

ARE ALL PATIENTS WITH SHORT SEGMENT HIRSCHSPRUNG'S DISEASE EQUAL? --- A RETROSPECTIVE MULTICENTER STUDY

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Aim of the study: Short segment Hirschsprung's disease (HSCR) carries a better prognosis than long segment disease but the definition of short is controversial. The objective of this study is to determine anatomically the extent of disease involvement that is associated with a better functional outcome.

Methods: This is a retrospective multicenter (n = 3) study with patients (≥ 3 years) who had transanal pullthrough operation done were reviewed. The extent of disease involvement was retrieved by reviewing the operative as well as histopathological reports of the specimens. Clinical assessment was performed with a seven-itemed bowel function score (BFS) (maximum score = 20). Manometric assessment was performed with anorectomanometry.

Main results: Forty-four patients were studied with median age at assessment = 52.0 months and operation = 3.0 months. The disease involvement was categorized into upper sigmoid/descending colon (DC) (n=8), sigmoid colon (SC) (n=12), upper rectum (UR) (n=14) and lower rectum (LR) (n=10). There was no significant difference in the age of assessment between the four groups. The median BFSs in the DC, SC, UR and LR were 13, 15, 17 and 17 respectively (p=0.01). Nine patients from the DC and SC groups reported soiling for more than twice per week. A higher proportion of patients in the UR and LR groups had a normal sphincter resting pressure but this was not statistically significant (DC vs SC vs UR vs LR = 62.5% vs 75.0% vs 85.7% vs 80.0%, p =0.10).

Conclusion: Patients with HSCR confined to the rectum appear to have a better functional outcome. Less bowel loss and colonic dissection maybe the underlying reasons. Although future studies with larger sample size and longer follow up period are required to validate the results of this study, it has provided a new insight to the definition of short segment disease in HSCR.