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Congenital dyserythropoietic anemia type II revealed in Georgia

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Objective: The congenital dyserythropoietic anemia (CDA) is a rare hereditary disorder, characterized by ineffective erythropoiesis and distinct morphologic abnormalities of erythroblasts in the bone marrow. This study was carried out to investigate infant with CDA type-II and make long-term observat on.

Methods: A full term infant, aged seven month, presented with pallor and jaundice of the skin, moderate splenomegaly and severe anemia was enrolled in the study. Pallor appeared in the five month of life, which was not interpreted correctly. Profound anemia revealed by seven month of age, at the time of admission. Investigations include: detailed history and physical examination, information about used medications, complete blood count with red cell indices, reticulocyte count, iron metabolism, bilirubin, liver and kidney function tests, bone marrow examination, abdominal ultrasound, parvovirus B19 antibodies, measurement of hemoglobin F, folic acid and vitamin B12 levels and acidified serum lyses test. We made packed red cell transfusions each month as a result of profound anemia.

Results: Based on clinical and para clinical data analyses and catamnestic observation, this case was diagnosed with CDA type-II. Diagnostic criteria were: Evidence of congenital chronic anemia with low reticulocyte count for the degree of anemia; increased serum iron and ferritin levels, indirect hyper-bilirubinemia and; typical morphologic abnormalities of the erythroblasts (CDA erythroblasts-30%) in the bone marrow. Acidified serum lyses test was positive.

Conclusion: We had a chance to observe infant with CDA type-II. This rare diagnosis was raised for the first time in Georgia. Anemia was non-responsive to iron, folic acid and vitamin B12. The diagnosis of CDA should be suspected in case of refractory anemia of long duration with a low reticulocyte count for the severity of anemia, features of iron overload, bi-nucleated normoblasts. Early diagnosis of the disease will allow us to prevent iron abundance caused by multiple blood transfusions. Only curative treatment is allogeneic bone marrow transplantation which is currently not possible due to the absence of HLA-compatible donor. The observation on this patient and blood transfusion has been continued per month.

Biography

Zaza Mtvarelidze is a Pediatric Hematologist of Children's New Clinic, Tbilisi, Georgia. For 20 years he has been working on the problems of inherited and acquired anemias in children. He has published 58 articles on this problem in Georgian and European medical Journals. He is the author of two Monographs: Anemia in Children and Iron Deficiency Anemia in Children. In recent years he had become interested in the beta-Thalassemia Major

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