, Faculty of Health Sciences, Sefako Makgatho Health Sciences University, Gauteng, South Africa

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Keywords	**Abstract**
Waugh Syndrome, intussusception, intestinal malrotation, intestinal obstruction	**Introduction:** Waugh Syndrome is a rare pediatric condition characterized by the synchronous occurrence of intussusception and intestinal malrotation. Both conditions are known causes of intestinal obstruction in children, but their coexistence poses a unique diagnostic and surgical challenge. The underlying pathophysiology is thought to involve abnormal fixation of the mesentery and defective intestinal rotation during embryogenesis, predisposing patients to intussusception. This study aims to describe the clinical presentation, diagnostic approach, surgical management, and outcomes of three pediatric patients diagnosed with Waugh Syndrome at our institution. Our objectives include identifying key diagnostic features, assessing the efficacy of surgical interventions, and contributing to the limited literature on this rare condition. **Method:** We conducted a retrospective case series review of three patients diagnosed and treated for Waugh Syndrome between July and

	November 2024. Clinical presentation, laboratory findings, imaging results, surgical intervention details, and postoperative outcomes were analyzed. **Results:** All three patients presented with symptoms of intestinal obstruction, including bilious vomiting, abdominal distension, and bloody stools. Diagnostic imaging revealed features of intussusception and malrotation. Surgical intervention involved laparotomy with manual reduction of intussusception and Ladd's procedure. Two patients required intensive care postoperatively, while one was managed in the surgical ward. All patients recovered well and were discharged with outpatient follow-up. **Conclusion:** Waugh Syndrome remains a potentially underdiagnosed entity that should be considered in pediatric patients with intussusception. Early recognition and surgical management are crucial to improving outcomes, and further studies are needed to enhance diagnostic accuracy and treatment strategies.
Abbreviations	CT: Computed Tomography ICU: Intensive Care Unit NGT: Nasogastric Tube

INTRODUCTION

Waugh Syndrome is a rare pediatric condition characterized by the synchronous occurrence of intussusception and intestinal malrotation. Although both intussusception and malrotation are known etiologies of intestinal obstruction in the pediatric population, the synchronous presentation of these 2 pathological conditions with different etiologies in a single patient is a rare phenomenon. It is postulated that the combination of poor fixation of the ascending and descending colon to the retroperitoneum, as first described by Waugh, along with abnormal intestinal rotation and fixation of the mesentery, may serve as predisposing factors for intussusception in patients with malrotation. [1-3] Waugh Syndrome can also present with additional diagnostic challenges such as Meckel's diverticulum, acute appendicitis and other pediatric gastrointestinal pathologies. [4]

Intestinal malrotation is described as any variation during embryogenesis from the normal 270° anticlockwise rotation of the midgut around the axis of the superior mesenteric artery. [2,5] This results in abnormal positioning of the small intestine and improper fixation of the mesentery, subsequently predisposing patients to develop intussusception. Intestinal malrotation is a common defect of embryological intestinal development observed in 1 in 6000 live births. [5] A majority (64-80%) of these cases present within the first month of life, and 90% by the first year. Symptomatic presentations usually involve midgut volvulus and obstruction secondary to the malrotation. [5,6] However, it is worth noting that some cases remain asymptomatic and are thus undiagnosed.

Intussusception is defined as telescoping of one section of intestines into the section immediately ahead of it. [7] In the pediatric population the most common etiology seen is idiopathic ileocolic intussusception. Intussusception is also 1 of the most common causes of pediatric intestinal obstruction, predominantly affecting children under the age of 2 years. [2] The peak incidence occurs between the ages of 5 and 9 months, with males being affected approximately twice as much as females. [8]

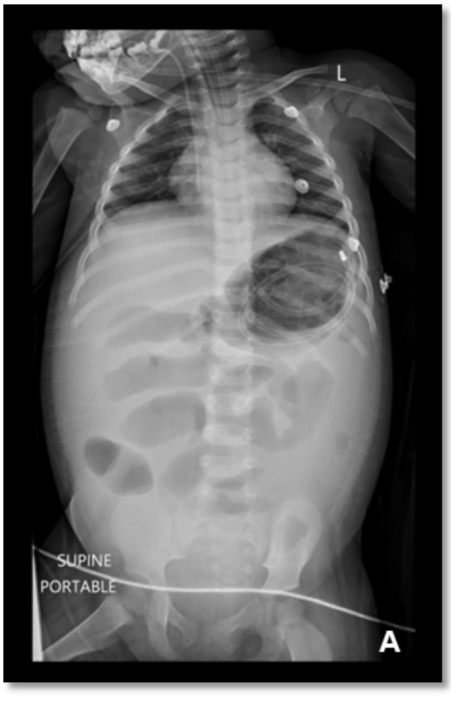
CASE REPORT

We diagnosed 3 patients with Waugh Syndrome at our facility between July and November 2024.

Patient A, a 1-year 7-month-old female, initially presented to a district hospital before being referred to our facility for specialist care. The guardian reported a 6-day history of fever, loss of appetite, intermittent colicky abdominal pain, bilious vomiting, and a 3-day history of mucoid bloody stools. On examination, the patient had a soft, distended abdomen and bilious nasogastric aspirates. Additionally, the guardian mentioned the use of unknown herbal medicine prior to the hospital visit, which contributed to the delayed presentation. The patient was otherwise previously well with no known chronic illnesses. The prolonged history of symptoms and herbal ingestion resulted in a diagnostic dilemma.

Patient B, a 3-year-old male, presented to our clinic with a 1-day history of vomiting, loss of appetite, fever and abdominal distension. He was known to our unit, having presented 2 years prior with intussusception requiring a laparotomy and right hemicolectomy post manual reduction. On examination, the patient had a distended and tender abdomen, with coffee-ground aspirates draining from the nasogastric tube.

Patient C, a 6-month-old female, was referred to our facility from a district hospital. The guardian gave a 2-day history of vomiting, diarrhea and colicky abdominal pain, which then progressed to mucoid bloody stools and a rectal mass. On examination, the patient's abdomen was distended and firm, with a prolapsed intussusception.



Biochemically, patient A had severe metabolic acidosis, acute renal injury, elevated septic markers and deranged liver function tests. A plain abdominal film demonstrated a distended stomach and dilated loops of small intestines. (Fig 1) These intestinal loops were central in location and there was no intestinal gas noted within the rectum. In addition, there was slight bulging of the flanks and ascites.

Fig 1. Abdominal X-ray of Patient A, with coiled NGT in the stomach

Due to extensive intestinal gas, there was reduced diagnostic yield of the abdominal ultrasound. Subsequently, a contrast enhanced Computed Tomography (CT) scan of the abdomen was performed, which demonstrated dilated loops of small intestines with air fluid levels. These intestinal loops demonstrated mural enhancement. The large intestinal loops were collapsed. Additionally, a "bowel within bowel configuration" was noted at the level of L3/L4. This refers to a pattern of concentric alternating layers of intestines suggestive of an intussusception. The intussuscipiens contained fat and mesenteric vessels, while the intussusceptum contained rounded enhancing nodules of soft tissue density suggestive of lymph nodes. (Fig 2-4)

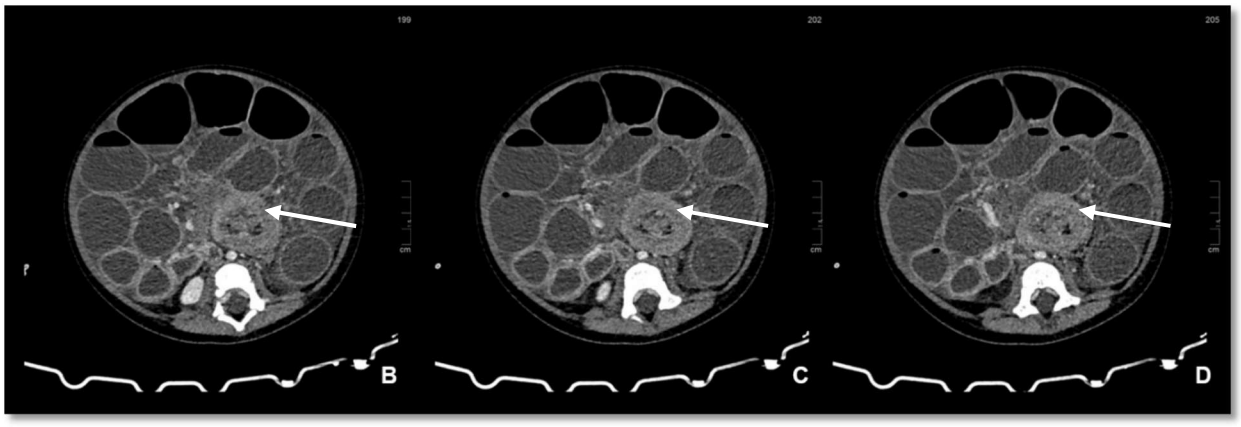


Fig 2-4 (labeled B, C and D). Contrast enhanced CT abdomen of Patient A, arrows demonstrate the bowel within bowel configuration

Features of malrotation were also appreciated on the CT abdomen. The duodeno-jejunal

junction was located to the right of the midline at the level of L2/L3. The superior mesenteric vein was noted anteriorly and slightly to the left of the superior mesenteric artery at the level of L2. Additionally, there was free fluid noted in the abdomen and pelvis, but no pneumoperitoneum or pneumatosis noted.

Patient B presented with a metabolic alkalosis that improved with fluid resuscitation. We were unfortunately unable to retrieve the imaging from the index presentation in 2022. Plain abdominal radiograph on this admission showed dilated loops of small intestine and features suggestive of intestinal obstruction.

Patient C was initially resuscitated before x-rays and ultrasound were done. The abdominal radiograph demonstrated proximal colon dilatation in the upper abdominal region with paucity of gas in the lower abdomen and pelvis. (Fig 5) There is no visible free air. The visualized chest is normal, with 11 pairs of ribs bilaterally.

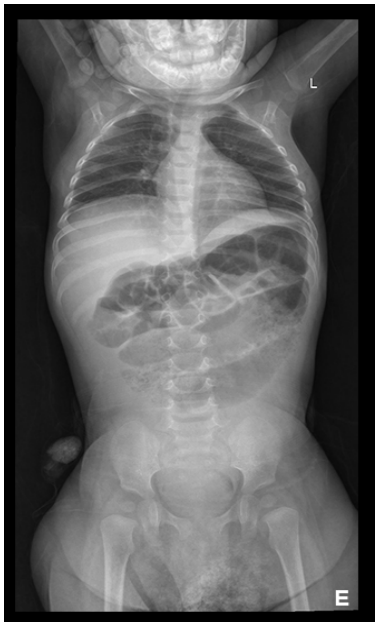


Fig 5. Chest and Abdominal X-ray of Patient C

An abdominal ultrasound was done which showed telescoping of one intestinal loop into another intestinal loop in the left inguinal fossa region. The intestines form an appearance of a target sign in the transverse plane and pseudo-kidney sign in the longitudinal plane. (Fig 6) The central core of intestines is hyper-echoic, and the outer wall is hypo-echoic. Doppler shows flow in the intussusceptum and intussuscipiens. No free fluid is noted within the intussusceptum.

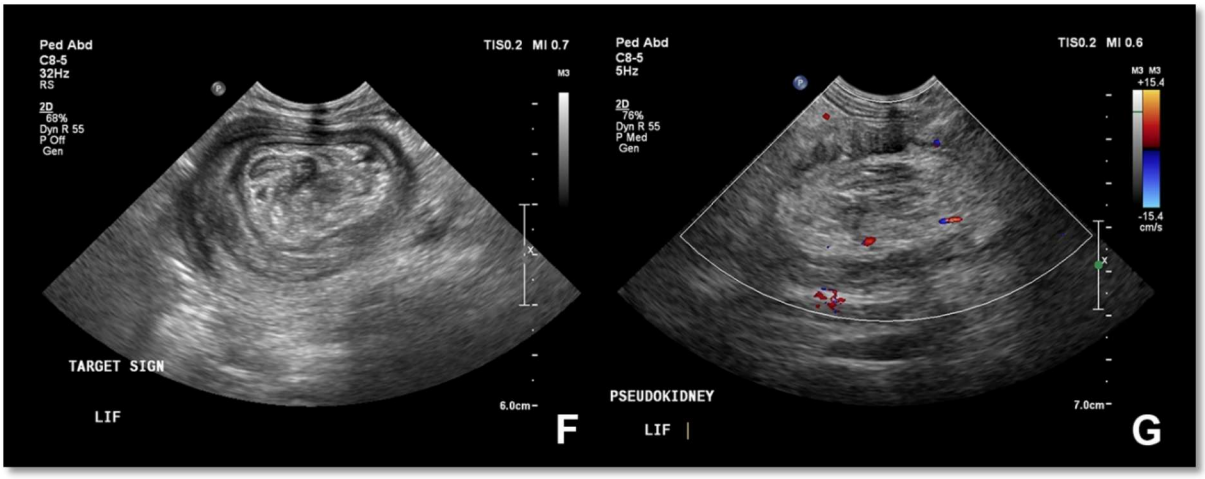


Fig 6. Abdominal ultrasound of Patient C

Patient A's condition was complicated by refractory hypovolemic shock and severe abdominal distension causing splinting of the diaphragm, which compromised the respiratory system and required preoperative ventilation. The patient was resuscitated and taken for an emergency laparotomy. A manual reduction of an ileocolic intussusception was successfully performed, revealing no intestinal necrosis or perforation. (Fig 7)

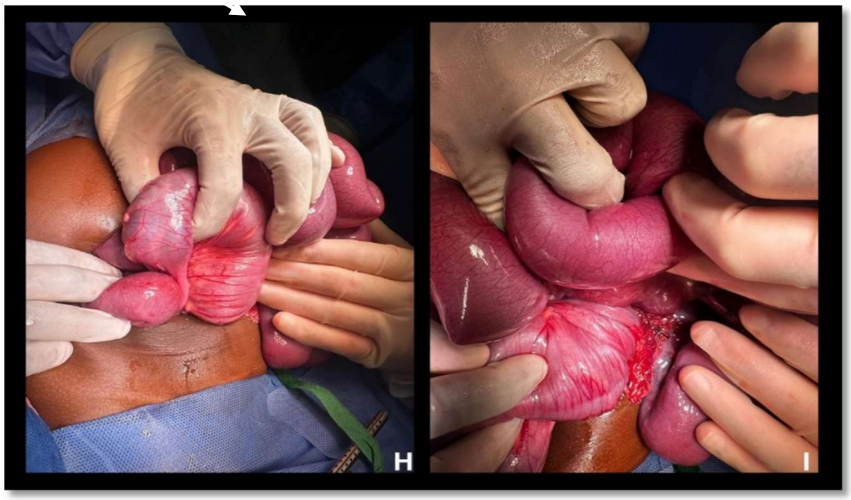


Fig 7. Intra-operative findings, Patient A

Further exploration identified an intestinal malrotation with midgut volvulus. (Fig 8) A Ladd's procedure was therefore performed, and the patient was transferred to the pediatric intensive care unit for postoperative care.

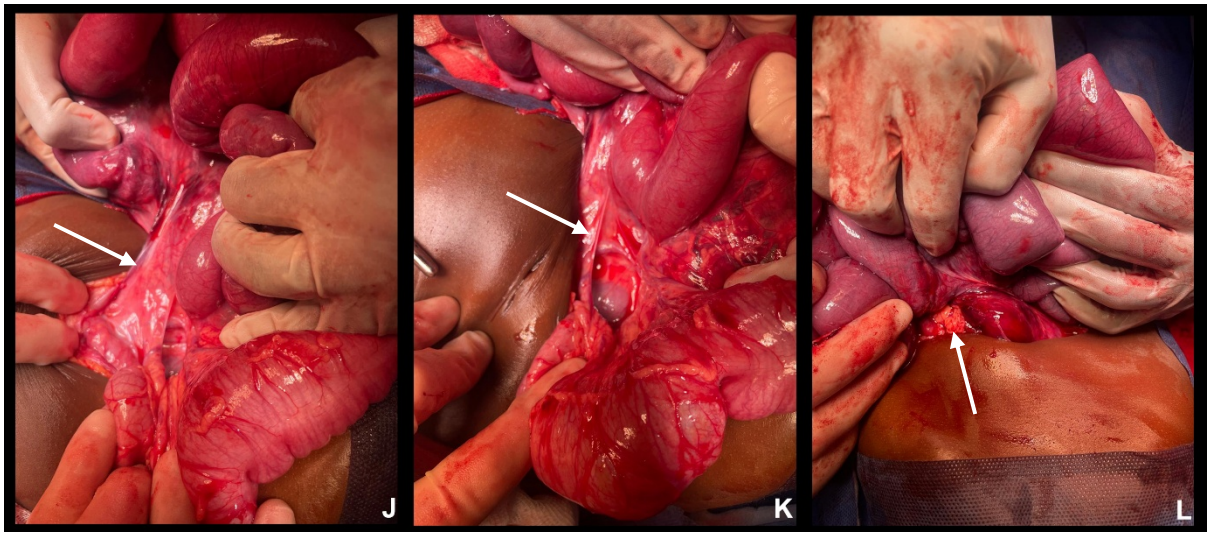


Fig 8. Narrow mesenteric base, Patient A

After initial resuscitation, Patient B underwent an exploratory laparotomy. A malrotation with a midgut volvulus was identified, with segmental necrosis of the affected area of small intestine. (Fig 9) After detorting the small intestine and performing a resection and primary end to end anastomosis, Ladd's procedure was performed for the narrow mesenteric base. (Fig 9)

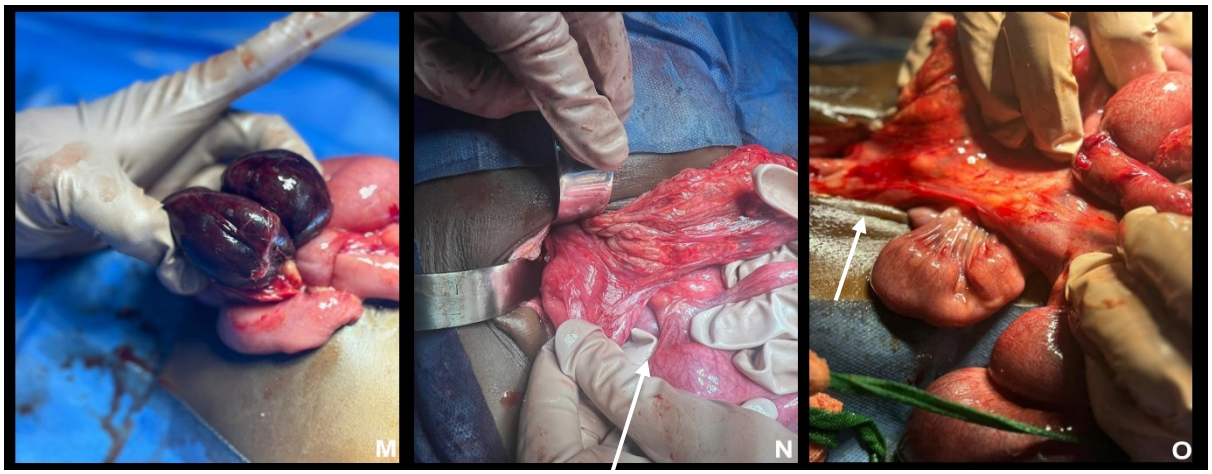


Fig 9. Intra-operative findings, Patient B: Midgut volvulus with necrotic bowel with a narrow mesenteric base

Patient C was resuscitated then taken for an emergency laparotomy. The prolapse and ileocolic intussusception were reduced, and upon examining the intestines it was found that the caecum and appendix were in the left upper quadrant of the abdomen. (Fig 10) The ascending colon was also found to be unattached, and Ladd's bands were present. After a Ladd's procedure, the patient was transferred to the pediatric intensive care unit for further care.

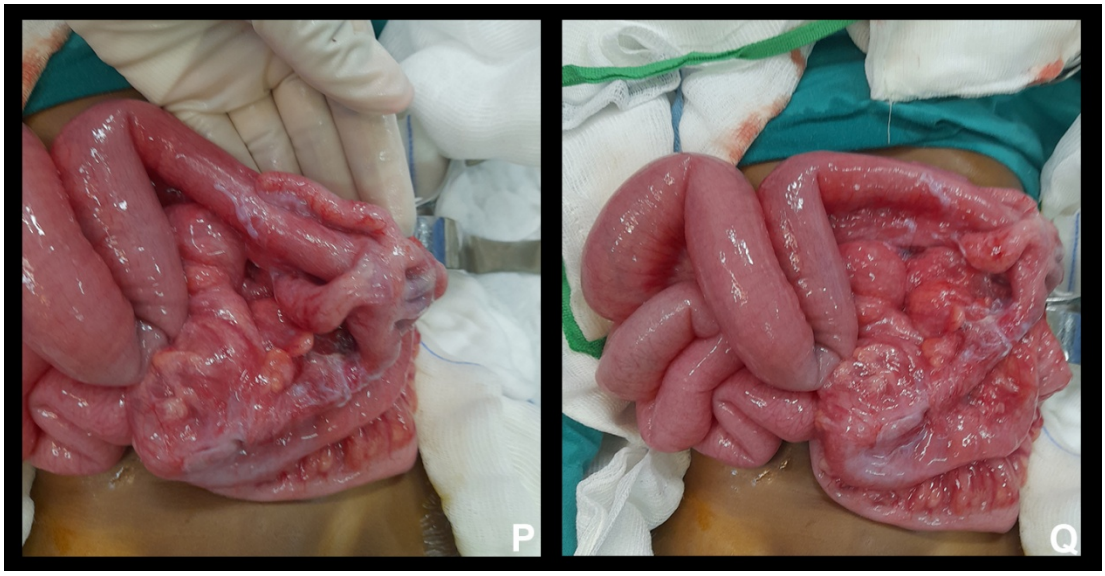


Fig 10. Intra-operative findings, Patient C: Intestinal malrotation with appendix and cecum in the left upper quadrant

Patients A and C spent a total of 6 days in the pediatric intensive care unit (ICU) after surgery, receiving treatment for metabolic acidosis and distributive shock. The ICU care involved a multidisciplinary team, including pediatric intensivists, ICU nurses, pediatric surgeons, dietitians and physiotherapists. After stepping down from the ICU, patients A and C spent an additional 3 and 2 days respectively in the pediatric surgical ward.
Patient B was cared for in the pediatric surgical ward for 8 days post-surgery. They required a transfusion for postoperative anemia and recovered well in the ward.

Throughout the admission and before discharge, the guardians received extensive counselling regarding the patients' conditions and outpatient follow up. All patients have been healthy without complications at their outpatient visits.

DISCUSSION

Since its initial description in 1911 by Waugh, and its name as Waugh Syndrome by Brereton et al., in a prospective study in 1986, fewer than 100 cases have been reported in medical literature. [1,3,9] In the South African setting, Breckon and Hadley identified and managed 6 patients with the condition at King Edward Hospital in Durban over a period of 4 months in 1998.[6] Another significant cluster of 4 patients were seen by Elkeir et al. at Khartoum North Teaching Hospital in Khartoum, Sudan over the course of 6 months in 2021.[2]

The most common approach today for definitive management involves laparoscopy or a laparotomy for manual reduction of the intussusception, followed by a Ladd's procedure for the malrotation. [2,3,10]

The scarcity of data on Waugh Syndrome can potentially be attributed to the fact that most patients are successfully managed nonoperatively. [4] Many authors consider Waugh Syndrome to be more common than recognized. Therefore, surgeons should keep this in mind when performing open surgery, pneumatic reduction of intussusception, and in cases of recurrent intussusception. [2-4,6]

CONCLUSION

Waugh Syndrome remains a potentially underdiagnosed entity that should be considered in pediatric patients with intussusception. Early recognition and surgical management are crucial to improving outcomes, and further studies are needed to enhance diagnostic accuracy and treatment strategies.

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