Correspondence

Received: April 4, 1981

Accepted: June 2, 1981

Acta haemat. 1982;67:229

Hemoglobin Koelliker (α_2 minus 141 Arg β_2) in Favism

G.		Schilirò
A.		Russo
N.		Azzia
M.S.		Digiaco
S.		Musumeci
G.		Russo

G. Schilirò, A. Russo, N. Azzia, M.S.S. Digiaco, S. Musumeci, G. Russo, First Pediatric Clinic, University of Catania, Italy

In 1965 Marti [1] first observed a new 'pseu-doabnormal' hemoglobin moving slightly faster than HbA in plasma and urine of patients with acute hemolysis of different etiologies. This hemoglobin, later called Koelliker, results from the action of a plasma carboxypeptidase which is capable of removing the C-terminal 141-arginine of the alpha-chain [2]. A massive hemolysis with saturation of haptoglobin is the condition which allows the enzymatic action on free plasmatic hemoglobin, leading to Hb Koelliker.

Samples of plasma and urine from 10 patients with acute hemolysis of favism were studied by electrophoresis on cellulose acetate in glycine buffer at pH 9. In all cases plasma contained a fast-moving fraction (more than 50% of the normal hemoglobin) which is likely to be Hb Koelliker. The same hemoglobin fraction was found in the urine. Electrophoresis of plasma stored at room temperature for 24 h after mechanical hemolysis showed the same hemoglobin fraction, using either HbA or HbF, HbS and HbG San José. It seems that formation of Hb Koelliker is a general phenomenon occurring during massive hemolysis, when free normal or abnormal hemoglobins are present in plasma.

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