# Studi neurofisiologi pada anak thalassemia mayor dan faktor-faktor yang memengaruhinya = Neurophysiology study and related risk factors in children with thalassemia major

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Abstrak

Thalassemia merupakan salah satu kelainan genetik paling umum di seluruh dunia. Tanpa managemen yang adekuat, komplikasi akan terjadi pada berbagai organ, termasuk sistem saraf. Tujuan penelitian ini adalah untuk menentukan proporsi abnormalitas Brain Auditory Evoked Potentials (BAEP), elektroensefalografi (EEG), dan elektroneurografi (ENG) pada anak thalassemia mayor dan hubungannya dengan faktor risiko terkait. Metode: Penelitian ini merupakan studi potong lintang deskriptif analitik yang dilakukan di Rumah Sakit Cipto Mangunkusumo (RSCM) Jakarta dan Rumah Sakit M Djamil (RSMDJ) Padang. Kriteria inklusi adalah anak thalassemia mayor berusia 12-18 tahun yang kontrol teratur minimal dalam 1 tahun terakhir. Pasien dengan epilepsi, palsi serebral, gangguan pendengaran, dan gangguan neurodevelopmental dikeluarkan dari penelitian. Pemeriksaan BAEP, EEG, dan ENG dilakukan pada semua subyek dan diinterpretasikan oleh konsultan neuropediatri. Dilakukan pencatatan usia onset, durasi transfusi, rerata hemoglobin (Hb) pra-transfusi, kadar feritin serum, saturasi feritin, dan kepatuhan konsumsi obat kelasi besi. Hubungan antar variabel dinilai menggunakan analisis bivariat dengan nilai $p < 0,05$ dikatakan bermakna. Hasil: Sebanyak 64 anak dengan rerata usia 15,1 tahun memenuhi kriteria penelitian, terdiri atas 29 anak laki-laki dan 35 anak perempuan. Rerata Hb pra transfusi, kadar feritin serum, dan saturasi transferin berturut-turut adalah 8,36 g/dL, 4495,3 ng/mL, dan 87,3%. Abnormalitas EEG ditemukan pada 28 (43,8%) subyek dan berhubungan bermakna dengan rerata Hb pra-transfusi < 9 g/dL (p=0,011, rasio prevalensi 3,014, interval kepercayaan 1,04-8,71). Abnormalitas BAEP ditemukan pada 4 (4,6%) subyek dan berhubungan bermakna dengan kadar feritin serum yang lebih tinggi (p=0,007). Hasil ENG abnormal hanya ditemukan pada 1 orang subyek. Tidak terdapat hubungan antara faktor risisko lainnya dengan masing-masing pemeriksaan neurofisiologi. Kesimpulan: Abnormalitas EEG ditemukan pada 43,8% anak thalassemia mayor dan berhubungan dengan rerata Hb pra-transfusi <9 g/dL, sedangkan abnormalitas BAEP ditemukan pada 4% subyek dan berhubungan dengan kadar feritin serum yang lebih tinggi.
.....Thalassemia is among the most common genetic disorders worldwide. Without adequate management, complications occur in various organs as well as neurology system. The aim of the study was to determine the proportion of abnormal electroencephalography, brain auditory evoked potentials (BAEP), and nerve conduction study (NCS) in children with thalassemia major and its association with related risk factors. Methods: This was a descriptive-analytic cross sectional study conducted in Cipto Mangunkusumo Hospital and M Djamil Hospital in January to March 2019 All children with thalassemia major aged 12 to 18 years were eligible for the study. Children with epilepsy, palsy cerebral, hearing disorder, or neurodevelopmental problems were excluded. Electroencephalography, BAEP, and NCS were performed in all subjects. Age of onset, transfusion duration, mean pre-transfusion hemoglobin, serum ferritin, transferrin saturation, and compliance to chelating agents therapy were recorded. Bivariate analysis was performed to determine the relationsip between variables with $p < 0,05$ was considered significant. Results: As many as 64 children with mean age 15,1 years fulfilled the study criteria during the study period, consisting of 29 boys and 35 girls.

Mean pre-transfusion hemoglobin, serum ferritin, and transferrin saturation was 8,36 g/dL, 4495,3 ng/mL, and 87,3% respectively. Abnormal EEG was found in 28 (43,8%) subjects and significantly associated with mean Hb below 9 g/dL (p = 0,011; prevalence ratio 3,014; confidence interval 1,04 - 8,71). Abnormal BAEP was found in 4 (4,6%) subjects and significantly associated with higher serum ferritin (p=0,007). Only 1 subject showed abnormal NCS. No association was found between other risk factors with each neurophysiology study. Conclusion: Abnormal EEG was found in 43,8% thalassemia major children and significantly associated with lower pre-transfusion hemoglobin level. Abnormal BAEP was found in 4% subjects and significantly associated with higher serum ferritin.