The Possible Association of Hirschsprung Disease and Inflammatory Bowel Disease in Children

Hirschsprung disease (HD) is caused by lack of migration of neural crest cells in the colon leading to impaired colonic motility with symptoms that include constipation, HD-associated enterocolitis, and potential life-threatening toxic megacolon. Intestinal inflammation is present in HD-associated enterocolitis, and prior case series have suggested that some patients with HD can have inflammatory bowel disease (IBD) as well. However, this association is unclear, and the authors of this study attempted to describe the risk of IBD occurrence in pediatric patients with HD.

This cohort study occurred over 50 years using data from the Swedish National Patient Register and the Swedish Drug Register. Patients were considered to have HD if HD was the main diagnosis and the patient had a surgery specific for HD, if the cause of at least two hospitalizations (with one hospitalization being greater than 4 days) was HD, or if the cause of one long admission (greater than 4 days) and more than one outpatient clinic visit was HD. Patients with IBD also were identified from the Swedish National Patient Register, and a cohort of 739 patients with HD was compared to 7390 age- and sex-matched controls. The median age of study participants was 25 years (range 16-49 years). No sex significant differences between children with or without HD were noted, and there was no difference in the median age of IBD diagnosis between these two groups. Forty-one patients in the cohort without HD had IBD while 20 patients in the group with HD had IBD. Significantly more subjects with HD had IBD compared to those patients without IBD (95% CI: 2.29–8.19). The group of subjects who had both HD and IBD consisted of 15 patients with Crohn disease and 5 patients with ulcerative colitis while the group of control subjects with IBD consisted of 18 patients with Crohn disease and 23 patients with ulcerative colitis.

Further investigation is needed to determine why such a potential association is present between HD and IBD. The authors speculate that genetic factors, environmental factors (such as smoking), or microbiome differences may explain these findings. It is important to remember to evaluate patients with HD for associated IBD if suspicious gastrointestinal symptoms are occurring.


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