ELSEVIER

Blood 142 (2023) 5250

The 65th ASH Annual Meeting Abstracts

ONLINE PUBLICATION ONLY

112.THALASSEMIA AND GLOBIN GENE REGULATION

Survival and Causes of Death in Patients with Alpha and Beta-Thalassemia

Adisak Tantiworawit, MD[1,2], Thansita Kamolsripat, MD[3], Pokpong Piriyakhuntorn, MD[4], Thanawat Rattanathammethee, MD[3], Sasinee Hantrakool, MD[3], Chatree Chaiadisaksopha, MD[3], Ekarat Ekarat Rattarittamrong, MD[3], Lalita Norasetthada, MD[3], Kanda Fanhchaksai, PhD[5,2], Pimlak Charoenkwan[5,2]

[1]Division of Hematology, Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai, Thailand
[2]Thalassemia and Hematology Center, Chiang Mai University, Muang Chiang Mai, Thailand
[3]Department of Internal Medicine, Chiang Mai University, Muang, Chiang Mai, Thailand
[4]Division of Hematology, Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai, Thailand
[5]Department of Pediatrics, Chiang Mai University, Muang, Chiang Mai, Thailand

Background: Thalassemia is the most common hereditary anemia worldwide. The prevalence is high in the malaria-endemic areas, including Southeast Asia, and Thailand. Severe thalassemia can result in shortened survival due to disease-related complications. In the last three decades, with regular red cell transfusions and iron chelators, the survivals of patients with thalassemia, especially for TDT, have significantly improved. The most distinctive change was the decrease in the iron-overloaded cardiomyopathy. Both alpha and beta-thalassemia are prevalent in Thailand. We aimed to study the survival, and the causes of death in the patients with thalassemia across the disease severity from NTDT to TDT, and to explore the factors influencing the survival.
Methods: A retrospective review of patients with thalassemia who were followed up at Chiang Mai University Hospital was conducted. The survival status was determined from medical records and the Thai National database (Official statistics registration systems). The survival and clinical factors associated with survival were analyzed.
Results: Our study cohort comprised 789 patients with thalassemia, who were registered in the Departments of Internal Medicine and Pediatrics. The median age (IQR) at the end of follow-up was 18•8 (11•7) years. There were 301 patients (38•1%) with Hb H disease, 279 patients with Hb E/beta-thalassemia (35•4%), and 209 patients (26•5%) with beta-thalassemia major, respectively. Half of the patients (395, 50•1%) had transfusion-dependent thalassemia (TDT). Sixty-five (8•2%) patients had died. The mean age at death was 17•0 ± 0•9 years. Multivariable analysis showed that TDT (adjusted HR 3.68, 95%CI 1.39-9.72, p 0•008) and mean serum ferritin level ≥3,000 ng/mL (adjusted HR 4•18, 95%CI 2.20-7.92, p<0•001) were independently associated with poorer survival. The two major cause of death were infection-related causes (36•9%), cardiac complications (27•7%), thalassemia/anemia (13•8%), trauma (6•2%), and other causes (15•4%).
Conclusions: TDT and mean serum ferritin level ≥3,000 ng/mL were the main factors associated with poorer survival in thalassemia patients. Infections and cardiac complications were the two major causes of death.

Disclosures No relevant conflicts of interest to declare.

https://doi.org/10.1182/blood-2023-179942