ABSTRAK

Latar Belakang: Penyakit Hirschsprung adalah suatu penyakit genetik yang kompleks, ditandai dengan tidak adanya ganglion enterik pada submukosa dan pleksus myenterikus yang bervariasi sepanjang usus bagian distal. Kebanyakan pasien mengalami perbaikan pasca *pull-through*, namun beberapa diantaranya mengalami gejala obstruksi persisten. Kami bertujuan mengidentifikasi faktor prognosis terhadap gejala obstruksi persisten pasca operasi Duhamel, *transanal endorectal* dan Swenson *like pull-through* pada pasien dengan penyakit Hirschsprung di institusi kami.

Metode: Penelitian ini menggunakan desain retrospektif. Kami menganalisis rekam medis pasien penyakit Hirschsprung dengan atau tanpa gejala obstruksi persisten pasca Duhamel, *transanal endorectal*, dan Swenson *like pull-through* yang datang ke rumah sakit dari Januari 2017 hingga Januari 2022.

Hasil: Seratus empat belas pasien masuk dalam penelitian: 79 laki-laki dan 35 perempuan. Kebanyakan pasien (90,4%) memiliki *short* aganglionosis. Pasien yang menjalani Duhamel (n = 38), *transanal endorectal* (n=63), and *transanal* Swenson *like pull-through* (n = 13). Sebanyak (21.9%) mengalami obstruksi persisten pasca operasi Duhamel, *transanal endorectal* dan Swenson *like pull-through*. Teknik *pull-through* dan usia saat operasi definitif berhubungan kuat dengan gejala obstruksi persisten pasca *pull-through* (masing-masing, *p*=0,032 dan 0,019), sedangkan jenis kelamin, tipe aganglionosis, *global developmental delay* dan sindrom Down, tidak (masing-masing, *p*=0,837; 0,525; 0,647 dan 0,301). Analisis multivariat menunjukkan bahwa usia saat operasi definitif merupakan faktor independent yang kuat terhadap kejadian gejala obstruksi persisten pasca *pull-through* (OR = 3.41 [95% CI 1.18-9.91]; p = 0.02).

Kesimpulan: Frekuensi gejala obstruksi persisten pasca *pull-through* di institusi kami relatif rendah. Selain itu, pasien yang menjalani *pull-through* diusia muda lebih mungkin mengalami gejala obstruksi persisten.

Kata Kunci: usia saat operasi definitif; penyakit Hirschsprung; gejala obstruksi persisten; *pull-through*, faktor prognosis.

ABSTRACT

Background: Hirschsprung disease is a complex genetic disorder characterized by the lack of enteric ganglia in the submucosal and myenteric plexuses, along variable portions of the distal gut. Most children had a good result after pull-through, but some of them had a persistent obstructive symptoms. We aimed to identify prognostic factors for persistent obstructive symptoms after Duhamel, transanal endorectal, and Swenson like pull-through in patients with Hirschsprung's disease in our institution.

Methods: This study was a retrospective design. We analyzed the medical records of Hirschsprung's disease patients with or without persistent obstructive symptoms after Duhamel, transanal endorectal, and Swenson like pull-through that admitted to our hospital from January 2017 to January 2022.

Results: One hundred and fourteen patients were included in this study: 79 males and 35 females. Most of them (90,4%) had short aganglionosis. They underwent the Duhamel (n = 38), transanal endorectal (n=63), and Swenson like pull-through (n = 13). Twenty one point nine (21,9%) patient showed persistent obstructive symptoms after Duhamel, transanal endorectal, and Swenson like pull-through. Pull-through approach and age at definitive surgery were strongly associated with persistent obstructive symptom after pull-through (p=0,032 and 0,019, respectively), while sex, aganglionosis type, presence of global developmental delay and Down syndrome were not (p=0,837; 0,525; 0,647 and 0,301, respectively). Multivariate analysis showed that age at definitive surgery was a strong independent factor for persistent obstructive symptoms after pull-through. (OR = 3.41 [95% CI 1.18-9.91]; p = 0.02).

Conclusions: The frequency of persistent obstructive symptoms after pull-through in our institution is considered low. Moreover, patients who underwent pull-through at a younger age might more likely suffer from persistent obstructive symptoms.

Keywords: age at definitive surgery; Hirschsprung disease; persistent obstructive symptoms; pull-through; prognostic factor.