

NRBC in Thalassaemia Syndromes

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In thalassaemia syndromes the suppression of ineffective erythropoiesis is one of the major goals of the therapy, together with the correction of the anaemia and the control of iron overload. The soluble transferrin receptor (s-Tfr) is now the most important parameter for the evaluation of total erythropoiesis. In the last decades the count of the nucleated red blood cells of the peripheral blood was considered a useful indirect parameter for the evaluation of the ineffective erythropoiesis and, therefore, for the definition of a correct transfusion regimen.

The availability of automated NRBC counts allowed us to evaluate the behaviour of this parameter in a large population of thalassaemia patients.

The population study included 32 thalassaemia major patients (14m - 18f), 56 thalassaemia intermedia patients (25m - 31 f), 59 Sß thalassaemia patients (30 m - 29 f) and, in addition, 18 hereditary spherocytosis patients (9 m - 9 f).

Results

As expected, the thalassaemia intermedia patients showed the highest values of s-TfR, while Sß thalassaemia patients and hereditary spherocytosis patients had intermediate values; the lowest values were obtained in thalassaemia major patients with mild differences between splenectomized and no-splenectomized patients of all groups.

NRBC were absent in all hereditary spherocytosis patients.

The counts were always increased in splenectomized patients of the other three groups. The highest values of NRBC were detected in Sß thalassaemia patients; thalassaemia intermedia patients showed intermediate values; the lowest counts were obtained in thalassaemia major patients.

The post- splenectomy increase of NRBC counts could likely be related to the splenic functions. The highest values of NRBC in non- splenectomized Sß thalassaemia patients could be related to the progressive functional asplenia observed in these patients, as suggested by the increased count age related, that whose detected in our patients.

In conclusion, s-TfR appears to be the best index of total erythropoiesis; NRBC could be more sensitive to the amount of the ineffective erythropoiesis: therefore both parameters could be useful to a correct management of transfusion regimen of thalassaemia patients.