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Hirschsprung's Disease Complicated by Sigmoid Volvulus: A Systematic Review

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Background: Hirschsprung's disease and sigmoid volvulus can sometimes be seen in the same patient.

Aims: To investigate the presence of Hirschsprung's disease in patients with sigmoid volvulus and to discuss the diagnosis and treatment methods.

Study Design: Systematic review.

Methods: This systematic review has been reported in line with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses and the methodological quality of systematic reviews guidelines. The PubMed and Scopus databases were scanned using the keywords "Hirschsprung* volvulus*" and "congenital aganglionic megacolon volvulus*". The reference list of the selected studies was reviewed for cross-checking. Two reviewers independently screened the available literature. Only the Hirschsprung's disease cases involving sigmoid volvulus were included, and cases of patients with volvulus in other sites was excluded. There was no restriction with respect to the publication language and type of writing. The primary outcome was morbidity and mortality.

Results: A total of 31 cases were analyzed in 22 articles; 97% of the patients were under the age of 40, 90% were male. There was a statistically significant difference in the necessity for relaparotomy between patients who were scheduled for sigmoid volvulus therapy with the suspicion of Hirschsprung's disease and patients who were treated without suspicion of Hirschsprung's disease (0% vs 37.5%, p=0.02). While there was no postoperative death in cases with suspected Hirschsprung disease, this mortality rate was 25% in cases without suspicion (p = 0.08).

Conclusion: Hirschsprung's disease should be excluded with rectal biopsy if a patient with sigmoid volvulus is under 40 years of age and has complaints of constipation from childhood.

Hirschsprung's disease and sigmoid volvulus are bowel diseases that are accompanied by chronic constipation. Both are more common in men (1). Hirschsprung's disease is more prevalent in newborns and in childhood, but a less aggressive form of a short-segment Hirschsprung's disease can be diagnosed in adolescence or in adulthood (2). Sigmoid volvulus is seen in patients at older ages, and it is rarely encountered in children (3). Sometimes both pathologies may occur in the same individual. Hirschsprung's disease is a congenital disease, and sigmoid volvulus is a complication of Hirschsprung's disease. It is important to diagnose an underlying Hirschsprung's disease in sigmoid volvulus cases.

The basis of the treatment of Hirschsprung's disease is resection of the aganglionic segment and construction of coloanal anastomosis between the normal colon and anal canal. To achieve this, various treatment modalities have been developed (4). The management approach of sigmoid volvulus includes derotation of the sigmoid colon by endoscopic, enema, or surgical approaches in patients with viable bowel. After this step, elective surgery can be planned for patients with acceptable anesthesia and operative risk. In patients with a failed derotation attempt, findings of acute abdomen, or a gangrenous bowel, emergency surgery is indicated (5, 6). If the associated Hirschsprung's disease is overlooked, the outcomes of the treatment of sigmoid volvulus alone are not clear. This study aimed to provide clarity with a comprehensive systematic review of the literature.

MATERIALS AND METHODS

This systematic review was prepared on the basis of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses.

Search strategy

The PubMed and Scopus databases were scanned using the keywords "Hirschsprung* volvulus*" and "congenital aganglionic megacolon volvulus*". The final screening took place on May 13,

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2020. The reference list of the selected studies was reviewed for cross-checking.

Selection criteria and intervention(s)

There was no restriction with respect to the publication language and type of articles. Only the Hirschsprung's disease cases involving sigmoid volvulus were included, and other volvulus cases (cecal, transverse, or splenic) were excluded. An e-mail was sent to the authors to obtain the missing data (such as surgical outcome, gender, and age) in their articles.

Data extraction and study quality

The methodological quality of the systematic reviews was used to assess the quality of this review (7). The Cochrane Handbook for Systematic Reviews of Interventions (the Handbook) was used for evaluating the risk of bias of the included studies in this review (8). This systematic review was registered in an international database (International Prospective Register of Systematic Reviews, CRD42020166692). Most studies reporting concomitant Hirschsprung's disease and sigmoid volvulus mainly include isolated case reports or small number of case series because this entity is rare. The year of publication, country of publication, age, sex, the presence of gangrene, surgical or nonsurgical treatments, and outcomes were reviewed. The data were evaluated separately by 2 researchers (UU and CK). A total of 31 cases were included in the analysis in 22 studies that met the defined inclusion criteria (Figure 1). Patients were grouped according to the following age groups: 0 to 2 years, newborn and infant; 2 to 17 years, pediatric; and ≥18 years, adults.

Statistical analysis

Statistical analyses were performed using the Statistical Package for the Social Sciences Version 22.0 (IBM SPSS Corp, Armonk, NY, USA). Quantitative variables were expressed as mean ± standard deviation (SD), median, minimum-maximum, and interval. Qualitative variables were reported as numbers and percentages. The Shapiro-Wilk test was used to assess the normality distribution of the quantitative variables. Although the means and SDs were used for homogeneous distributions, median and range were used for heterogeneous distributions. The Fisher's Chi-square test was used to compare the qualitative variables. The Mann-Whitney *U* test was used for heterogeneous distributions, and the Student's *t* test was used for homogeneous distributions. $p<0.05$ was considered statistically significant.

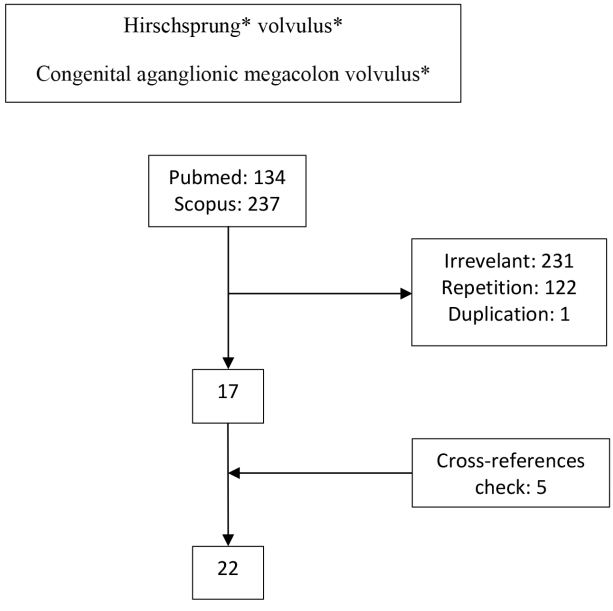

FIG. 1. Flowchart of the systematic review.

RESULTS

The selected publications included 16 cases from Asia, 8 cases from Europe, 4 cases from Africa, and 3 cases from North America (9-26). The age distribution of the patients was not homogeneous (Shapiro-Wilk test, $p<0.05$). The age range was 4 hours to 82 years (median=10 years). Median age for newborn/infant, child, and adult was 1 day (range, 1-42 days), 7 years (range, 3-12 years), and 24 years (range, 18-82 years), respectively; 97% of the patients were younger than 40 years and 90% were men (Table 1).

Nonoperative detorsion was attempted in 14 of 25 patients (56%) (7 with enema, 2 with rectal tube, and 5 with endoscopy), with a success rate of 64.3% (9/14). A total of 11 patients (failed nonoperative derotation in 5 patients; nonoperative intervention 6 patients was not attempted) underwent surgical detorsion.

In 19 (70.4%) patients, Hirschsprung's disease was diagnosed or suspected during the treatment of sigmoid volvulus (Table 2). In these 19 patients, the treatment of Hirschsprung's disease was successfully performed in a single procedure or stepwise surgical intervention. In the remaining 8 patients in whom the Hirschsprung's disease diagnosis was overlooked, 3 patients required early relaparotomy after the first operation, and 2 of these patients died. There was a difference in the necessity for relaparotomy (37.5% vs 0%, p=0.02) between patients who were scheduled for sigmoid volvulus therapy with the suspicion of Hirschsprung's disease and patients who were treated without suspicion of Hirschsprung's disease. Although mortality was not observed in 19 patients who were suspected to have Hirschsprung's disease, mortality was observed in 2 of the 8 patients who received only sigmoid volvulus treatment without considering Hirschsprung's disease (25% vs 0%, p=0.08). In 4 patients, there were no data on this issue.

Intraoperative bowel gangrene was observed in only 3 (9.7%) patients. In 1 patient, gangrene was observed in the entire colon. The surgical procedures performed for Hirschsprung's disease showed heterogeneity. The Soave procedure was performed in 6 patients, Swenson procedure in 11 patients, sphincterotomy in 1 patient, and restorative proctocolectomy in 1 patient.

Mortality was observed in a total of 2 (8%) patients, where 1 was of pediatric age and the other was an adult. Both the patients did not have gangrene. However, neither of the patients had undergone definitive surgery for Hirschsprung's disease because of unawareness of a Hirschsprung's disease diagnosis. One of these patients underwent only surgical detorsion, and the cause of death was unknown. Resection and anastomosis with transverse colostomy were performed in the other patient. After the closure of the colostomy, relaparotomy was necessary owing to massive abdominal

TABLE 1. Reports of 31 Hirschsprung's diseases complicated by sigmoid volvulus in chronological order

Author	Country	Year	Age	Gender	Perioperative findings
Dean (9)	United States of America	1952	4 years	Male	SV
Shepherd (10[a])	Uganda	1969	12 years	Male	SV
Ciardimi et al. (10[a])	Italy	1977	3 years	Male	SV
Ciardimi et al. (10[a])	Italy	1977	5 years	Male	SV
Buts et al. (11)	Belgium	1980	10 years	Female	SV
Valla et al. (10a)	France	1982	5 days	Male	SV
Valla et al. (10a)	France	1982	5 years	Male	SV
McCalla et al. (12)	United States of America	1985	2 days	Male	SV
Henales et al. (10[a])	Spain	1993	5 years	Male	SV
Erdener et al. (13)	Turkey	1995	3 years	Male	SV
Venugopal et al. (14)	United Kingdom	1997	4 hours	Male	SV
Sarioglu et al. (15)	Turkey	1997	11 years	Male	SV
Rachid (16)	Morocco	1997	Adult[b]	Male[b]	SV
Puneet et al. (17)	India	2000	12 years	Male	Gangrenous SV
Bach et al. (18)	Malawi	2003	Adult[c]	N/A	SV
Tan et al. (19)	Singapore	2006	38 years	Male	Gangrenous SV[d]
Ghaemi et al. (20)	Iran	2010	18 years	Female	SV
Ghaemi et al. (20)	Iran	2010	21 years	Male	SV
Ghaemi et al. (20)	Iran	2010	19 years	Male	SV
Ghaemi et al. (20)	Iran	2010	24 years	Male	SV
Alagumuthu et al. (21)	India	2011	33 years	Male	SV
Alagumuthu et al. (21)	India	2011	18 years	Female	SV
Alagumuthu et al. (21)	India	2011	24 years	Male	SV
Zeng et al. (10)	United States of America	2012	12 years	Male	SV
Ibrahim et al. (22)	Egypt	2013	7 years	Male	SV
Khalayleh et al. (23)	Israel	2016	1.25 days	Male	SV
Khalayleh et al. (23)	Israel	2016	1.25 days	Male	SV
Khalayleh et al. (23)	Israel	2016	1 days	Male	SV
Ranjan et al. (24)	India	2016	11 years	Male	SV
Van Horebeek et al. (25)	Belgium	2017	42 days	Malee	SV
Wu (26)	China	2018	82 years	Male	Gangrenous SV

N/A: not applicable, SV: sigmoid volvulus
aReference found in cited review article; bFrom Scopus; cAdulthood of the patient was understood from the picture in the article; dGangrenous sigmoid colon, duskiness involving the entire colon; eThe author of the study reported by e-mail.

distension. There was anastomotic leakage, and primary repair and a transverse colostomy were performed. Hirschsprung's disease was detected in the rectal biopsy, but the patient died from the septic shock of fecal peritonitis. No mortality was seen in patients undergoing a definitive surgery for Hirschsprung's disease.

DISCUSSION

The incidence of Hirschsprung's disease is approximately 1 in 5000 live births (27, 28), which may change among ethnic groups. The incidence of Hirschsprung's disease is 1.5 in 10,000 live births in Northern Europe and 2.1 in African American and 2.8 in Asian populations (29). In this analysis, most patients with sigmoid volvulus had combined Hirschsprung's disease, 50% of the cases (15/30) were reported from Asia, and the lowest rate was from the United States, with 10% of the cases (3/30). The incidence of Hirschsprung's disease also varies between sexes. The male-to-female ratio is approximately 4:1 (1, 30). This study found a similarly high incidence of the disease in men (90%).

Approximately 95% of patients with Hirschsprung's disease are diagnosed before the age of 5 years (31). Mild cases can be diagnosed after the age of 10 years and are called adult Hirschsprung's disease (32-34). The actual incidence of patients with adult Hirschsprung's

TABLE 2. Reports of surgical approach for 31 cases of Hirschsprung's disease complicated by sigmoid volvulus in a chronological order

Author	Derotation	Biopsy	Surgery	Doubt on Hirschsprung's disease	Outcome
Dean (9)	Enema/yes	N/A	Surgical details are not available	N/A	Recovered
Shepherd (10[a])	Surgery/yes	N/A	Only derotation	No	Died
Ciardimi et al. (10[a])	N/A	N/A	Surgical details are not available	N/A	N/A
Ciardimi et al. (10[a])	N/A	N/A	Surgical details are not available	N/A	N/A
Buts et al. (11)	Enema/failed	Yes	First sigmoidopexy, later Swenson	No	Recovered
Valla et al. (10[a])	Surgery/yes	N/A	Only sigmoidectomy	No	Recovered
Valla et al. (10[a])	Surgery/yes	N/A	Only sigmoidectomy	No	Recovered
McCalla et al. (12)	Surgery/yes	Yes	First only derotation, relaparotomy[b], later Soave	No	Recovered
Henales et al. (10[a])	Surgery/yes	N/A	Enterostomy and sphincterotomy	Yes	Recovered
Erdener et al. (13)	Surgery/yes	Yes	First colostomy later Soave	Yes	Recovered
Venugopal et al. (14)	Enema/yes	Yes	Soave	Yes	Recovered
Sarioglu et al. (15)	Surgical/yes	Yes	First colostomy later Swenson	Yes	Recovered
Rachid (16)	N/A	N/A	Surgical details are not available	N/A	N/A
Puneet et al. (17)	N/A	Yes	Hartmann's procedure	No	Recovered
Bach et al. (18)	N/A	Yes	First mesosigmoidoplasty, then relaparotomy[c]	No	Recovered
Tan et al. (19)	Surgical/yes	Yes	Restorative proctocolectomy	Yes	Recovered
Ghaemi et al. (20)	Endoscopy/yes	Yes	Swenson	Yes	N/A
Ghaemi et al. (20)	Endoscopy/failed	Yes	First derotation, later Swenson	Yes	N/A
Ghaemi et al. (20)	Endoscopy/failed	Yes	First derotation, later Swenson	Yes	N/A
Ghaemi et al. (20)	Endoscopy/failed	Yes	First derotation, later Swenson	Yes	N/A
Alagumuthu et al. (21)	Surgical/yes	Yes[d]	Anastomosis and colostomy[e] and relaparotomy	No	Died
Alagumuthu et al. (21)	Surgical/yes	Yes	First colostomy,[f] later Swenson	Yes	Recovered
Alagumuthu et al. (21)	Rectal tube/yes	Yes	Soave[g]	Yes	Recovered
Zeng et al. (10)	Rectal tube/yes	Yes	Surgical details are not available	Yes	Recovered
Ibrahim et al. (22)	Enema/failed	Yes	First colostomy, later Soave[g]	Yes	Recovered
Khalayleh et al. (23)	Enema/yes[h]	Yes	Swenson	Yes	Recovered
Khalayleh et al. (23)	Enema/yes	Yes	Swenson	Yes	Recovered
Khalayleh et al. (23)	Enema/yes	Yes	Swenson	Yes	Recovered
Ranjan et al. (24)	N/A	N/A	Soave[i]	Yes	Recovered
Van Horebeek et al. (25)	Endoscopy/yes	Yes	Swenson[j]	Yes	Recovered
Wu (26)	Surgical/yes	Yes	First Hartmann's procedure later none	Yes	Recovered

N/A: not applicable
[a]Reference found in a cited review article; [b]After 12 days emergency laparotomy and colostomy; [c]Relaparotomy but surgical technique not definition; [d]Second transverse colostomy; [e]Transverse colostomy done. After 1 week of colostomy closure; primer repair, transverse colostomy plus rectal biopsy owing to perforation; [f]Double barrel colostomy; [g]Modified Soave; [h]Derotation with enema owing to recurrence of 2 times; [i]Primary Scott Boley's; [j]The author was reached via mail.

disease is unknown because Hirschsprung's disease is often overlooked in adult patients. There are cases of Hirschsprung's disease even at the ages of 74 and 82 years (26, 35). Approximately half of the adult patients with Hirschsprung's disease are younger than 30 years (2). In our study, 93% of the patients were younger than 30 years and 97% were younger than 40 years.

Hirschsprung's disease is seen in 0.6% to 3% of sigmoid volvulus cases (15, 23). Hirschsprung's disease frequency increases to 17% in the sigmoid volvulus cases below the age of 18 years (3). In a study by Ghaemi et al. (20), Hirschsprung's disease was detected in approximately 1 of 3 patients with sigmoid volvulus whose ages ranged between 14 and 30 years. The aganglionic segment in Hirschsprung's disease starts from the distal rectum, and the extent of the proximal diseased part varies. In 80% of individuals, aganglionosis is restricted to the rectosigmoid colon (short-segment disease); in 15% to 20% of patients, aganglionosis extends proximally to the sigmoid colon (long-segment disease); and in approximately 5%, aganglionosis affects the entire large intestine (total

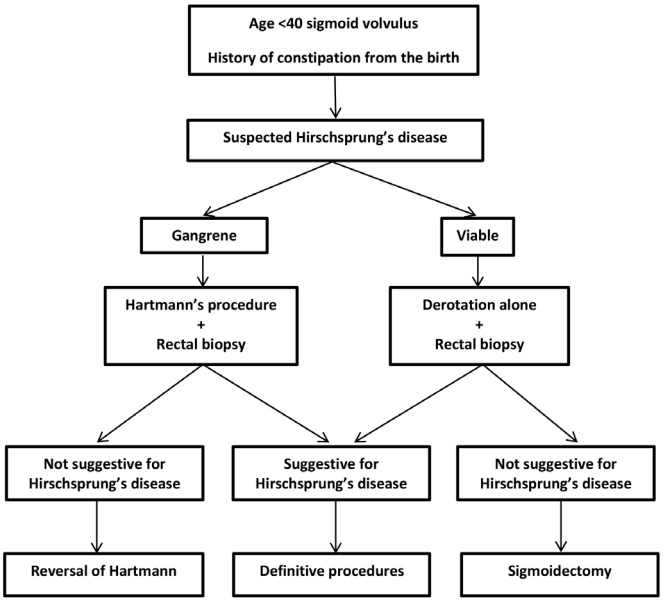

FIG. 2. Management of Hirschsprung's disease complicated by sigmoid volvulus.

colonic aganglionosis). Rarely, the aganglionosis extends into the small bowel or even more proximally to encompass the entire bowel (total intestinal aganglionosis). In most cases of late-diagnosed Hirschsprung's disease, the short segment of the bowel is affected. In these patients, prolonged constipation increases the likelihood of developing sigmoid volvulus (27).

Rectal biopsy for the diagnosis of Hirschsprung's disease is a worldwide accepted entity. Rectal biopsy is recommended from 1 to 3 cm above the dentate line (36). Although it is frequently used as a result of consensus, routine use of anorectal manometry or barium enema in Hirschsprung's disease is not necessary (37). Anorectal manometry has been reported to be more useful for neurogenic achalasia of the internal anal sphincter rather than for Hirschsprung's disease (37). It is difficult to detect the presence of Hirschsprung's disease using radiological evaluation techniques in a patient with sigmoid volvulus. In the same consensus, it was decided that barium enema is not necessary to confirm the diagnosis of Hirschsprung's disease, but it aids in the determination of the extent of involved aganglionic bowel segment to guide the operative strategy (37).

On rectal examination, a massive discharge of flatus and stool is expected in Hirschsprung's disease, but this may not be observed when sigmoid volvulus is present. Most patients (97%) in this study were younger than 40 years, which suggests that Hirschsprung's disease should be suspected in patients younger than 40 years with cases of sigmoid volvulus (Figure 2). Previously published guidelines from Salas et al. (3) on the management of sigmoid volvulus with suspected Hirschsprung's disease do not mention the aspects of age or colon viability. Therefore, we recommend that in patients who have signs of suspected Hirschsprung's disease (complaints of constipation since childhood and patients younger than 40 years) at the time of sigmoid volvulus diagnosis, rectal biopsy should be performed. This is more likely to be recommended if the patient is male.

During the evaluation of patients with sigmoid volvulus together with megacolon, physicians should consider not only Hirschsprung's disease but also degenerative myopathies, such as Bantu's pseudo-Hirschsprung's disease or isolated hypoganglionosis. Degenerative myopathies are inherited diseases, such as Hirschsprung's disease, and there are also adult forms of the disease that can be seen in patients in the age range of 30 to 34 years. In these patients, diagnoses can also be made by rectal biopsy (38-41).

In patients with sigmoid volvulus with suspected Hirschsprung's disease, rectal biopsy is needed before sigmoid resection. Definitive surgery should be planned according to the result of the rectal biopsy. In the first stage, detorsion should be performed by endoscopic, enema, or surgical means. When Hirschsprung's disease is excluded by an intraoperative frozen biopsy, the surgeon can continue definitive surgery. In this study, only 4 of the 21 patients (19.0%) were diagnosed through an intraoperative frozen biopsy. A colostomy was performed, and definitive surgery was performed in the second session in 3 patients. In the remaining 1 patient, owing to the presence of gangrene in the entire colon, total colectomy with J pouch ileoanal anastomosis was performed in the initial operation.

In this study, the incidence of sigmoid gangrene was lower in patients with sigmoid volvulus with Hirschsprung's disease (9.7%). We think that the presence of gangrene is not an obstacle to rectal biopsy. In these patients, resection is essential and this colon resection can make definitive surgery more difficult. Moreover, 3 cases of patients with gangrene were reported in the literature, and only 1 of these patients underwent rectal biopsy. The Hartmann's procedure was performed in a 12-year-old male patient with gangrenous sigmoid volvulus (17). A 38-year-old male patient with gangrenous sigmoid volvulus was diagnosed as having a gangrenous sigmoid colon extending to the transverse and ascending colon. This patient underwent proctocolectomy with ileal pouch-anal anastomosis (19). An 82-year-old man was initially followed up conservatively. Laparotomy was performed owing to evolving acute abdominal findings. The Hartmann's procedure was performed because a gangrenous sigmoid colon was observed during the exploration (26).

When Hirschsprung's disease is suspected in patients with sigmoid volvulus, colostomy as the first-step surgical procedure is more appropriate if a sigmoid resection is necessary. If the patient has overlooked Hirschsprung's disease, the risk of anastomotic leakage is high owing to the distal aganglionic segment. If there is suspicion of Hirschsprung's disease in a patient and anastomosis was performed, rectal tube placement may be recommended.

The limitations of this review are that most studies included in this review were case reports and the operations performed for Hirschsprung's disease were heterogeneous. Sigmoid volvulus association with Hirschsprung's disease is a rare event. Therefore,

this review is not appropriate for meta-analysis, or inclusion of randomized controlled trials is needed to make it appropriate. Although there is a low risk of bias for the included studies, this review suggests a low-quality recommendation, but we think that the recommendations are easily applicable and will assist in avoiding mortality and morbidity.

In patients with constipation since childhood, especially in those with sigmoid volvulus who are younger than 40 years, the likelihood of an underlying Hirschsprung's disease should be considered, and this diagnosis should be excluded before resection and anastomosis of the colon.

Ethics Committee Approval: Since it was a systematic review, its approval to the ethics committee was not required. However, this systematic review was registered in an international database (PROSPERO, CRD42020166692).

Patient Consent for Publication: N/A.

Author Contributions: Concept - U.U.; Design - U.U., C.K.; Supervision - C.K.; Resources - U.U., Ö.G.; Materials - U.U., Ö.G.; Data Collection and/or Processing - U.U., C.K.; Analysis and/or Interpretation - U.U., C.K.; Literature Search - U.U., C.K.; Writing Manuscript - U.U., C.K.; Critical Review - C.K.

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