

Indeks transferin sebagai penanda perubahan status besi pasien thalassemia mayor pasca transfusi dan terapi kelasi = Index transferrin as a marker of iron status changes in major thalassemia patients post transfusion and post chelation therapy

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Abstrak

Pendahuluan: Thalassemia adalah suatu kelainan genetik akibat kegagalan sintesis rantai globin, mengakibatkan terjadinya anemia berat akibat peningkatan aktivitas eritropoiesis yang inefektif dan hemolisis. Peningkatan aktivitas eritropoiesis akan memacu peningkatan absorpsi besi di usus sehingga terjadi kelebihan besi dalam tubuh. Transfusi darah dilakukan secara berkala untuk mengatasi anemia yang timbul pada pasien thalassemia mayor. Pemberian transfusi berulang akan mempercepat terjadi secondary iron overload, untuk mengatasinya diberikan terapi kelasi rutin.

Tujuan : Mendapatkan perubahan nilai indeks transferin, saturasi transferin, dan feritin sebelum dan sesudah transfusi dan juga sebelum dan sesudah terapi kelasi pada pasien thalassemia mayor. Mendapatkan perbedaan indeks transferin dan saturasi transferin, dan feritin sebagai parameter untuk menilai perubahan status besi pada pasien thalassemia mayor pasca transfusi dan terapi kelasi.

Metode: Desain penelitian kohort prospektif. Subjek penelitian terdiri dari 35 pasien thalassemia mayor usia 7-18 tahun yang mendapat transfusi berulang dan kelator besi rutin. Dilakukan pemeriksaan kadar besi serum, UIBC, TIBC, feritin, transferin, saturasi transferin dan indeks transferin pre transfusi, pasca transfusi dan pasca terapi kelasi.

Hasil: Rerata indeks transferin pasca transfusi 124 ± 22 % lebih rendah secara bermakna dari pre transfusi dengan nilai $p=0,016$, sedangkan pasca kelasi 123 ± 34.5 % ($p=0,045$). Saturasi transferin pasca transfusi 96 ($51 - 100$)% meningkat secara bermakna dibandingkan pre transfusi 87 ($69-100$)% dengan nilai $p=0,026$, namun tidak berbeda bermakna pada pasca kelasi 87 ($39-100$). Kadar feritin serum pasca transfusi 3737 ($649 -17.094$) mg/dL, meningkat secara bermakna dibandingkan pre transfusi 3315 ($544,7-14.964$) mg/dL ($p=0,018$). Perbedaan indeks transferin dan saturasi transferin pre transfusi 45 ($22-153$)% lebih tinggi secara bermakna dibandingkan pasca transfusi 35 ($6-89$)% dengan nilai $p=0,000$, sedangkan pasca kelasi adalah 41 ± 25 dengan nilai $p=0,036$.

Kesimpulan: pemeriksaan indeks transferin untuk pemantauan efektifitas terapi kelasi pada pasien thalassemia mayor dapat dipertimbangkan.

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Introduction: Thalassemia syndromes are a group of hereditary disorder characterized by genetic deficiency in the synthesis of -globin chains. It is associated in severe anemia caused by an increase in ineffective erythropoiesis activity and hemolysis. Erythropoiesis activity will spur increased iron absorption in the intestine so there will be an excess of iron in the body. Blood transfusion is used routinely to treat anemia arising in patients with thalassemia major. Repeated transfusion will accelerate occur secondary iron overload, to solve given chelation therapy routine.

Objective: To know the index value changes transferrin, transferrin saturation, and ferritin before and after transfusion and also before and after chelation therapy in patients with thalassemia major. To know

difference transferrin index and transferrin saturation, and ferritin as a parameter to assess changes iron status in patients thalassemia major post-transfusion and chelation therapy.

Methods: This was prospective cohort, There were 35 patients with thalassemia major who receive repeated transfusions and iron kelator routine, with age 7-18 years. Examination of serum iron levels, UIBC, TIBC, ferritin, transferrin, transferrin saturation and transferrin index before transfusion, after transfusion, and after chelation therapy.

Results: Mean transferrin index post-transfusion $124 \pm 22\%$ was significantly lower than pre transfusion ($p=0.016$), as well as post-chelation $123 \pm 34.5\%$ with a value of $p=0.045$. Transferrin saturation post-transfusion $96 (51-100)\%$ increased significantly with pre transfusion $87 (69-100)\%$ with a value of $p=0.026$, However no significant difference were observed in post chelation therapy $87 (39-100)$. Post-transfusion serum ferritin level $3737 (649-17094)$ mg/dL, increased significantly compared to pre transfusion $3315 (544.7-14,964)$ mg/dL ($p=0.018$). Differences transferrin index and transferrin saturation pre transfusion was $45 (22-153)\%$ significantly higher than the post-transfusion $35 (6-89)\%$ with a value of $p=0.000$, while the post chelation therapy was $41 \pm 25\%$ ($p=0.036$).

Conclusion: Transferrin index can be considered for monitoring the effectiveness of chelation therapy in patients with thalassemia major.