Gambaran Profil Asam Amino Pada Pasien Thalassemia Mayor Di Rscm Menggunakan Liquid Chromatography Tandem Mass Spectrometry (Lc-Msms)

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Abstrak

ABSTRAK
Secondary iron overload pada thalassemia mayor terjadi karena eritropoiesis inefektif dan tranfusi berkala. Besi melebihi transferin sehingga banyak non transferin bound iron NTBI yang mengkatalisasi terjadinya ion radikal bebas yang merusak jaringan. Pengendapan besi pada saluran cerna mengakibatkan perubahan fungsi, kerusakan organ, gangguan ketersediaan asam amino. Iron overload dikurangi dengan kelasi besi. Transferin merupakan kelator alami tubuh terdiri asam amino dominan alanin, leusin, glisin, asam aspartat. Berdasarkan penelitian, pasien iron overload memiliki transferin lebih rendah dibandingkan non iron overload. Penelitian bertujuan mengetahi perubahan status besi, profil asam amino dan hubungan iron overload dengan profil asam amino. Parameter yang diteliti : besi serum, unsaturated iron binding capacity UIBC , total iron binding capacity TIBC , feritin, saturasi transferin, indeks transferin, alanin, leusin, glisin, asam aspartat. Desain penelitian kohort dengan 21 subjek, yaitu 13 thalassemia beta mayor dan 8 thalassemia beta HbE. Hasil penelitian didapatkan perubahan status besi bermakna yaitu peningkatan feritin pasca transfusi, penurunan feritin pasca kelasi 1 bulan, peningkatan kadar besi pasca kelasi 3 bulan. Perubahan asam amino bermakna yaitu penurunan alanin, leusin, serta peningkatan glisin pasca kelasi 1 bulan Terdapat hubungan kuat, bermakna searah antara indeks transferin dan alanin pre transfusi. Terdapat hubungan kuat, bermakna, searah antara indeks transferin dengan alanin dan glisin pasca transfusi.
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ABSTRACT
Secondary iron overload in thalassemia major occurs due to ineffective erythropoiesis and periodic transfusions. The excess of iron exceed transferrin so there are many non transferrin bound iron NTBI that induce tissue damaging free radical ion. Accumulation of iron in intestine can lead to changes in the function, organ damage, lack of amino acid availability. Iron overload can be reduced by iron chelation. Transferrin is the body 39 s natural chelator comprising of dominant amino acid alanine, leucine, glycine, aspartic acid. Research found that transferrin were lower in iron overload patients. This study aims to acquire the changes of iron status, amino acid profile, and correlation between iron overload and amino acid profile. Studied parameter were serum iron, unsaturated iron binding capacity UIBC , total iron binding capacity TIBC , ferritin, transferrin saturation, transferrin index, alanine, leucine, glycine, aspartic acid. The study design were cohort with 21 subjects consisted of 13 beta major thalassemia and 8 beta Hbe thalassemia. The result showed significant iron status changes ferritin increased post transfusion, ferritin decreased after 1 month chelation and serum iron increased after 3 months chelation. Significant amino acid profile changes decreased of alanine and leucine, and glycin increased after 1 month chelation. There rsquo s significant correlation between transferrin index and alanine pre transfusion. There rsquo s significant correlation between transferrin index and alanine, glycine after 3 month chelation.