

Profil fungsi tubulus ginjal pada transfusion dependent thalassemia remaja dan faktor yang memengaruhi = Renal tubule function in transfusion dependent thalassemia adolescent

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Abstrak

Latar belakang: Thalassemia merupakan kelainan hemoglobin yang diturunkan secara autosomal resesif terbanyak di dunia, termasuk di Indonesia. Eritropoiesis inefektif dan hemolisis yang mengakibatkan terjadinya anemia, disertai kelebihan besi, dan toksisitas kelasi besi dapat mengganggu fungsi organ, salah satunya ginjal.

Tujuan: Mengetahui profil tubulus ginjal dan faktor yang memengaruhi pada transfusion dependent thalassemia (TDT) remaja

Metode: Penelitian deskriptif analitik yang dilakukan dengan potong lintang. Subyek adalah TDT remaja usia 10-18 tahun yang datang ke RSCM pada Januari 2024 sampai Februari 2024. Subyek dieksklusi jika terdapat riwayat penyakit ginjal, demam, gejala infeksi saluran kemih, serta mendapatkan kortikosteroid, trimetoprim, aminoglikosida atau sefalosporin generasi pertama dalam 1 minggu terakhir berdasarkan anamnesis dan catatan medik. Setelah informed consent didapatkan dari orangtua, dilakukan pengambilan data demografi dasar, pengukuran status antropometri, dan pengukuran tekanan darah. Pengambilan sampel darah dilakukan sebelum transfusi rutin dan pengambilan sampel urin pertama di pagi hari untuk dilakukan pemeriksaan fungsi ginjal. Hasil: Dari 55 subyek, proporsi jenis kelamin 1:1 dengan median usia 14 (10-17) tahun. Median usia terdiagnosis thalassemia adalah 24 (2-180) bulan. Mayoritas subyek adalah thalassemia-! (72,7%), sedangkan thalassemia-!/HbE 27,3%. Status nutrisi gizi baik 45,5%, gizi kurang 34,5%, dan gizi buruk 20%. Jenis kelasi besi yang diberikan antara lain, deferipron (DFP) 61,8%, deferasiroks (DFX) 29,1%, dan kombinasi (DFP dan DFX) 7,3%, sedangkan 1 pasien tanpa terapi. Rerata kadar hemoglobin adalah 8,5 (SD 1,1) g/dL dengan nilai median feritin 4.604,2 (753,86-30.472,67) ng/dL. Prevalens disfungsi tubulus ginjal adalah 96,3%, meskipun prevalens penurunan fungsi ginjal hanya 1,8%. Profil fungsi tubulus ginjal berupa hiperurikosuria (94,5%), proteinuria (39,1%), peningkatan NGAL/kreatinin urin (23,6%), hiperkalsiuria (21,8%), dan hiperfosfaturia (20%). Tidak terbukti hubungan rasio NGAL/kreatinin urin terhadap anemia pre- transfusi, status kelebihan besi, dan jenis kelasi besi. Hiperfiltrasi glomerulus terjadi pada 63,6% subyek dan NGAL/kreatinin urin dan proteinuria memiliki hubungan yang bermakna terhadap hiperfiltrasi glomerulus dini ($p=0,029$ dan $p=0,025$).

Kesimpulan: Disfungsi tubulus banyak ditemukan pada TDT remaja. Tidak terbukti hubungan rasio NGAL/kreatinin urin terhadap anemia pre-transfusi, status kelebihan besi, dan jenis kelasi besi. Pemeriksaan fungsi ginjal sebaiknya dilakukan secara rutin pada TDT remaja.

.....Background: Thalassemia is the most common hemoglobin disorder in Indonesia. Ineffective erythropoiesis and haemolysis result in anemia, accompanied by iron overload, and iron chelation toxicity can disrupt organ function, one of which is the kidneys.

Objective: To determine renal tubule function and its contributing factors in transfusion dependent thalassemia (TDT) adolescents.

Method: This is an analytical descriptive study in cross-sectional. Subjects were TDT adolescent aged 10-18

years who came to RSCM from December 2023 to January 2024. Subjects were excluded if they had a history of kidney disease, fever, symptoms of urinary tract infection, and received corticosteroids, trimethoprim, aminoglycosides or first generation cephalosporins within the last 1 week based on anamnesis and medical records. Informed consent was obtained from the parents and basic demographic data, anthropometric status measurements and blood pressure measurements were taken. Blood samples are taken before routine transfusions and first morning urine samples are taken to check kidney function.

Results: Of the 55 subjects, the gender proportion was 1:1 with a median age of 14 (10- 17) years. The median age at diagnosis was 24 (2-180) months. The majority of subjects were α -thalassemia (72.7%), while α -thalassemia/HbE was 27.3%. The nutritional status was normal 45.5%, wasting 34.5%, and severe wasting 20%. The types of iron chelation given included deferiprone (DFP) 61.8%, deferasirox (DFX) 29.1%, and combination (DFP and DFX) 7.3%, while 1 patient had no therapy. The mean hemoglobin level was 8.5 (SD 1.1) g/dL with a median ferritin value of 4,604.2 (753.86-30,472.67) ng/dL. The prevalence of renal tubular dysfunction is 96.3%, although the prevalence of decreased renal function is only 1.8%. The renal tubular function profile consisted of hyperuricosuria (94.5%), proteinuria (39.1%), increased urinary NGAL/creatinine (23.6%), hypercalciuria (21.8%), and hyperphosphaturia (20%). There is no correlation between the urine NGAL/creatinine ratio and pre-transfusion anemia, iron overload status, and type of iron chelation. Glomerular hyperfiltration occurred in 63.6% subjects and urinary NGAL/creatinine and proteinuria had a significant correlation to early glomerular hyperfiltration ($p=0.029$ and $p=0.025$).

Conclusion: Tubular dysfunction is often found in adolescent TDT. There is no relationship between the urine NGAL/creatinine ratio and pre-transfusion anemia, iron overload status, and type of iron chelation. Kidney function evaluation should be carried out routinely in adolescent TDT.