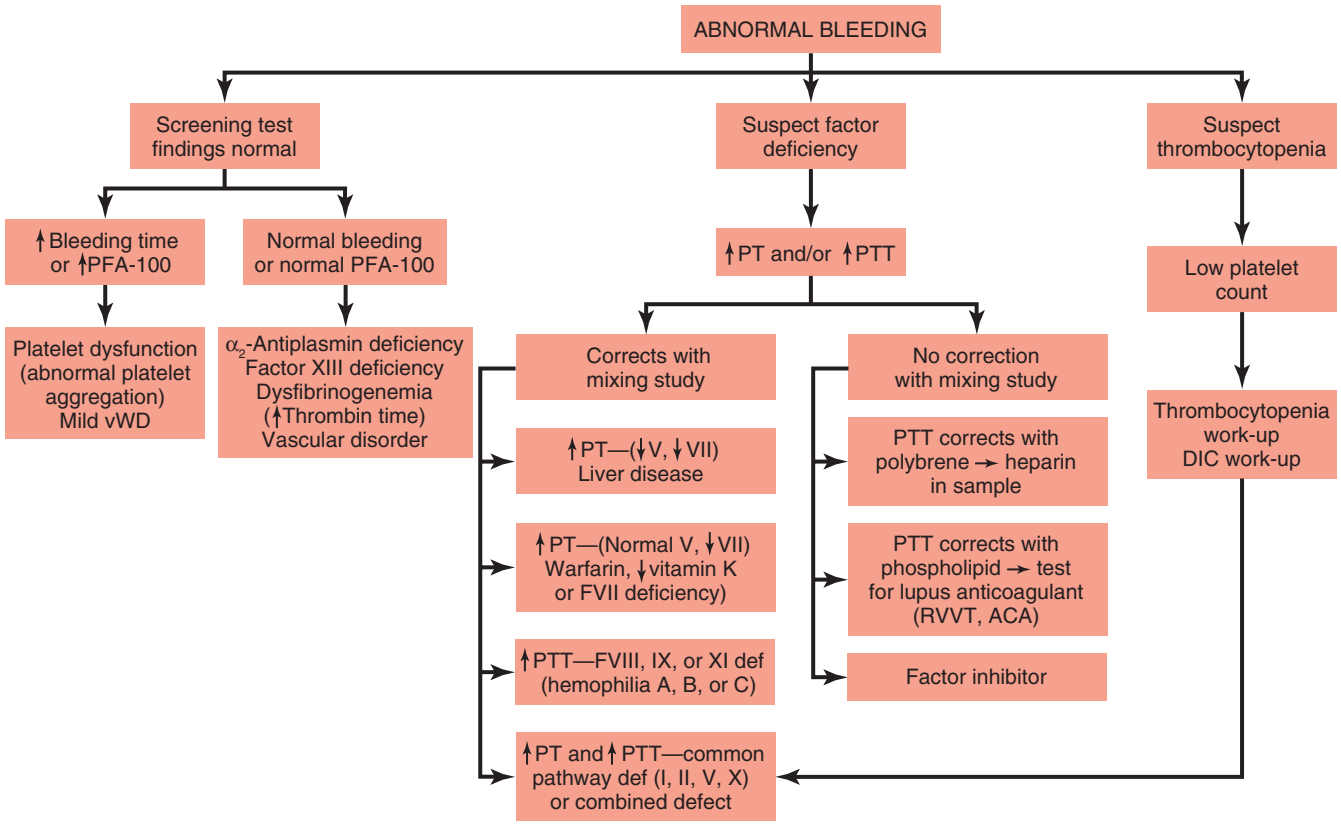

FIGURE 51-3 Algorithm for the evaluation of bleeding. Screening laboratory tests for platelet and factor deficiencies are used to narrow the work-up for bleeding, followed by specific factor and other coagulation studies (e.g., mixing studies, D-dimer) to confirm the diagnosis. ACA, Anticardiolipin antibody; DIC, disseminated intravascular coagulation; FVIII, factor VIII; PFA-100, Platelet Function Analyzer-100; PT, prothrombin time; PTT, partial thromboplastin time; RVVT, Russell viper venom time; vWD, von Willebrand disease; ↑, increased; ↓, decreased.

thrombocytopenia, or abnormal platelet function; (2) low levels of multiple coagulation factors resulting from vitamin K deficiency, liver disease, or disseminated intravascular coagulation (DIC); (3) single-factor deficiency (usually inherited); and, more rarely, (4) an acquired inhibitor to a coagulation factor such as factor VIII. The laboratory evaluation is most efficient when it is performed in this context.

BLEEDING CAUSED BY VASCULAR DISORDERS

Vascular purpura (i.e., bruising) is defined as bleeding caused by intrinsic structural abnormalities of blood vessels or by inflammatory infiltration of blood vessels (i.e., vasculitis). Although vascular purpura usually causes bleeding in the setting of normal platelet counts and normal coagulation study results, vasculitis and vessel damage may be severe enough to cause secondary consumption of platelets and coagulation factors.

Collagen breakdown and thinning of the subcutaneous tissue that overlies blood vessels is often observed in older patients (i.e., senile purpura), and similar atrophic skin changes are a common effect of steroid therapy. Another acquired cause of vascular purpura is scurvy (i.e., defieicney of vitamin C [ascorbic acid]). Patients with scurvy have bleeding around individual hair fibers (i.e., perifollicular hemorrhage) and corkscrew-shaped hairs. Bruising occurs in a classic saddle pattern over the upper thighs. The bleeding gums are caused by gingivitis and not by the subcutaneous tissue defect. Edentulous patients with scurvy do not have bleeding gums, and scurvy should not be excluded on this basis.

Congenital defects of the vessel wall can cause bruising. These rare syndromes include pseudoxanthoma elasticum, a defect of the elastic fibers of the vasculature that is associated with severe GI and genitourinary bleeding, and Ehlers-Danlos syndrome, which is characterized by abnormal collagen molecules in blood vessels and subcutaneous tissue. Both syndromes cause bruising in the skin, but only patients with pseudoxanthoma elasticum develop significant GI bleeding.

Another inherited vessel wall defect associated with GI bleeding is hereditary hemorrhagic telangiectasia (i.e., Osler-Weber-Rendu syndrome). This disorder is characterized by degeneration of the blood vessel wall that results in angiomatous lesions resembling blood blisters on mucous membranes, including the lips and GI tract. The frequency of bleeding caused by breakdown of these lesions increases with age, and GI lesions commonly cause significant chronic bleeding, often resulting in iron deficiency.

The sudden onset of palpable purpura (i.e., localized, raised hemorrhages in the skin) associated with rash and fever may be caused by aseptic or septic vasculitis. Septic vasculitis can be caused by meningococcemia and other bacterial infections and is often accompanied by thrombocytopenia and prolongation of clotting times. One cause of aseptic vasculitis in young children and adolescents is Henoch-Schönlein purpura, a vasculitis of the skin, GI tract, and kidneys that is usually accompanied by