The Turkish Journal of Pediatrics 1997; 39: 91-98

CLINICAL RISK FACTORS OF HIRSCHSPRUNG – ASSOCIATED ENTEROCOLITIS II: POSTOPERATIVE ENTEROCOLITIS*

*Akıle Sarıoğlu MD***, F. Cahit Tanyel MD***, Nebil Büyükpamukçu MD***
*Akgün Hiçsönmez MD****

SUMMARY: Sarıoğlu A, Tanyel FC, Büyükpamukçu N, Hiçsönmez A. (Department of Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Clinical risk factors of Hirschsprung-associated enterocolitis. II: Postoperative enterocolitis. Turk J Pediatr 1997; 39: 91-98.

Among 302 Hirschsprung patients diagnosed between 1976 and 1993, 213 patients who had undergone definitive operations for Hirschsprung's disease (HD) were reviewed for the occurrence of postoperative Hirschsprung-associated enterocolitis (HAEC). The role of 42 parameters were analyzed. We detected 34 patients (14.0%) with postoperative HAEC with no mortality. In nine patients (26.5%) HAEC persisted postoperatively but no effect of preoperative HAEC on postoperative HAEC was detected. The length of the aganglionic segment did not have any effect on HAEC. Down's syndrome was not present in any of our patients with HAEC. Enterocolitis appeared after Swenson, Duhamel and Boley operations in 23, 10 and one patient, respectively. The incidence of HAEC was inversely proportional to age and weight at the time of definitive operation. The presence of colostomy before or during definitive operation had no effect on postoperative HAEC. Complications of definitive operations, including stricture, did not have any effect on HAEC. *Key words: Hirschsprung's disease, postoperative enterocolitis.*

The surgical principles of Hirschsprung's disease (HD) were put forward by Swenson and Bill in 1948[1]. Later, Duhamel[2], Soave[3], State[4], Martin[5], Boley[6] and others proposed different types of pull-through operations and modifications of these[7]. These operations carry the risk of complications related to both the type of the procedure and the nature of the disease itself[8]. Among these, Hirschsprung-associated enterocolitis (HAEC) is accepted to have the highest incidence and mortality rate[9-11]. However, it is still a matter of debate whether postoperative HAEC is a result of the natural course of the disease, a complication of definitive operations, or both. Some authors have stated that the spasm of the aganglionic sphincter might be the cause of HAEC[9, 12, 13]. Anastomotic stricture and insufficient operation are the other causes that have been attributed to the operation[14]. Although some surgeons still prefer to perform the colostomy procedure before definitive operation, nowadays there is a tendency towards operating on Hirschsprung patients in the newborn period and avoiding a colostomy[15-17]. The incidence of HAEC reported after this sort of operation is very low. A retrospective

* From the Department of Pediatric Surgery, Hacettepe University Faculty of Medicine, Ankara.

** Pediatric Surgeon, Hacettepe University Faculty of Medicine.

*** Professor of Pediatric Surgery, Hacettepe University Faculty of Medicine.

study was planned to evaluate the clinical risk factors of postoperative HAEC, its natural course and whether any relation exists between preoperative and postoperative HAEC.

Material and Methods

Among 302 Hirschsprung patients admitted to Hacettepe University Children's Hospital between 1976 and 1993, 213 patients who had undergone definitive operations for HD in the same period of time were analyzed for the presence of postoperative Hirschsprung associated enterocolitis (HAEC). The patients were compared with others who did not have postoperative HAEC in order to determine clinical risk factors related postoperative HAEC. The diagnosis of HAEC was based on foul-smelling, explosive diarrhea.

The patients were evaluated according the following data: a) history and symptoms on admission: 1. sex, 2. age of diagnosis, 3. birthweight, 4. gestational age, 5. family history, 6. history of death of a sibling, 7. consanguinity, 8. associated anomaly, 9. delay in the passage of meconium, 10. abdominal distention on physical examination, 11. defecation pattern, 12. intestinal obstruction, and 13. intestinal perforation; b) the relation to preoperative HAEC: 1. the presence of preoperative HAEC, and 2. the number of attacks of preoperative HAEC; c) length of aganglionic segment and factors related with definitive operation: 1. length of aganglionic segment, 2. the presence of colostomy before definitive operation, 3. diarrhea after colostomy, 4. type of definitive operation, 5. period between definitive operation and colostomy closure, 6. age at the time of definitive operation, 7. weight at the time of definitive operation, 8. colostomy closure during definitive operation, 9. type of suture used in definitive operation, 10. leakage of anastomosis, 11. mucosal disruption of anastomosis, 12. adhesive intestinal obstruction, 13. postoperative sepsis, 14. postoperative pelvic abscess, 15. stricture of anastomosis, 16. period between definitive operation and colostomy closure, and 17. date of definitive operation; d) clinical course of postoperative HAEC: 1. time of first attack after colostomy, 2. paracolic abscess, 3. perforation, 4. sepsis, 5. number of attacks, 6. number of hospitalizations, 7. period of hospitalization, 8. microorganism, 9. type of antibiotic, 10. period of antibiotic usage.

Short-segment aganglionosis (SS) was accepted as aganglionosis extending from the rectum up to the sigmoid colon, long-segment disease (LS) above the sigmoid up to the ascending colon, and extensive aganglionosis (EA) as total colonic aganglionosis and beyond. For statistical comparisons, chi-square and Fisher's exact chi-square tests were used, and p values less than 0.05 were considered to be significant.

Results

Among 302 Hirschsprung patients diagnosed between 1976 and 1993 at Hacettepe University Children's Hospital, 213 patients underwent definitive operations for HD. In 34 (16.0%) of these patients, postoperative HAEC without mortality was detected. Reliable data about the details of postoperative HAEC could not be obtained from all the files so these data were excluded.

Five (14.7%) female and 29 (85.3%) male patients comprised the postoperative HAEC group ($p > 0.05$). The age of diagnosis was similar in both groups as well ($p > 0.05$). In patients with postoperative HAEC, the age at diagnosis was the neonatal period in 16 cases (47.1%) one to 12 months in eight cases (23.5%) one to six years in nine cases (26.5%), and greater than seven years in one case (2.9%).

The majority of patients in both groups had birth weights of 2500 to 3500 g (60.0% in the HAEC group and 66.7% in the group without HAEC). Similarly, the majority were term babies (95.8% and 93.9% in the groups with and without HAEC, respectively). No statistical test was applied for these two criteria.

HD was detected in only one of the brothers of the postoperative HAEC patients who died after definitive operation for HD outside our hospital. Four other siblings of patients with postoperative HAEC had a history of a disease resembling HD but no diagnostic verification. Consanguinity was detected in only one patient (2.9%) with postoperative HAEC. Consanguinity was present in 16.1 percent of the group without HAEC, and the difference was statistically significant ($p < 0.05$). In seven (20.6%) patients with HAEC and 20 patients (17.7%) without HAEC, an associated congenital anomaly was detected ($p > 0.05$). None of the patients with HAEC had Down's syndrome.

Three patients (11.1%) with postoperative HAEC reportedly passed meconium in the first 24 hours of life, six (22.2%) of them between 24 and 48 hours, and 18 (66.7%) after 48 hours ($p > 0.05$).

Abdominal distention on first admission was present in 94.1% of HAEC patients and 93.8% of patients without HAEC ($p > 0.05$). Defecation patterns before definitive operation were similar in the two groups. Intestinal obstruction on first admission was detected in 50.0 percent of patients with HAEC and 38.1 percent of patients without HAEC.

Among postoperative HAEC patients, only nine (26.5%) had HAEC preoperatively. There was no statistical difference between the groups with and without HAEC in terms of having preoperative HAEC. Six (66.7%) of the nine patients had more than four attacks of preoperative HAEC and 18 (52.9%) patients without postoperative HAEC also had more than four attacks of preoperative HAEC ($p > 0.05$).

The length of the aganglionic segment was not an important factor in the development of postoperative HAEC. Twenty-five (73.5%) patients with HAEC had SS disease, as did 97 (85.8%) of those without HAEC ($p > 0.05$).

In all the patients with postoperative HAEC the definitive operation was preceded by a colostomy procedure.Among patients without postoperative HAEC, 105 (92.9%) had a colostomy before definitive operation ($p > 0.05$). Diarrhea after colostomy was present in 11.8 and 8.7 percent of the patients with HAEC and without HAEC, respectively ($p > 0.05$).

Postoperative HAEC appeared after Swenson operation in 23 patients (67.7%), after Duhamel operation in 10 patients (29.4%), and after Boley operation in one patient (2.9%). Only Swenson and Duhamel operations were taken into consideration for statistical comparison ($p > 0.05$).

The period between colostomy opening and definitive operation was less than six months in nine (26.5%) patients, six to 12 months in 10 (29.4%) patients, 12 to 24 months in seven (20.6%) patients, and more than 24 months in eight (23.5%) patients ($p > 0.05$).

The ages of patients at the time of definitive operation varied significantly ($p < 0.05$). In the HAEC group eight patients (23.5%) were operated on before the age of 15 months, 21 patients (61.8%) at 15 months to three years, and five patients (14.7%) at three years or greater. These percentages were 8.0, 62.0 and 30.1 percent respectively, for the group without HAEC. The weights of patients varied as well. Eight patients (24.2%) were less than 9 kg, 24 (72.7%) were 10-20 kg and one (3.0%) was greater than 21 kg in the HAEC group; these percentages were 10.7 75.0 and 14.3 percent, respectively, in the group without HAEC.

At the end of definitive operation no colostomy was present in seven patients with HAEC and 27 patients without HAEC ($p > 0.05$). Silk was used in 72.0 percent of patients with HAEC for the anastomosis of the pull-through operation, and 52.5 percent of those without it. Silk and stapler was used in 16.0 percent of the HAEC group and 17.5 percent of the group without HAEC. Vicryl and stapler was used in 6.3 percent of patients with HAEC but none without it. Other suture material and a crushing clamp were also used in some patients.

Leakage of the anastomosis was detected in one patient with postoperative HAEC and two patients without it ($p > 0.05$). Mucosal disruption of the anastomosis was present in two patients with HAEC and two patients without it ($p > 0.05$), and adhesive intestinal obstruction after definitive operation in five and seven patients, respectively ($p > 0.05$). Sepsis after definitive operation was found in one of the patients with HAEC and one without it ($p > 0.05$). Of six patients who had pelvic abscess after definitive operation, two developed HAEC ($p > 0.05$). Anastomotic stricture was present in 25 patients, eight of whom developed HAEC ($p > 0.05$).

The period between definitive operation and colostomy closure was three to six weeks in seven patients (25.9%), seven to 12 weeks in five (18.5%) and more than 13 weeks in 15 (55.6%) patients ($p > 0.05$). The patients' definitive operations were performed between 1976 and 1980, 10 between 1981 and 1985, eight between 1986 and 1990, and five between 1991 and 1993 ($p > 0.05$).

Neither pericolic abscess nor perforation developed in postoperative HAEC. The first attack of HAEC started one month after the colostomy closure in 13 patients (38.2%), two months after in six (17.7%), three months after in eight (23.5%), and after one year in seven (20.6%). Twenty-two patients (64.7%) had one to three attacks, seven (20.6%) had four to six attacks, and three patients (8.8%) had more than seven attacks of HAEC. The number of hospitalization due to HAEC was as follows: 15 patients (44.1%) once, six patients (17.7%) two times, five patients (14.7%) three times, and four patients (11.8%) four or more times. Four of these patients (11.8%) were hospitalized for four to seven days, six patients (17.7%) eight to 15 days, and 20 patients (58.8%) more than 16 days.

Microorganisms detected were as follows: E.coli (12 patients), E.coli + Klebsiella (5 patients), Klebsiella (1 patient), Klebsiella + Citrobacter (1 patient), Proteus + E.coli (1 patient), Proteus + E.coli + Klebsiella + Candida albicans (1 patient), Proteus + Enterobacter aerogenes (1 patient), Proteus + Enterobacter aerogenes + E.coli (1 patient), and Pseudomonas (1 patient).

Different combinations of antibiotics were used in patients with HAEC, mainly penicillin + tobramycin (gentamicin) + metronidazole (ornidazole), or trimethoprim-sulphamethoxazole (TMS) + metronidazole. In 11 patients antibiotics were used for five to seven days, in three patients eight to 12 days, and in 17 more than 12 days.

The treatment of postoperative HAEC was as follows: twelve patients underwent several dilatations, two patients sphincterotomy, five patients dilatations + sphincterotomy, four patients dilatations + colostomy opening, one patient dilatations + redo operation, two patients dilatations + ileorectal fistula repair, and two patients dilatations + bridectomy.

Discussion

Postoperative HAEC was detected in 34 (16.0%) patients. Different centers have reported different incidences of postoperative HAEC, some of which were consistent with our results[18-25]. Female patients constituted 14.7 percent of the HAEC patients (female: male ratio 1:5.8). Other parameters such as age on admission, birthweight, gestational age, family history and associated congenital anomalies either were not significantly different or were not suitable for statistical analysis. We encountered consanguinity less frequently in the HAEC group. Although this might suggest that postoperative HAEC did not have a congenital origin, some other parameters such as family history and associated congenital anomaly did not necessarily support this.

The time of passage of meconium, defacation patterns, and the presence of abdominal distention on physical examination on first admission, intestinal obstruction and intestinal perforation before definitive operation was almost the same in the two groups.

The presence of preoperative HAEC and the number of attacks did not create a risk factor for postoperative HAEC. Similar results were present in the series of Tietelbaum et al.[14] In the presented series, 6.1 percent of patients with postoperative HAEC also had preoperative HAEC. Elhalaby et al.[26] reported a 4.8 percent occurrence of postoperative HAEC after preoperative HAEC.

In the present series we report a high incidence of SS disease (73.5%) with postoperative HAEC, with no statistical difference from the group without HAEC.

In all of the patients with postoperative HAEC, definitive operation was performed after colostomy procedure. No difference was found between the patients with and without HAEC in terms of the presence of colostomy before definitive operation. In older children and in cases with considerable colonic dilatation, the colostomy procedure is necessary to facilitate the definitive operation. Surgeons who prefer performing definitive operation in the newborn period without colostomy report a very low incidence of postoperative HAEC.

No relation was found between the diarrhea after colostomy and postoperative HAEC. Similarly, we did not detect a relation between preoperative HAEC and diarrhea after colostomy in the first part of our study. In our study colostomy closure during definitive operation or soon after definitive operation did not have any effect on the occurrence of postoperative HAEC.

We detected postoperative HAEC in 22 patients (63.6%) after Swenson operation, 11 (33.3%) after Duhamel operation, and one (3.0%) after Boley operation. Previous series have reported different percentages for postoperative HAEC in different types of operations for HD[10, 19, 20, 23]. In the present series no difference was detected between the two groups in terms of type of operation (Swenson and Duhamel operations were compared).

Both the age and weight of the patient were risk factors for postoperative HAEC. It seems that if the symptoms of the disease are mild enough to enable the patient to survive with the disease with little or no complaints until the age of 15 months or a weight of greater than 10 kg, the risk of postoperative HAEC decreases.

Complications due to definitive operation, including anastomotic stricture, had no effect on the development of HAEC. Tietelbaum et al.[14] reported that among their four patients with postoperative HAEC, three had anastomotic stricture.

HAEC attacks could start as early as within the first postoperative month and as late as one year later. In 64.7 percent of patients, only one to three HAEC

attacks were detected, and the number of HAEC attacks decreased with time. Similar results were reported by Elhalaby et al.[26] The number of hospitalizations decreased in accordance with the number of attacks, but the period of hospitalization was still high (more than 16 days in 58.8% of patients). In most of the patients the microorganism detected was E.coli. Klebsiella and Proteus were the other two common microorganisms. We preferred penicillin + tobramycin (gentamicin) + metronidazole in severe cases, and TMS + metronidazole or metronidazole alone in mild cases. In the surgical treatment of postoperative HAEC we started with dilatations; other modes of treatment included sphincterotomy, colostomy opening, ileorectal fistula repair, bridectomy and redo operation with success. The colostomies of the four patients were closed later when the barium enema of the distal colon returned to normal. In two patients the signs and symptoms of HAEC obscured those of ileorectal fistula, causing a delay in diagnosis and treatment. Two patients returned to normal after dilatations and bridectomy. In one patient Duhamel operation was performed after Boley operation to treat intractable enterocolitis. None of these patients had the complaint of diarrhea at that moment.

Since postoperative HAEC is frequently encountered following definitive operation among patients weighing less than 10 kg or younger than 15 months of age, delaying the definitive operation beyond these criteria might be preferable.

REFERENCES

1. Swenson O, Bill AH. Resection of rectum and rectosigmoid with preservation of the sphincter for benign spastic lesions producing megacolon. Surgery 1948; 24: 212-220.
2. Duhamel B. A new operation for the treatment of Hirschsprung's disease. Arch Dis Child 1960; 35: 38-39.
3. Soave F. A new surgical technique for the treatment of Hirschsprung's disease. Surgery 1964; 56: 1007-1014.
4. State D. Segmental colon resection in the treatment of congenital megacolon (Hirschsprung's disease). Am J Surg 1963; 105: 93-101.
5. Boley SJ. New modification of the surgical treatment of Hirschsprung's disease. Surgery 1964; 5: 1015-1017.
6. Martin LW. Surgical management of Hirschsprung's disease involving the small intestine. Arch Surg 1968; 183-188.
7. Grob M, Genton N, Ventobel V. Experiences with surgery of congenital megacolon and suggestions of a new surgical technique. Zentralbl Chir 1959; 44: 1781-1789.
8. Sieber WK. Hirschsprung's Disease. In: Welch KJ, Randolph JG, Ravitch MM, O'Neill JA Jr, Rowe MI (eds). Pediatric Surgery (4th ed) Vol: 2. Chicago: Year Book Medical Publishers Inc; 1986; 995-1019.
9. Bill AH Jr, Chapman ND. The enterocolitis of Hirschsprung's disease. Its natural history and treatment. Am J Surg 1962; 103: 70-74.
10. Kleinhaus S, Boley SJ, Sheran M, Sieber WK. Hirschsprung's disease. A survey of the members of the Surgical Section of the American Academy of Pediatrics. J Pediatr Surg 1979; 14: 588-597.

11. Ikeda K, Goto S. Diagnosis and treatment of Hirschsprung's disease in Japan. An analysis of 1628 patients. Ann Surg 1988; 199: 400-405.

12. Swenson O, Fisher JH, Scott JE. Diarrhea following rectosigmoidectomy for Hirschsprung's disease. Pediatr Surg 1960; 48: 419-421.

13. Clausen EG, Davies OG Jr. Early and late complications of the Swenson pull-through operation for Hirschsprung's disease. Am J Surg 1963; 106: 372-380.

14. Tietelbaum DH, Qualman SJ, Caniano DA. Hirschsprung's disease: identification of risk factors for enterocolitis. Ann Surg 1988; 207: 240-244.

15. Klein MD, Coran AG, Wesley JR, Drongowsk RA. Hirschsprung's disease in the newborn. J Pediatr Surg 1984; 19: 370-374.

16. Carcassonne M, Morisson-Lacombe G, Letourneau JN. Primary corrective operation without decompression in infants less than three months of age with Hirschsprung's disease. J Pediatr Surg 1982; 17: 241-243.

17. Carcassonne M,Guys JM, Morisson-Lacombe G, Kreitmann B. Management of Hirschsprung's disease: curative surgery before three months of age. J Pediatr Surg 1989; 24: 1032-1034.

18. Rescorla FJ, Morrison AM, Engles D, West KW, Grosfeld JI. Hirschsprung's disease: evaluation of mortality and long-term function in 260 cases. Arch Surg 1992; 127: 934-942.

19. Polley TZ Jr, Coran AG, Wesley JR. A ten-year experience with ninety-two cases of Hirschsprung's disease. Ann Surg 1985; 202: 349-355.

20. Grosfeld JL, Ballantinell TV, Csicsko JF. A critical evaluation of the Duhamel operation for Hirschsprung's disease. Arch Surg 1978; 113: 454-460.

21. Foster P, Cowan G, Wrenn EL Jr. Twenty-five years' experience with Hirschsprung's disease. J Pediatr Surg 1990; 25: 531-534.

22. Puri P. Hirschsprung's disease: clinical and experimental observations. World J Surg 1993; 17: 374-384.

23. Sherman JO, Snyder ME, Weitzman JS, et al. A 40-year multinational retrospective study of 880 Swenson procedures. J Pediatr Surg 1989; 24: 833-845.

24. Surana R, Quinn FM, Puri P. Evaluation of risk factors in the development of enterocolitis complicating Hirschsprung's disease. Pediatr Surg Int 1994; 9: 234-236.

25. Carnerio PM, Brereton RJ, Drake DP, et al. Enterocolitis in Hirschsprung's disease. Pediatr Surg Int 1992; 7: 356-360.

26. Elhalaby EA, Coran AG, Blane CE, et al. Enterocolitis associated with Hirschsprung's disease: a clinical-radiological characterization based on 168 patients. J Pediatr Surg 1995; 30: 76-83.