



ABSTRACT

Background: Hirschsprung's Disease (HSCR) is a congenital birth defect characterised by the absence of ganglion cells in the distal colon and thus resulting in intestinal obstruction. Aganglioneosis begins in the rectum and continues proximally across varying lengths. It is categorised into categories based on the extent of aganglioneosis. Many studies have been done in other countries but it is not done in DR. Sardjito Hospital yet.

Objectives: The main objective of this study is know the incidence of HSCR in DR. Sardjito Hospital. The secondary objectives are to know the male to female ratio of this disease, the clinical manifestations of HSCR such as abdominal distention and meconium passage more than 24hours after birth, the types of HSCR, the types of definitive surgery and also post-operative complications and mortality rate.

Method: This research is an observational descriptive study and 77 subjects were involved. The subject involved the patients of DR. Sardjito Hospital, Yogyakarta from January 2013 to October 2014. Subjects were recruited using consecutive sampling and included all patients that present to these hospitals with HSCR, as confirmed by clinical manifestation, barium enema and/or biopsy. Data were obtained from patients' medical records. Subjects with incomplete data were excluded.

Results: The incidence of HSCR in DR. Sardjito Hospital in 2013 was 1:789. In this research, the male (62): female (15) ratio was 4.13:1, mostly presenting with short segment disease (96.1%) and neonatal age of presentation (79.2%). The most common presenting feature was abdominal distention (94.9%), with Transanal Endorectal Pull Through (28.6%) method being the most commonly used surgical method. The common post-operative complications were enterocolitis (15.6%) and constipation (10.4%). The mortality rate was at 6.5%.

Conclusions: HSCR is not an uncommon disease in DR. Sardjito Hospital. There were more male patients and most were short segmented disease. Patients commonly presented with abdominal distention during the neonatal period. The common post operative complications were enterocolitis and constipation with low mortality rate.

Keywords: Hirschsprung disease; incidence; clinical manifestation; enterocolitis; constipation.