Chirurgia (Bucur). 2017; 112(2):110-116
http://dx.doi.org/10.21614/chirurgia.112.2.110

Hereditary Spherocytosis – Diagnosis, Surgical Treatment and Outcomes. A Literature Review

Simona Manciu[1,2], Emil Matei[1,2], Bogdan Trandafir[2]
[1]Carol Davila University of Medicine and Pharmacy, Bucharest, Romania
[2]Department of General Surgery and Liver Transplantation, Fundeni Clinical Institute, Bucharest, Romania

Abstract
Hereditary spherocytosis (HS) is a disease affecting the red blood cells membrane and belongs to the congenital hemolytic anemias. The clinical spectrum ranges from asymptomatic patients to severe forms requiring transfusions in early childhood. The diagnosis can be based on the physical examination, complete red blood cell count, reticulocytes count, medical history and specific tests, preferentially the EMA test (eosin-5-maleimide binding) test and AGLT (Acidified Glycerol Lysis Time). Splenectomy is considered the standard surgical treatment in moderate and severe forms of hereditary spherocytosis. Total splenectomy exposes the patient to a life - long risk of potentially lethal infections and thus, its usage was reconsidered. Because of this reason, a feasible alternative is the partial splenectomy. The use of partial splenectomy aims to retain splenic immunologic function, while at the same time to decrease the rate of hemolysis. The long - term outcomes of patients with total or subtotal splenectomy for congenital hemolytic anemia, still remain unclear, but the majority of the studies showed a qualitative resolution of anemia and reduction of transfusion rate. Despite the well known advantages of conservative surgery, the optimal choice of treatment and outcomes should be confirmed with the patient.
Key words: spherocytes, hemolysis, flow cytometry, splenectomy, long-term outcomes