

## INTISARI

**Latar belakang.** Hirschsprung (HSCR) adalah penyakit kelainan genetik kompleks berupa aganglionosis pleksus submukosa (Meissner) dan mienterikus (Auerbach) serta hipertrofi serabut saraf. HSCR ditemukan pada 1/5.000 kelahiran dengan rasio laki-laki dibanding perempuan adalah 4:1. Diagnosis ditegakkan melalui biopsi rektal ketebalan penuh dan pemeriksaan histopatologis menggunakan pengecatan hematoxylin-eosin (HE) dan calretinin.

**Tujuan.** Untuk mengetahui akurasi dan konkordansi pengecatan HE dibandingkan calretinin dalam mendeteksi sel ganglion pada penyakit HSCR di RSUP Dr. Sardjito.

**Metode.** Desain penelitian adalah observasional dengan pendekatan potong-lintang melalui data pasien HSCR RSUP Dr. Sardjito tahun 2017-2021. Uji diagnostik yang ditelaah adalah akurasi, sensitivitas, spesifisitas, nilai ramal positif, dan nilai ramal negatif. Signifikansi uji diagnostik dianalisis menggunakan Mc Nemar Test. Analisis kesepakatan antara kedua teknik diuji menggunakan Cohen's Kappa Statistic.

**Hasil.** Terdapat 29 pasien suspek HSCR yang dibagi menjadi tiga kelompok usia, yaitu neonatus, 29 hari -  $\leq 1$  tahun, dan  $> 1$  tahun. Hasil akurasi, sensitivitas, spesifisitas, nilai ramal positif pada keseluruhan sampel adalah 79.31% (95% CI: 0.60-0.92), 83.30% (95% CI: 0.62-0.96), 72.70% (95% CI: 0.44-0.92), 83.30% (95% CI: 0.62-0.96), dan 72.70% (95% CI: 0.44-0.92) secara berurutan. Pada kelompok neonatus (n=8) adalah 87.50% (95% CI: 0.47-0.99), 85.70% (95% CI: 0.42-0.99), 100% (95% CI: 0.25-1.00), 100%, dan 50% (95% CI: 0.14-0.85), sedangkan pada kelompok usia 29 hari -  $\leq 1$  tahun (n=20) adalah 75% (95% CI: 0.50-0.91), 80% (95% CI: 0.44-0.97), 70% (95% CI: 0.35-0.93), 72.73% (95% CI: 0.50-0.87), dan 77.78% (95% CI: 0.49-0.92) secara berurutan. Uji diagnostik tidak menunjukkan perbedaan yang signifikan secara statistik ( $p=1.00$ ) dengan indeks Kappa Cohen adalah 0.56 (persetujuan moderat). Hasil ini menunjukkan bahwa pengecatan HE dan calretinin konkordan dengan derajat persetujuan moderat.

**Kesimpulan.** Akurasi pengecatan HE dibandingkan calretinin adalah 79.31%. Kedua pengecatan konkordan dengan derajat persetujuan moderat, sehingga HE masih dapat digunakan dalam penegakan diagnosis HSCR.

**Kata Kunci.** Hirschsprung, hematoxylin-eosin, calretinin, sel ganglion, akurasi

## ABSTRACT

**Background.** Hirschsprung (HSCR) is a genetic disorder with a submucosal (Meissner) and myenteric (Auerbach) plexus aganglionosis and nerve fiber hypertrophy. It is found in 1/5.000 births with a male to female ratio of 4:1. HSCR diagnosis is confirmed by full-thickness rectal biopsy and histopathological examination using hematoxylin-eosin (HE) and calretinin staining to identify ganglion cells.

**Objective.** to determine the accuracy and concordance of HE staining compared to calretinin in identifying ganglion cells in HSCR disease at Dr Sardjito General Hospital, Yogyakarta.

**Method.** This study was an observational with cross-sectional approach using a HSCR patient data at Dr Sardjito General Hospital Yogyakarta in 2017 - 2021. Accuracy, sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) were studied. The significance of those tests was analyzed using a Mc Nemar Test. Analysis of agreement between two techniques was tested using a Cohen's Kappa Statistics.

**Result.** There were 29 patients with suspected HSCR divided into three age groups, namely neonates, 29 days -  $\leq 1$  year, and  $> 1$  year. The results for accuracy, sensitivity, specificity, PPV, and NPV for the entire sample were 79.31% (95% CI: 0.60-0.92), 83.30% (95% CI: 0.62-0.96), 72.70% (95% CI: 0.44-0.92), 83.30% (95% CI: 0.62-0.96), and 72.70% (95% CI: 0.44-0.92) respectively. In the neonate group (n=8) was 87.50% (95% CI: 0.47-0.99), 85.70% (95% CI: 0.42-0.99), 100% (95% CI: 0.25-1.00), 100%, and 50% (95% CI: 0.14-0.85), whereas in 29 days -  $\leq 1$  year group (n=20) was 75% (95% CI: 0.50-0.91), 80% (95% CI: 0.44-0.97), 70% (95% CI: 0.35-0.93), 72.73% (95% CI: 0.50-0.87), and 77.78% (95% CI: 0.49-0.92) respectively. The diagnostic test showed no statistically significant difference (p=1.00) with the Cohen's Kappa index being 0.56 (moderate agreement). These results indicate that HE and calretinin staining is concordant with a moderate degree of agreement.

**Conclusion.** HE staining accuracy compared to calretinin is 79.31%. Both staining are concordant with a moderate degree of agreement, so that HE staining can still be used in the diagnosis of HSCR.

**Keywords.** Hirschsprung, hematoxylin-eosin, calretinin, ganglion cells, accuracy