Waugh syndrome is the association of intussusception and intestinal malrotation. The association is rarely reported in the literature though intussusception is a commonly encountered problem in pediatric patients as a cause of intestinal obstruction. We present our experience in 7 patients with a review of published reports.

**BACKGROUND AND OBJECTIVES:** Waugh syndrome (WS) is the association of intussusception and intestinal malrotation. The association is rarely reported in the literature though intussusception is a commonly encountered problem in pediatric patients as a cause of intestinal obstruction. We present our experience in 7 patients with a review of published reports.

**DESIGN AND SETTING:** Retrospective analysis of 7 patients with the diagnosis of Waugh syndrome who were treated at our department between February 1982 to December 2012.

**PATIENTS AND METHODS:** Seven patients with Waugh syndrome presented to our unit during the period February 1982 to December 2012. The clinical findings and management are presented and discussed.

**RESULTS:** Seven patients (three males and four females) presented with intussusception in association with malrotation. The age range was from 4 to 11 months; the patients had bilious vomiting and blood in the stool; the diagnosis was confirmed by ultrasound (2), Ba enema (2) and intraoperatively (3). All required operative intervention; either manual reduction or bowel resection and Ladd procedure; one patient died of sepsis; recurrence of obstruction was seen in another patient while the rest did well postoperatively.

**CONCLUSION:** The relationship between intestinal malrotation and intussusceptions may be more frequent than is reported; failure of non-operative management of intussusception may be due to this association and hence brings the attention to its existence. A prospective study is needed to look for intestinal malrotation in patient with intussusceptions who undergo abdominal sonographic examination to determine the true incidence of this association. The anomaly is suspected by presence of a reversed anatomic relationship of the superior mesenteric artery and vein and in such cases to perform an upper gastrointestinal contrast study to define the exact location of the duodenojejunal (DJ).
in 2 patients, but it failed and suggested the diagnosis of malrotation in 1 of these patients. Ultrasound examination confirmed the diagnosis of malrotation in 2 patients. The rest of the patients underwent exploratory laparotomy because of the presence of signs suggestive of peritonitis, as abdominal wall guarding and tenderness were observed right from the beginning. Ultimately all of the patients had undergone exploratory laparotomy.

RESULTS
Seven patients of Waugh were found among 106 intussusception patients who were admitted to our unit. Five patients were found to have an ileocolic intussusception, 1 patient had ileocoloanal intussusception, and 1 patient had ileocolorectal intussusception. One of the patients had intussusceptions with perforated large bowel that required the repair of the perforations and ileostomy formation. The small bowel was ischemic and dusky in color but not frankly gangrenous, which was left unresected and the patient was scheduled for possible second-look surgery. Fortunately, he did well and continued to improve over the next days; therefore, the second-look surgery was not done.

All of the patients were found to have malrotation as evidenced by the intraoperative finding of the location of the duodenojejunal junction, which was located to the right of the spine.
The outcome was good except for 1 patient who died of sepsis and another patient who developed recurrence of obstruction 2 weeks later, but the family refused admission and he was lost to follow-up. This particular patient did not have a Ladd procedure for an unknown reason mentioned in his record. Table 2 summarizes the operative findings and the operative procedure performed.

## DISCUSSION

The association of intussusception and malrotation has been rarely described in the published reports. Brereton...
et al. in a prospective study reported 15 patients with intussusceptions and intestinal malrotation among 37 patients with intussusception in whom the position of the duodenojejunal flexure was determined (40%); they called this association Waugh syndrome after George E Waugh, who was the first to describe the association in 1911 in a report of 3 boys with simultaneous intussusception and malrotation. A search of the published reports revealed a total of 63 cases up till now (Table 3).

In 2000, Breckon and Hadley described 6 infants with Waugh syndrome among 12 intussusception patients. They suggested that malrotation by its nature is associated with a mobile right colon, which may be a prerequisite for intussusceptions. In 2004, Inan et al reported 2 patients with Waugh syndrome. Both patients were found to have an unfixed cecum and mobile colon and agreed with Breckon and Brereton opinions about the etiology of intussusception.

The age of our patients ranged from 4-11 months, which is comparable with that reported in the published reports (4-36 months). In 3 different reports, 3 cases with an age outside the usual range were reported, which included a 28-week preterm and an 8-year-old child as well as a 56-year-old adult patient.

Ladd procedure and manual reduction of the intussusception have been the treatments of choice in Waugh syndrome to deal with both complications (i.e., intussusception and malrotation); all of our patients had manual reduction and Ladd procedure except 1 who had manual reduction only for an unknown reason and later she developed recurrence of obstruction, but the parents refused admission and she was lost to follow-up. A laparoscopic approach to Waugh syndrome was performed successfully in a 3-year-old full-term, male patient.

The disparity between the high incidence of Waugh syndrome in Brereton study and the rarity of cases in the published reports suggest that many cases remained undiagnosed, possibly because most cases of intussusception are reduced nonsurgically by air or contrast enema and the radiological signs of malrotation are not clear in these situations.

The failure rate of nonsurgical reduction is high (40% – 100% in Ref. 5 and Ref. 3, respectively) in patients with Waugh syndrome as seen in 2 patients who had a trial of enema reduction in our cases as well as in others. This might be an indication of the presence of malrotation in patients with intussusceptions who fail the nonsurgical reduction.

In conclusion, the relationship between intestinal malrotation and intussusceptions may be more frequent than is reported. A prospective study is needed to look for intestinal malrotation in patients with intussusceptions who undergo abdominal sonographic examination to determine the true incidence of this relationship. The anomaly is suspected by the presence of a reversed anatomic relationship of the superior mesenteric artery and vein, and it is advised to perform an upper gastrointestinal contrast study in such cases to define the exact location of the duodenojejunal junction.
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