

## Renal impairment in $\beta$ thalassemia major patients receiving repeated blood transfusion

Deskripsi Lengkap: <https://lib.ui.ac.id/detail?id=105510&lokasi=lokal>

---

### Abstrak

Thalassemia  $\beta$  mayor adalah penyakit yang disebabkan oleh kelainan sintesis rantai polipeptida  $\beta$  yang diturunkan secara otosomal resesif. Pengobatan thalassemia  $\beta$  mayor pada umumnya berupa pemberian transfusi berulang, yang mengakibatkan penumpukan besi dan berakhir dengan hemokromatosis. Penumpukan besi dapat terjadi pada organ tubuh antara lain ginjal. Tujuan penelitian ini untuk mengetahui adanya gangguan fungsi ginjal pada penderita thalassemia  $\beta$  mayor berumur 15-28 tahun yang telah mendapatkan 6 unit packed red cells. Pada penelitian ini telah diperiksa kadar besi serum (SI) dan daya ikat besi total (TIBC) serta kadar mikroalbumin dan  $\beta_2$ -mikroglobulin ( $\beta_2$ -m) dalam urin. Hasil yang didapat 94,7% penderita menunjukkan peningkatan saturasi transferin dan 40% diantaranya disertai hemokromatosis; 73,4% disertai mikroalbuminuria, 1,3% dengan albuminuria dan 21,3% dengan peningkatan  $\beta_2$ -m urin. Jumlah kasus dengan kelainan ginjal dijumpai pada 78,6%. (Med J Indones 2003; 12: 215-223)

*$\beta$ -thalassemia major is a disease caused by  $\beta$  polypeptide chain synthesis disorder which is inherited in an autosomal recessive manner from both parents and which is marked by little or no  $\beta$ -globin chain synthesis. Treatment for  $\beta$ -thalassemia major patients is by giving repeated blood transfusions, which causes iron accumulation, leading to hemochromatosis. Iron accumulation can occur in various body organ, including the kidneys. The aim of this study was to investigate the existence of renal impairment in  $\beta$ -thalassemia major patients. The subjects of this study were  $\beta$ -thalassemia major patients aged 15 - 28 years old who had received 6 units of packed red cells or more within 6 months. In this study, urine and serum samples of the subjects were taken and examined. Assay of serum iron was performed with Hitachi 737. Results were that 94.7% patients showed an increase in transferrin saturation and 40% of them had hemochromatosis; 73.4% had microalbuminuria; 1.3% had albuminuria and 21.3% had increased urinary  $\beta_2$ -microglobulin ( $\beta_2$ -m). A total of 78.6% of patients showed renal impairment. Conclusion of this study suggested that glomerular dysfunction happens in an earlier stage of the disease process. The high incidence of microalbuminuria is also attributed to defective ability of the proximal tubular cells to reabsorb protein besides dysfunction of the glomeruli. (Med J Indones 2003; 12: 215-223)*