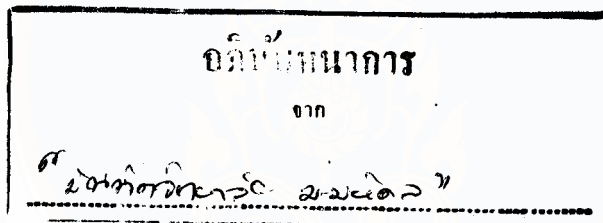


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CALPAIN AND CALPASTATIN IN THALASSEMIC RED CELLS

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ผู้ป่วยจะสูงกว่าในคนปกติประมาณ 2-3 เท่า และจะลดระดับลงจนกระทั่งอยู่ในระดับใกล้เคียงกัน
เคียงกันในชั้นที่มีความหนาแน่นสูงสุด ในขณะที่แอคติวิตีของคาลเพนในชั้นที่มีความ
หนาแน่นต่ำสุดในคนปกติมีค่าสูงกว่าในผู้ป่วยด้วย ดังนั้นจึงสรุปได้ว่าแอคติวิตีของ
โปรตีนเอสไม่ได้มีส่วนสำคัญในการบอกความแตกต่างของระดับความรุนแรงของโรคนี้



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Abstract

In Thailand, β^0 -thalassemia/Hb E is the most common genotype of β -thalassemia disease. However, a wide spectrum of severity is observed, ranging from mild to severe, with a mean hemoglobin level of 7.7 g/dl. One of the contributing factor which may ameliorate the severity is red cell protease. Red cell calpain activity (using casein as substrate) and its inhibitor, calpastatin level (EIA kit) were measured in cytosol and membrane components of 16 normal subjects and 33 patients. Cytosolic calpain activity and calpastatin amount were always higher than in membrane for all cases. Patients (both mild and severe) had higher amounts of calpastatin than normal controls, but calpain activity was not different. However, calpastatin level from patients with mild anemia was lower than that of the severe cases, but no difference in calpain activity could be observed. When erythrocytes (5 normals and 5 patients) were separated into 5 fractions on a Percoll density gradient, calpain activity of normals in the least dense fraction was highest and decrease more than 2-fold in the higher density fraction, but calpastatin level slightly

decreased, whereas in the patients calpain activity stable and calpastatin level decreased slightly. Comparing fractions of similar density, patients calpastatin level in the highest fraction was 2-3 fold higher than normal controls and declined to the same level in the most dense fraction, normal erythrocytes in the highest fraction also contained higher calpain activity than those of patients. It is concluded that red cell protease activity does not play a significant role in determining the severity of β^0 -thalassemia/Hb E disease.

