



## Case report

## An unusual cluster of Waugh syndrome as a cause of intestinal obstruction in children - A case series

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## ABSTRACT

**Introduction and importance:** The association of intussusception with intestinal malrotation is a rare phenomenon named Waugh syndrome by Brereton et. It is a rare disease entity with less than 100 cases described so far. A cluster presentation is something unusual for rare diseases.

**Case presentation:** All four patients have presented with features suggestive of bowel obstruction as all of them have had colicky abdominal pain. Three patients have vomiting as an additional complaint, so as the diarrhea. Fever has also been reported in one patient. Patients have presented in 6 months period, non-consecutively. Abdominal pain, diarrhea and vomiting are alarming in the pediatric population. When each case presented, an immediate resuscitation was performed including IV lines, nasogastric suctioning, electrolytes correction, and blood and FFP transfusion as necessary. Abdominal ultrasonography was performed revealing target sign. After optimization for surgery, manual reduction of the intussusception was done followed by a Ladd's procedure for the malrotation was performed for all 4 patients with excellent outcomes.

**Clinical discussion:** With the surgery, after case optimization, remains the standard of care; the presence of cluster presentation for Waugh syndrome could suggest that some environmental factors might be implicated and further observations can be carried out to look more into such phenomenon.

**Conclusion:** Waugh syndrome, a combined presentation of intestinal malrotation and intussusception, is a rare syndrome that can present in children. Optimization for surgery followed by a 2-step surgical approach is the usual treatment with excellent outcomes described.

## 1. Introduction

Intussusception is one of the most common causes of pediatric intestinal obstruction. Although it can affect children at any age, it mainly affects children less than 2 years, with only 10–25% cases happen in children older than that [1]. It has its major incidence in the age group between 5 and 7 months of age [2]. Malrotation of the gut is a condition in which there is an error in the normal rotation of the midgut during the intra-uterine period (Normally the rotation is 270° anti-clockwise which can result in abnormal mesenteric fixation. Intestinal Mal-rotation (IM) is defined as any deviation from the normal 270° counter clockwise rotation of the midgut during embryological development, resulting not only in the mal-position of the small intestine but also in the mal-fixation of the mesentery [3].

The association of intussusception with intestinal malrotation is a

rare phenomenon. It is named Waugh syndrome (WS) by Brereton et al. on their prospective study. They have named this association as WS because it was first mentioned in the literature in early twentieth century by George E Waugh. [4,5]. In a rare disease entity like Waugh syndrome in which less than 100 cases reported so far, a cluster presentation of 4 patients in a single center in less than 6 months; although have been described once in the literature, is still an unusual incident. Expanding the literature and knowing more about the syndrome presentation, management and outcome is crucial (Figs. 1 and 2).

With paucity of the reported cases, no clear guidelines have been established. Preoperative optimization including intravenous fluids and fresh frozen plasma along with electrolytes correction are necessary before surgical intervention. Open surgery for the purpose of manually reducing the intussusception followed by a Ladd's procedure for intestinal malrotation is the usual and most commonly used treatment

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method. This case has been reported in line with the SCARE criteria.

## 2. Patient information

We report a case series of 4 children presented to the emergency department with features of intestinal obstruction in a single teaching hospital, Khartoum North Teaching Hospital – KNTH, located in Khartoum, Sudan. KNTH is a teaching and public hospital and one of the major pediatric surgery centers in the country. Their ages were very diverse, which have ranged from as young as 8 months up to as old as 12 years old. They have no significant family or genetic history. Families of 2 children reported previous presentation for which they have not sought medical care. No significant drug history was found for either patient and none of these four patients has any known genetic diseases. All of our patients belong to the low socioeconomic state.

## 3. Clinical findings

With regard to their clinical presentation, all of our patients have presented with colicky abdominal pain. Three patients have vomiting as an additional complaint so as the diarrhea. Their diarrhea was bloody in nature in two children and watery in only one child. Fever has been reported in one child (Table 1).

## 4. Timeline

Patients have presented between in the period from 1st January – 30th June 2021, non-consecutively and managed with surgery after successfully been optimized for it. All of our four patients have presented, assessed and managed in a single teaching hospital, Khartoum North Teaching Hospital – KNTH, located in Khartoum, Sudan. When each case presented, an immediate resuscitation was performed if necessary and patients were then optimized for surgery.

With regard to the presentation timeframe, the first 2 patients have a history of greenish vomiting few months and 2 years before their

presentations, respectively. They have otherwise no other complaints and no chronic illnesses. The families of the latter two patients have not reported previous presentation like this in their children.

## 5. Diagnostic assessment and interpretation

All patients have been admitted to the ward prior to surgery for optimization where IV lines and nasogastric tubes were inserted. All patients have received fluid replacement for their dehydration as well as fresh frozen plasma (FFP). Two patients received blood for low hemoglobin. Associated electrolytes abnormalities were corrected for both sodium and potassium. Abdominal imaging in form of ultrasonography was performed which showed a target sign that was suggestive for intussusception for all four cases. Renal function tests and urinalysis for all four patients were normal.

## 6. Intervention

After optimization for surgery with IV fluids, nasogastric suctioning and FFP, patients have undergone laparotomies. When observing their abdomens, a midgut malrotation was observed in addition to the intussusception. Manual reduction of the intussusception was done followed by a Ladd's procedure for the malrotation. Hypertrophied mesenteric lymph nodes were observed during the procedure and normal bowel thickness was described.

Surgical intervention was done under the effect of general anesthesia. Presence of 2 pathologies has required a two-step approach to management intra-operatively; the first step is to manage the intussusception through manually reducing the affected part and the second step is to manage the malrotated part of the gut through a Ladd's procedure.

Each one of the four surgeries has been performed in the presence of an experienced pediatric surgeon assisted by a surgical trainee. Establishing a learning curve for both procedures at the same time is somehow difficult considering we are talking about 4 patients only.

The procedure was performed by a pediatric surgery consultant/

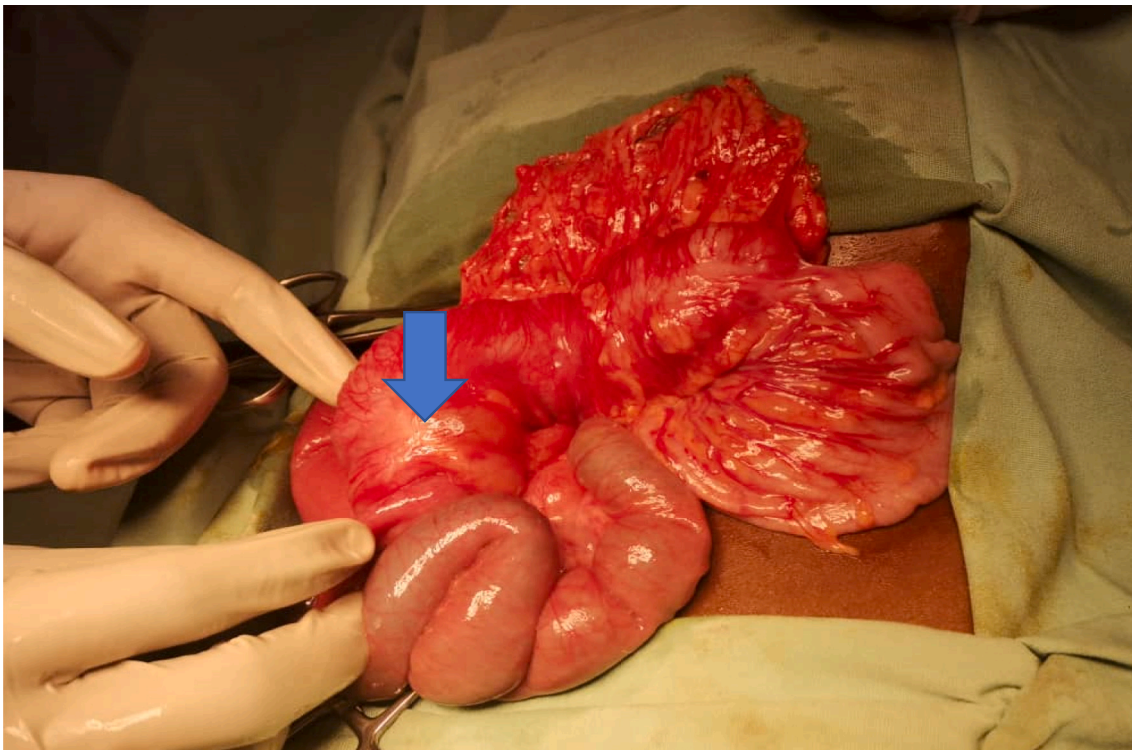


Fig. 1. Telescoping of one bowel segment into another featuring an intussusception.

specialist assisted by a pediatric surgery trainee. No specific or special post-surgical considerations were needed for patients and they lead a successful post-operative recovery course.

## 7. Follow-up and outcome

During operation the intussusception was manually reduced and the malrotation was corrected using Ladd's procedure relieving the obstruction. As a result, all of patients' symptoms of abdominal pain and vomiting have resolved. Patients returned back to the ward in good conditions and managed a successful post-operative course. Patients stayed for up to 10 days maximum based on their initial presentation and their hospital course.

The outcome for surgery to correct both conditions is very good. All four patients have been followed-up in the referred clinic for possible complications. There was no mortality among patients and there was no need for re-exploration or revision surgery and there was no wound-related complications of wound infection or dehiscence. They have been followed-up for a period of 6 months in the following intervals post-operatively (2 weeks, 1 month, 3 months and 6 months) with no complications either related to the wound or to the need for revision surgery.

None of our four patients have faced any problem with the procedure in terms of adhering to the recommended treatment plan or tolerating it and they lead a successful hospital course.

## 8. Discussion

Waugh's syndrome is a rare syndrome in which patients present with combined disease pathologies of acute-onset intussusception and gut malrotation. Its incidence has not truly established, but less than 100 cases worldwide have been reported so far as of 2017 [6]. Chronic

intussusception constitutes about 3% of all intussusception cases in children less than 1 year and around 10% is older children [7]. Association of chronic intussusception with intestinal malrotation, although could be a variable of Waugh's syndrome has only been described once in the literature [8].

With regard to the etiology, there is some currently available evidence which postulates that malrotation of the gut may increase the risk of concurrent acute intussusception as Waugh and Lond suggested. Their suggestion has posited that both the ascending and descending colon are relatively free and lacks adequate posterior wall attachment which may increase the risk of ileocecal intussusception. Failure of primitive mesenteric folds to be have the enough strength to close the mesenteric vessels may result in the typical presentation of recurrent symptoms without complications from the pathology [8].

What caught our attention in these set of cases is that all 4 cases presented of Waugh's syndrome in a 6-month period only. While this might be considered as unusual for a rare syndrome, there has been a report of 6 cases of such a syndrome in a period of 4 months by Breckon and Hadley at King Edward Hospital in Durban – 1998. There was a male preponderance of WS, as numbers suggested (4 out of 6 at King Edward Hospital and 3 out of 4 at our teaching hospital at Khartoum North. Breckon and Hadley's patients were reported to have an age range of 3–9 months, in cases we report here there is a diversity in their ages of presentation. Same preoperative optimization including intravenous fluids and fresh frozen plasma along with electrolytes correction were provided for all patients, here and in the previous cases.

When it comes to management of such presentation, open surgery for the purpose of manually reducing the intussusception followed by a Ladd's procedure for intestinal malrotation is the usual and most commonly used treatment method. In Breckon's report of cases, these steps were performed in one surgical approach while one patient underwent a redo-laparotomy [9]. In our 4 reported cases, we approached

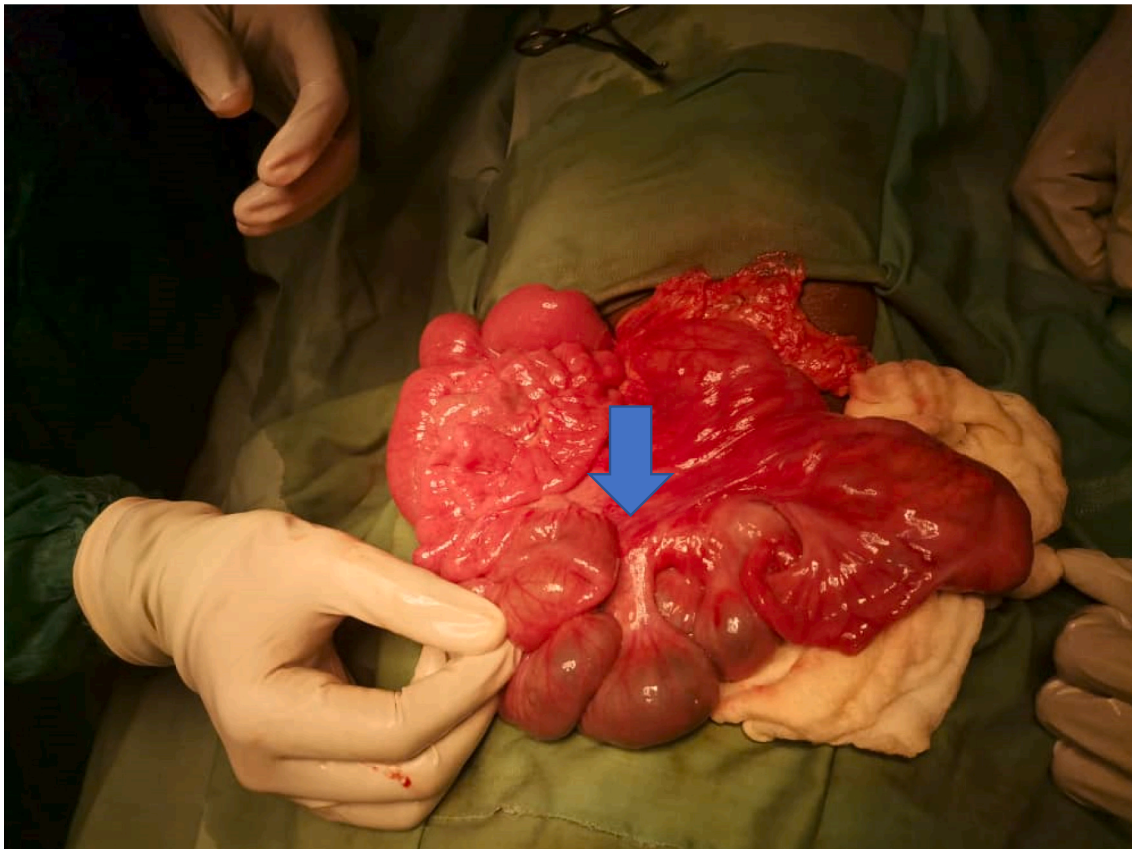


Fig. 2. Rotation of the bowel anti-clockwise around its mesentery featuring gut malrotation.

**Table 1**

Featuring the clinical findings for each patient.

	Age at presentation	Abdominal pain	Diarrhea	Bilious vomiting	Fever	Non-surgical reduction
First patient	12 years	Present	Not present	Present	Not present	Not attempted
Second patient	3 years	Present	Present (Bloody)	Present	Present	Not attempted
Third patient	8 months	Present	Present (Bloody)	Present	Not present	Not attempted
Fourth patient	3 years	Present	Present (Watery)	Not present	Not present	Not attempted

all of them through a one-time laparotomy procedure including detwisting of the segment underwent intussusception that was then followed by a Ladd's procedure. Laparoscopic approach to the same procedure has also been performed once [10]. The possibility of combined pathologies has not been considered in many cases and it was identified intraoperatively. Non-surgical approach with enema can be detrimental as it may conceal the associated malrotation resulting in the condition being recurrent and chronic [5]. One fact that should be stressed here is that any child with bilious vomiting and subtle abdominal sign has to carry out upper GIT contrast as this may help identifying or at least give us a clue as what could be the cause(s) before surgery. Additionally, in light of the above presentation and looking for potential causes or associated disorders, although not performed, we recommend upper GIT contrast in patients with recurrent intussusception as it could be of a great value in terms of the etiology.

The learning objective from this case is that the presence of cluster presentation for Waugh syndrome could suggest that some environmental factors might be implicated and further observations can be carried out to look more into such phenomenon. Additionally, this might bring into attention the vital role that pediatric surgeon can play in diagnosing this phenomenon to prevent delayed presentations and lowering morbidity and mortality. Moreover, in a low-resource setting and especially when patients are old enough, the possibility of a secondary cause should be considered and surgery can be considered the best option here.

This case has been reported in line with the SCARE criteria [11].

### Consent

Written informed consent was obtained from the patient for publication of this case series and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Ethical approval

Ethical approval was obtained from Ethical committee at Khartoum North Teaching Hospital.

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### Guarantor

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### CRediT authorship contribution statement

1. Ibrahim S Elkeir: Involved in study design, data acquisition, drafting the article, revising it critically and finally approved the manuscript.
2. Walaa Balla: Involved in conception of the study design, drafting the article and finally approved the manuscript.
3. Helenja Gurr: Involved in conception of the study design, drafting the article and finally approved the manuscript.
4. Moh Fatih: Involved in the design of the study, revising it critically and finally approved the manuscript.
5. Suliman Gabir Abdalla Mohammed: Involved in the design of the study, revising it critically and finally approved the manuscript.
6. Mohamed Abdulkarim: Involved in study design, data acquisition, drafting the article, revising it critically and finally approved the manuscript.

### Declaration of competing interest

Authors report no conflict of interest of any sort.

### References

- [1] M. Waseem, H.K. Rosenberg, Intussusception, *Pediatr. Emerg. Care* 24 (11) (2008 Nov) 793–800, <https://doi.org/10.1097/PEC.0b013e31818c2a3e>. PMID: 19018227.
- [2] E.G. Ford, M.O. Senac Jr., M.S. Srikanth, J.J. Weitzman, Malrotation of the intestine in children, *Ann. Surg.* 215 (2) (1992) 172–178, <https://doi.org/10.1097/00006558-199202000-00013>.
- [3] A. Cusimano, A.M.A.B. Abdelghany, A. Donadini, Chronic intermittent abdominal pain in young woman with intestinal malrotation, fitz-Hugh-Curtis syndrome and appendiceal neuroendocrine tumor: a rare case report and literature review, *BMC Womens Health* 16 (2015) 3, <https://doi.org/10.1186/s12905-015-0274-2>.
- [4] G. Baltazar, C. Sahyoun, J. Sime, M. Bitar, J. Bitar, A.C. Rao, Discovery of a case of Waugh's syndrome during a mission to Haiti, *Int. J. Surg. Case Rep.* 3 (1) (2012) 22–24, <https://doi.org/10.1016/j.ijscr.2011.09.006>. Epub 2011 Oct 25. PMID: 22288034; PMCID: PMC3267255.
- [5] H. Al-Momani, Waugh syndrome: a report of 7 patients and review of the published reports, *Ann. Saudi Med.* 34 (6) (2014) 527–531, <https://doi.org/10.5144/0256-4947.2014.527>.
- [6] Y.A. Khan, S.K. Yadav, A. Elkholy, Waugh's syndrome: report of two children with intussusception, *Eur. J. Pediatr. Surg. Rep.* 5 (1) (2017) e29–e31, <https://doi.org/10.1055/s-0037-1604264>.
- [7] D. Macauley, T. Moore, Subacute and chronic intussusception in infants and children, *Arch. Dis. Child.* 30 (1955) 180–183.
- [8] N. Zavras, K. Tsilikas, G. Vaos, Chronic intussusception associated with malrotation in a child: a variation of Waugh's syndrome? *Case Rep. Surg.* 2016 (2016), 5638451 <https://doi.org/10.1155/2016/5638451>.
- [9] V.M. Breckon, G.P. Hadley, Waugh's syndrome: a report of six patients, *Pediatr. Surg. Int.* 16 (5–6) (2000) 370–373, <https://doi.org/10.1007/s003830000349>. PMID: 10955564.
- [10] G. Harper, D. Hardy, C. Howell, R. Hatley, W. Pipkin, Laparoscopic approach to Waugh's syndrome, *Am. Surg.* 77 (4) (2011 Apr) E78–E79. PMID: 21679542.
- [11] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CASE REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.