

Faktor risiko timbulnya inhibitor faktor VIII pada anak dengan Hemofilia A = The risk factors of factor VIII inhibitor in children with Hemophilia A / Grace Natalia Adriana Simatupang

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Abstrak

ABSTRAK

Latar belakang. Proses timbulnya inhibitor bersifat multifaktorial baik genetik maupun lingkungan. Beberapa studi telah dilakukan untuk mengetahui faktor risiko terbentuknya inhibitor namun masih terdapat kontroversial pendapat. Tidak seperti di negara maju, di Indonesia skrining inhibitor tidak rutin dilakukan karena keterbatasan biaya dan alat sehingga diperlukan suatu penelitian yang dapat dijadikan acuan pemeriksaan inhibitor selektif.

Tujuan. Mengetahui prevalens, karakteristik klinis dan faktor risiko timbulnya inhibitor pada anak dengan hemofilia A di departemen IKA- RSCM.

Metode. Uji potong lintang dilakukan pada anak usia ≤18 tahun dengan perdarahan akut di pusat hemofilia terpadu IKA-RSCM. Pada subjek dilakukan pengambilan darah vena dan dilakukan pemeriksaan inhibitor menggunakan metode Bethesda assay. Orangtua diminta mengisi kuesioner mengenai usia saat pertama kali didiagnosis hemofilia, mendapat terapi faktor VIII, jenis terapi pengganti, derajat hemofilia, jenis perdarahan, dan suku bangsa ibu penderita. Analisis bivariat dilakukan dengan uji Fisher. Analisis multivariat tidak dilakukan karena tidak memenuhi syarat.

Hasil penelitian. Dari 40 subjek penelitian, didapatkan prevalens inhibitor sebanyak 37,5% (15/40) dengan inhibitor high responder sebanyak 3/15 dan low responder 12/15. Median (rentang) usia subjek penelitian adalah 10 (1,5-18) tahun. Median usia saat diagnosis hemofilia pertama kali ditegakkan dan saat pertama kali mendapat terapi faktor VIII pada inhibitor positif adalah 8 dan 9 bulan. Hampir seluruh subjek (39/40) mendapat terapi konsentrat plasma, 11/15 subjek dengan inhibitor positif mendapat terapi pertama kali sebelum berusia 1 tahun, 14/15 subjek merupakan hemofilia berat dan sebagian besar (12/15) mendapat manifestasi perdarahan sendi. Suku bangsa ibu Jawa lebih sering ditemukan pada inhibitor positif (8/15). Tidak ditemukan hasil yang bermakna secara statistik antara faktor risiko dengan timbulnya inhibitor.

Simpulan. Prevalens inhibitor pada penelitian ini sebesar 37,5%. Inhibitor positif lebih sering ditemukan pada penderita hemofilia berat yang mendapat terapi pertama kali sebelum berusia 1 tahun. Penelitian ini tidak berhasil membuktikan faktor risiko bermakna untuk timbulnya inhibitor pada anak dengan hemofilia A.

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ABSTRACT

Background. Several factors may influence inhibitor incidence including genetics and environment. Several studies have been conducted to determine the risk factors for inhibitor formation but there is still a

controversial opinion. Unlike in developed countries, in Indonesia inhibitor screening is not routinely performed due to limited funds thus required a research that can be used as reference checks selective inhibitors.

Objective. To find out the prevalence, clinical characteristics and risk factors of factor VIII inhibitor in children with hemophilia A in Child Health Department- Cipto Mangunkusumo Hospital

Methods. A cross sectional descriptive study conducted in children aged 1; 18 years old with acute bleeding at the National Hemophilia Care Center, Cipto Mangukusumo Hospital. All the subjects performed venous blood sampling and the examination of inhibitor using the Bethesda assay. Parents were asked to fill out questionnaires on age at first diagnosis of hemophilia, treated with factor VIII replacement therapy type, degree of hemophilia, types of bleeding, and the patient's mother tribes. Bivariate analysis performed by Fisher's test. Multivariate analysis was not performed because it does not qualify.

Results. Out of 40 children study, showed prevalence inhibitor 37.5% (15/40) with a high responder inhibitor 3/15 and low responders 12/15. Median (range) age of subjects was 10 (1.5 to 18) years. The median age at diagnosis of hemophilia was first established and the first time the subjects get a factor VIII inhibitor therapy positive was 8 and 9 months. Almost all subjects (39/40) treated with plasma concentrates, 11/15 subjects with a positive inhibitor therapy gets first time before age 1 year, 14/15 subjects is severe hemophilia and most (12/15) of them had joint bleeding manifestations. Java native tribes more often found in the positive inhibitor (8/15). No results found a statistically significant association between the risk factors with the onset of inhibitor.

Conclusion. The prevalence of inhibitors in this study was 37.5%. Positive inhibitors was more frequent in patients with severe hemophilia who received therapy for the first time before the age of 1 year old. This study failed to prove significant risk factor for the onset of inhibitors in children with hemophilia A.