

thalassemia major because there is less ineffective erythropoiesis, or none at all.

TABLE 100-2 Features Common to Most Patients with a Hemolytic Disorder

General examination	Jaundice, pallor
Other physical findings	Spleen may be enlarged; bossing of skull in severe congenital cases
Hemoglobin level	From normal to severely reduced
MCV, MCH	Usually increased
Reticulocytes	Usually increased
Bilirubin	Almost always increased (mostly unconjugated)
LDH	Increased (up to 10× normal with intravascular hemolysis)
Haptoglobin	Reduced to absent if hemolysis is at least in part intravascular

Abbreviations: LDH, lactate dehydrogenase; MCH, mean corpuscular hemoglobin; MCV, mean corpuscular volume.

The laboratory features of HA are related to (i) hemolysis per se, and (ii) the erythropoietic response of the bone marrow. In most cases hemolysis is largely extravascular, and it produces an increase in unconjugated bilirubin and aspartate aminotransferase (AST) in the serum; urobilinogen will be increased in both urine and stool. If hemolysis is mainly intravascular, the telltale sign is hemoglobinuria (often associated with hemosiderinuria); in the serum there is free hemoglobin, lactate dehydrogenase (LDH) is increased, and haptoglobin is reduced. In contrast, the serum bilirubin level may be normal or only mildly elevated. The main sign of the erythropoietic response by the bone marrow is an increase in reticulocytes (a test all too often neglected in the initial workup of a patient with anemia). Usually the increase will be reflected in both the percentage of reticulocytes (the more commonly quoted figure) and in the absolute reticulocyte count (the more definitive parameter). The increased number of reticulocytes is associated with an increased mean corpuscular volume (MCV) in the blood count. On the blood smear, this is reflected in the presence of macrocytes;