IS-9  Hirschsprung's disease in Korea—Clinical experience of 233 cases in single tertiary medical center—

Division of Pediatric Surgery, Department of Surgery, Yonsei University College of Medicine, Seoul, Korea

Seong Min Kim, Jung Oh, Seung Hoon Cho

BACKGROUND/METHOD: Treatment outcome of Hirschsprung's disease has much improved during past two decades in Korea. From May 1986 to May 2005, we performed operation for 233 cases of Hirschsprung's disease. The aim of this study is analysis of clinical experience of Hirschsprung's disease. We retrospectively reviewed records of operation and follow-up data of the patients. RESULTS: Male to female ratio was 3.57: 1. Median age at diagnosis was 1.7 month (2 day - 114.4 month). 9.4% (22/233) showed associated anomaly (six showed chromosomal anomaly (2.6%), six showed genitourinary anomaly (2.6%), four showed small bowel atresia (1.7%)). Transitional zone was located in upper and lower rectum in 21.5% (50/233), sigmoid colon in 60.9% (142/233), transverse colon in 10.7% (25/233), ileum (total colonic aganglionosis) in 6.4% (15/233), upper jejunum in 0.4% (1/233). Of the 233 patients, Duhamel operation was performed in 151 patients (64.8%), initial myectomy in 41 patients (17.6%), one stage transanal Soave pull-through in 15 patients (6.4%), Martin-Duhamel operation in 10 patients (4.3%). Median follow-up period was 95.2 month. Major complications include postoperative intestinal obstruction in 19/233 (8.2%), recurrent enterocolitis in 18/233 (7.7%), severe constipation in 13/233 (5.6%). Mortality was confined to long segmental aganglionosis 8/233 (3.4%). There was no significant difference of functional benefit among various pull-through methods for each kind of aganglionosis. CONCLUSION: Overall long term ability to show proper bowel movement was 88.8%. The highest morbidity occurred among patients who had long segmental aganglionosis or who had immediate postoperative bowel movement problem. Early transanal one stage pull-through operation during neonatal period has become our standard procedure for selected cases of rectosigmoid aganglionosis.