original article

Waugh syndrome: a report of 7 patients and review of the published reports

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Ann Saudi Med 2014; 34(6): 527-531

DOI: 10.5144/0256-4947.2014.527

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 February1982 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 duodenojejuonal (DJ).

Waugh syndrome is the association of intussusception and malrotation. The association has been named Waugh syndrome by Brereton et al after George E Waugh who was the first to describe this entity In the only prospective study of this association of which we are aware of Brereton et al found a high incidence of malrotation among patients with intussusception.[1] Since then a few case reports were published describing this rarely reported entity. Seven patients with this association presented to our hospital and formed the basis of this study.

PATIENTS AND METHODS

Seven patients (3 males and 4 females) presented with intussusception in association with malrotation to our unit during the period February1982-December 2012 were retrospectively reviewed. The age range was from 4-11 months. **Table 1** summarizes the clinical presentation of these patients.

The clinical presentation of the involved patients was as follows: all of the patients had bile-stained vomiting and passage of blood in the stool. A sausage shaped abdominal mass suggestive of an intussusception could be palpated In 4 patients; the intussusceptum was seen prolapsing through the anus in 1 of the patients; and in the other the intussusception was felt reaching the rectum. The duration of symptoms ranged from 24 hours to 2 weeks.

All the patients were initially resuscitated with intravenous fluids. Contrast enema reduction was tried

Table 1. Summary of clinical presentation.

Age (months)	Sex	Vomiting	Rectal bleeding	Distension	Mass	Duration (d)	Enema reduction trial
4	F	Yes	Yes	No	Yes	7	No
4	F	Yes	Yes	No	No	1	No
8	F	Yes	Yes	Yes	No	4	Yes, malrotation was suggested
8	M	Yes	Yes	Yes	Yes	1	Yes
9	F	Yes	Yes	No	Yes	1	No
11	M	Yes	Yes	No	Yes	2	No
11	M	Yes	Yes	Yes	No	14	No

Table 2. Summary of the operative findings and the operative procedures performed.

Age (months)	Sex	Operative findings	Operative procedure	Outcome
4	F	Ileocolic intussusception with malrotation and volvulus with gangrenous bowel	Resection of gangrenous bowel and ileostomy and Ladd procedure	Died of sepsis
4	F	Ileocolic intussusception and malrotation	Operative manual reduction and Ladd procedure	Improved
8	F	Ileocolic intussusception and malrotation	Operative manual reduction and Ladd procedure	Improved
8	M	Ileocolic intussusception and malrotation	Operative manual reduction and Ladd procedure	Improved
9	F	Ileocolic intussusception and malrotation	Operative manual reduction only	Recurrence two weeks later, but the family refused admission and was lost to follow-up
11	M	ileocoloanal intussusception and malrotation	Operative manual reduction and Ladd procedure	Improved
11	M	Ileocorectal intussusception and malrotation	Resection of gangrenous bowel and repair of colon perforation, and ileostomy and Ladd procedure	Improved

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 duodenojejuonal junction, which was located to the right of the spine.

Table 3. Patients with Waugh syndrome reported in the published reports.

	Author	Journal	Y	Number of cases	Enema reduction
1	Waugh[2]	The Lancet	1911	3	None
2	Perrin and Lindsay[9]	British Journal of Surgery	1921	2	None
3	van Meurs[10]	British Journal of Surgery	1946	1	None
4	Tabibi et al[11]	Journal of the American Osteopathic Association	1971	1	None
5	Berry and Ray[12]	Southern Medical Journal	1972	1	None
6	Stewart et al[13]	Surgery	1976	1	None
7	Filston and Kirks[14]	Pediatric Surgery International	1981	3	None
8	Ornstein and Lund[15]	British Journal of Surgery	1981	1	None
9	Welch et al[16]	Annals of the Royal College of Surgeons of England	1983	1	None
10	Burke and Fitzgerald[17]	Australian and New Zealand Journal of Surgery	1985	1	None
11	Brereton et al[1]	British Journal of Surgery	1986	15	None
12	Jain[18]	Archives of Surgery	1989	2	None
13	Ward and Brereton[19]	European Journal of Pediatric Surgery	1992	2	None
14	Sarin and Singh[20]	Indian Pediatrics	1995	1	None
15	Lobo et al[21]	Pediatric Radiology	1997	7	None
16	Breckon and Hadley[3]	Pediatric Surgery International	2000	6	Failed 6
17	Luo et al[22]	Pediatric Surgery International	2003	1	Failed 1
18	Inan et al[4]	Journal of Pediatric Surgery	2004	2	None
19	Dawrant et al[6]	Pediatric Surgery International	2005	1	None
20	Rao and Kumar[23]	Indian Journal of Pediatrics	2005	1	None
21	Rangel et al[24]	Medicina Universitaria	2007	1	None
22	Lukong et al[27]	South Africa Journal of Surgery	2007	1	None
23	Domínguez-Pérez et al[25]	Acta Pediátrica de México	2008	5	Successful, 3 2 failed
24	Al-Jahdali et al[5]	Journal of Pediatric Surgery	2009	1	None
25	Hardy et al[8]	The American Surgeon	2011	1	None
26	Gerard et al[26]	International Journal of Surgery Case Reports	2012	1	None
	Total			**63**	

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 duodenojejuonal 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.[1,2] 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.[3] 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.[4]

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.[5-7]

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.[8]

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.[3]

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.[3,5,22] 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 patient 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 duodenojejuonal junction.

REFERENCES

1. Brereton RJ, Taylor B, Hall CM. Intussusception and Malrotation in infants: Waugh's syndrome. Br J Surg 1986;73:55-7.
2. Waugh GE. Referred penile pain in intussusception with notes of three cases. Lancet I .1911;1492-4.
3. Breckon VM, Hadley GP. Waugh's syndrome: a report of six patients. Pediatr Surg Int 2000;16:370–3.
4. Inan M, Basaran UN, Ayvaz S, Pul M. Waugh's syndrome: report of two cases. J Pediatr Surg 2004;39:110–1.
5. Al-Jahdali A, Lees GM, Gay DP, et al. Colocolic intussusception in a preterm infant with intestinal Malrotation. J Pediatr Surg 2009;44: e17-e18.
6. Dawrant MJ, Lee JC, Ho CP, et al. Complex presentation of intussusception in childhood. Pediatr Surg Int 2005;21:730–2.
7. Hsieh TM, Sheen-Chen SM, Lin CC. Combination of adult intussusception and intestinal malrotation: an unusual presentation. Dig Dis Sci 2008;53:3037–9.
8. Hardy D, Howell C, Hatley R, et al. Laparoscopic approach to Waugh's Syndrome. Am Surg 2011;77:78–9.
9. Perrin WS, Lindsay EC. Intussusception: a monograph based on 400 cases. Br J Surg 1921-1922;9:46-71.
10. Van Meurs Dp. Ileocolic intussusception associated with non-rotation of the midgut, congenital heart disease, and congenital hydrocele. Br J Surg 1946;34:91-3.
11. Tabibi M, Thompson RB, Gipe JF, et al. Malrotation syndrome with secondary intussusception: report of a case. J Am Osteopath Assoc 1971;70:686–91.
12. Berry BE, Ray JE. Malrotation of the colon with prolapsed ileocolic intussusception in familial polyposis. South Med J 1972;65(9):1075-8.
13. Stewart DR, Colodny AL, Daggett WC. Malrotation of the bowel in infants and children: a 15 year review, Surgery 1976;79(6):716-20.
14. Filston HC, Kirks DR. Malrotation - the ubiquitous anomaly. J Pediatr Surg 1981;16(4 Suppl 1):614-20.
15. Ornstein MH, Lund RJ. Simultaneous occurrence of malrotation volvulus and intussusception in an infant. Br J Surg 1981;68(6):440-1.
16. Welch GH, Azmy AF, Ziervogel MA. The surgery of malrotation and midgut volvulus: a nine year experience in neonates. Ann R Coll Surg Engl 1983;65(4):244-7.
17. Burke TE, Fitzgerald RJ. Intussusception, volvulus and malrotation. Aust N Z J Surg 1985;55(1):73-4.
18. Jain SK. Intestinal malrotation and midgut volvulus with intussusception: an unusual association. Arch Surg 1989;124(4):509.
19. Ward HC, Brereton RJ. Intussusception and duodenal stenosis: an infrequent but expected association. Eur J Pediatr Surg 1992;2(4):239-40.
20. Sarin YK, Singh VP. Waugh's syndrome. Indian Pediatr 1995;32:108–9.
21. Lobo E, Daneman A, Fields JM, et al. The diagnosis of malrotation during air enema procedure. Pediatr Radiol 1997 Jul; 27(7):606-8.
22. Luo CC, Wang CR, Chiu CH. Intussusception and intestinal malrotation in an infant: a case report. Pediatr Surg Int 2003;19: 413–4.
23. Rao PLNG, Kumar V. Waugh's syndrome, Indian J Pediatr 2005;72:E21–2.
24. Joel Cazares Rangel, Inti Bocanegra Cedillo,Hector, N. Morales Sauceda, et al. Sindrome de Waugh. Reporte de un caso Medicina Universitaria 2007;9(36):141-3.
25. Domínguez-Pérez ST, Baeza-Herrera C, Jaimes G et al. Síndrome de Waugh. Primeros informes en México Acta Pediatr Mex 2008;29(6):355-8.
26. Baltazar G, Sahyoun C, Sime J et al. Discovery of a case of Waugh's syndrome during a mission to Haiti, Int J Surg Case Rep 2012;3:22–24.
27. Lukong CS, Maitama MI, Aminu B, Mshelbwala PM, Ameh EA. Intestinal malrotation and midgut volvulus coexisting with intussusception. S Afr J Surg 2007;45:30.