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HEMATOLOGY BOARD REVIEW MANUAL

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Immune Hemolytic Anemia

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INTRODUCTION

Hemolytic anemias are a diverse group of clinical disorders characterized by decreased survival of erythrocytes in the circulation. Because of their multiple causes, hemolytic anemias are often difficult for hematologists, as well as internists and primary care physicians, to diagnose. This clinical confusion can be lessened somewhat by following a logical, structured approach to diagnosis in patients suspected of having a hemolytic anemia.

First, it must be determined that hemolysis is actually present and that the anemia is not caused instead by bleeding or bone marrow failure. The laboratory and other tests most useful in determining whether or not hemolysis exists are listed in **Table 1**. Next, the most common causes of hemolysis (ie, acquired autoimmune hemolytic anemia, hypersplenism, congenital hemoglobinopathies) should be ruled in or out, with the specific cause identified, if possible. If a common cause is not easily ascertained, a “checklist” approach to the diagnosis of hemolytic anemia can be pursued. The numerous specific causes of hemolysis for the most part fall into 1 of 4 general categories: (1) processes outside the erythrocyte, (2) alterations of the erythrocytic membrane, (3) abnormalities in the hemoglobin molecule, or (4) decreased levels of an enzyme in erythrocytes (**Table 2**).¹ When evidence of one of the disorders that commonly cause hemolytic anemia cannot be found, this checklist is useful as a guide to other problems to consider in making the diagnosis.

In discussing hemolytic anemias, this review will focus specifically on autoimmune hemolytic anemia (AIHA), reviewing its definition, classifications, etiology, pathophysiology, clinical features, diagnosis, and management. The case of a 34-year-old man with a hemolytic anemia and spherocytosis will be presented to illustrate the major clinical points.

CLINICAL PRESENTATION

INITIAL EVALUATION OF CASE PATIENT

A 34-year-old man is seen in the emergency department reporting shortness of breath with exertion. He says that he has been unable to climb the stairs to his third-floor apartment during the past 2 weeks without stopping several times; previously, he could climb to the third floor very easily. Medical history is unremarkable except for arthroscopic knee surgery performed 2 years ago following a skiing injury. He takes no medicine other than an occasional nonsteroidal anti-inflammatory drug to treat headaches and knee pain. Family history reveals that his mother was treated for breast cancer 6 years ago; his father and 2 siblings are in good health. The patient does not smoke or use illicit drugs but does drink 2 to 3 beers each week.

Physical examination reveals a pale man in no distress at rest. Blood pressure is 140/76 mm Hg, pulse is 124 bpm, and respiratory rate is 22 breaths/min. He is afebrile. Nail beds and conjunctivae are pale; the sclerae