Correspondence

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RES Phagocytosis in β -Thalassemia and Sickle Cell Anemia

G.		Drivas
J.		Kafezas
A.		Kalos
G.		Theodoropoulos
K.		Melissinos

Athens

G. Drivas, J. Kafezas, A. Kalos, G. Theodoropoulos, K. Melissinos, Department of Pathologic Physiology, Athens University, Athens-609 (Greece)

Increased susceptibility to infections is thought to be one of the common problems in hereditary hemolytic anemias. The phagocytic capacity of the reticuloendothelial system (RES) is the first system of defense against infections [1]. The clearance of micro-aggregated human serum albumin labeled with radioactive iodine is a way to explore in vivo the RES phagocytosis [2, 3], and we use it in β -thalassemia and sickle cell anemia, both homozygous and heterozygous. The microaggregated iodinated human serum albumin was prepared, using a tracer amount of albumin radioiodinated with radioactive iodine-125 and used at a dose of 5.0 mg/kg [4]. It has a molecular weight of 670,000 daltons. The removal of colloid from plasma by the RES was plotted semilogarithmically with the time and the second exponential component that forms a straight line from the 6th to the 15th min was used to calculate the half-time of clearance by the RES cells. 44 adult patients were compared to 30 adult normal subjects: 14 with homozygous β -thalassemia (TT), 19 with β -thalassemia trait (tt), 12 with sickle cell anemia (SS) and 9 with sickle cell trait (AS). None of these patients have been splenectomized, and at the time of the study they all were in good health and without blood transfusions. The data are shown in table I and figure 1. The clearance of iodinated human serum albumin is normal in TT subjects and diminished in tt, SS and AS subjects. Such results are expected in SS subjects, where RES impairment can be explained by several factors: functional asplenia [7], chronic hemolysis [8], increased susceptibility of cirrhosis which reflects impaired Kupffer cell function [9,10] and opsonin deficiency [11]. A normal clearance of ISA is more surprising in TT subjects. Because factors such as chronic tissue ischemia [5], chronic hemolysis [6] and increased storage of iron in the tissue [7] exist in β -thalassemia we should expect a diminished phagocytosis. Probably, since autopsy studies [8,9] had shown an obvious hyperplasia of the RES in this condition, the above factors are balanced by hepatosplenomegaly. Finally, the decreased clearance of iodinated human serum albumin in tt and AS

Table I. RES clearances (values are mean half-lives $\pm$ SD)

Cases	RES	p	t
n	clearance	values	
min			
Controls	30	13.8 ± 1.9	
β-Thalassemia (TT)	14	15.1 ± 3.7 > 0.1	1.55
β-Thalassemia trait (tt)	19	17.9 ± 3.4 < 0.001	5.0
Sickle cell anemia (SS)	12	20.0 ± 3.4 < 0.001	7.2
Sickle cell trait (AS)	9	17.9 ± 2.3 < 0.001	4.9

subjects needs further studies to elucidate its significance.

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References

- Twomey, J. J.: The macrophage and host defense mechanism. Biomedicine 22: 1-4(1975).
- Benacerraf, B.; Halpern, B.N.; Stiffel, C; et al.: Phagocytose d'une fraction du serum chauffé et iodé par le système réticulo-endothelial system function. J. infect. dis. 123: 465-469(1971).
- 4 Drivas, G.; Uldall, P.R.; Wardle, E.N.: Réticulo-endothelial function in renal allograft recipients. Br.med.J. Hi: 743-745(1975).

Fig. 1. Distribution of RES clearances. Horizontal lines indicate means.

Valdivieso, M.: Bacterial infection in haematological diseases. *Clin. Haematol.* 5: 230 (1976).

Kaye, D.; Gill, F.A.; Hook, E.W.: Factors influencing host resistance to Salmonella infections: the effects of hemolysis and erythrophagocytosis. *Am. J. med. Sci.* 254: 205 (1967).

Dubin, J.N.: Idiopathic hemochromatosis and transfusion siderosis. A review. *Am. J. clin. Path.* 25: 514-542 (1955).

Bhamarapravati, N.; Na-Nakorn, S.; Wasi, P.; Juchinda, S.: Pathology of abnormal hemoglobin diseases seen in Thailand. *Am. J. clin. Path.* 47: 745 (1967).

Green, T.W.; Conley, C.L.H.; Berthrong, M.: The liver in sickle cell anaemia. *Bull. Johns Hopkins Hosp.* 92: 99 (1953).

Yianitsiotis, A.: Liver function in hereditary haemolytic anaemias; MD thesis Athens (1964).

Johnston, R.B.; Newman, S.L.; Struth, A.C.: An abnormality of the alternate pathway of complement activation in sickle cell disease. *New Engl. J. Med.* 288: 803 (1973).