



ABSTRAK

Latar Belakang: Penyakit Hirschsprung adalah penyakit kongenital di mana terdapat keabsenan sel ganglia pada usus, sehingga usus tidak bisa berelaksasi saat peristaltik. Tidak jarang penyakit Hirschsprung menyebabkan penurunan kualitas hidup penderita melalui komplikasinya, seperti *Hirschsprung-associated enterocolitis*. Jika dibiarkan, HAEC dapat menyebabkan kematian. Manajemen klinis yang tepat telah terbukti dapat mengurangi risiko komplikasi, bahkan mengobati kondisi aganglia pasien Hirschsprung. Alur manajemen yang efektif dimulai dari proses penegakan diagnosis yang tidak hanya akurat, tetapi juga tepat waktu.

Tujuan: Penelitian ini bertujuan untuk mengidentifikasi hubungan *late diagnosis* HSCR dengan kejadian HAEC pasca prosedur *pull-through*.

Metode: Penelitian ini menggunakan metode kohort retrospektif dengan populasi studi terdiri dari seluruh pasien penyakit Hirschsprung di RSUP Dr. Sardjito dari tahun 2020—2025 yang telah menjalani prosedur *pull-through*. Sampel yang diikutsertakan sudah memenuhi kriteria dan dianalisis menggunakan uji bivariat serta multivariat agar variabel *confounding* dapat terkendali.

Hasil: Jumlah kasus HAEC pada penelitian ini tidak dalam jumlah besar (0-16,3%) dengan median skor diagnosis HAEC berupa nol yang disertai pola distribusi *right-skewed*. Dari analisis bivariat dan multivariat, didapatkan p dari hubungan *late diagnosis* dengan kejadian HAEC lebih besar dari 0,05. Skor HAEC pasien *early* dan *late diagnosis* tidak terdapat perbedaan yang bermakna ($p = 0,245$) dengan *effect size* yang kecil (0,133).

Kesimpulan: Pada pasien Hirschsprung RSUP Dr. Sardjito periode 2020-2025, *late diagnosis* tidak didapatkan meningkatkan risiko HAEC pasca prosedur *pull-through*. *Late diagnosis* juga tidak didapatkan memiliki hubungan dengan skor HAEC pasien. Akan tetapi, temuan perlu diinterpretasikan dengan hati-hati karena jumlah sampel yang minim.

Keyword: Hirschsprung; *late diagnosis*; HAEC; prosedur *pull-through*



ABSTRACT

Background: Hirschsprung's disease (HSCR) is a congenital disorder where there is an absence of ganglion cells in the intestine, which prevents intestine from relaxing during peristaltic. It is not uncommon for the disease to decrease the patients' quality of life through its complication, one of it being Hirschsprung-associated enterocolitis (HAEC). If left untreated, HAEC can lead to death. Proper managements has been proven to reduce risk of complications and definitively treat the aganglionic condition in Hirschsprung's disease patients. An effective management flow begins with a diagnostic process that is not only accurate but also timely.

Objective: This study aims to identify the relationship between late diagnosis of HSCR and the occurrence of HAEC post pull-through procedure.

Methods: This study used a retrospective cohort method with a study population consisting of Hirschsprung's disease patients in RSUP Dr. Sardjito from 2020-2025 who have undergone pull-through procedure. Samples included in this study have fulfilled the criterias and were analyzed using bivariate and multivariate tests so that confounding variables could be controlled.

Results: The number of HAEC cases in this study was not large (0-16,3%) with a median HAEC diagnosis score of zero accompanied by a right-skewed distribution pattern. From bivariate and multivariate analyses, the p value of the relationship between late diagnosis and HAEC occurrence was greater than 0,05. There was no significant difference in HAEC scores between early and late diagnosis patients ($p = 0,245$) with a small effect size (0,133).

Conclusions: In Hirschsprung's disease patients at Dr. Sardjito General Hospital during the period 2020-2025, late diagnosis was not found to increase the risk of HAEC post procedure. Late diagnosis was also not found to be associated with patient's HAEC scores. However, findings need to be interpreted with caution due to small sample size.

Keyword: Hirschsprung; late diagnosis; HAEC; pull-through procedure

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