due to a contaminated blood unit. Rarely, and if at all in children, HA is seen with sepsis or endocarditis from a variety of organisms. In addition, bacterial and viral infections can cause HA by indirect mechanisms (see Table 100-6).

Immune Hemolytic Anemias These can arise through at least two distinct mechanisms. First, when an antibody directed against a certain molecule (e.g., a drug) reacts with that molecule, red cells may get caught in the reaction (the so-called innocent bystander mechanism: see section below on Hemolytic Anemia from Toxic Agents and Drugs), whereby they are damaged or destroyed. Second, and more frequently, a true autoantibody is directed against a red cell antigen, i.e., a molecule present on the surface of red cells. Autoimmune hemolytic anemias have been originally classified into two types, depending on the thermal amplitude of the autoantibodies involved: this classification is valid, because the two types have different pathophysiological and clinical features.

AUTOIMMUNE HEMOLYTIC ANEMIA, warm type (wAIHA: for simplicity we will use the acronym AIHA) This type has an estimated incidence in the United States of about 1–3:100,000 per year, and a prevalence of 17:100,000. AIHA can be serious since even with appropriate management the mortality is of the order of 5–10%.

Clinical Features and Diagnosis The onset is often abrupt and can be dramatic. The hemoglobin level may drop, within days, to as low as 4 g/dL; the massive red cell removal will produce jaundice, and sometimes the spleen is enlarged. When this triad is present, the suspicion of AIHA must be high. The reticulocyte count is typically elevated, except when erythroid precursors are also targeted by the autoantibody attack. LDH may also be elevated. In some cases, AIHA can be associated, on first presentation or subsequently, with autoimmune thrombocytopenia. This double autoimmune condition, referred to as Evans syndrome, may be a manifestation of common variable immune deficiency, and in children it may suggest one of several primary immune deficiency syndromes. Evans syndrome signals high-risk disease. Other predictors of the outcome and of the probability of relapse of AIHA are severe anemia (Hb <6 g/dL),