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Patient with bleeding diathesis in the emergency room: principles of management

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Summary

The study aim was to develop an algorithm supporting decision-making at the Hospital Emergency Department or the Admission Room for the management of unexpected bleeding suggestive of a bleeding disorder.

Key words: bleeding disorder, acquired hemophilia A (AHA), bleeding, Hospital Emergency Department (HED)

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Bleeding disorders are conditions characterized by defects in hemostasis that lead to increased susceptibility to bleeding. Pursuant to the defective element of the hemostatic mechanism, bleeding disorders may be classified into vascular, platelet, coagulation factor deficiencies, fibrinolytic defects as well as complex (more than one defective hemostatic element). Pursuant to the cause of coagulopathy, bleeding disorders fall into two main categories: congenital (genetically determined) and acquired. Typically, the latter presents suddenly, in individuals with no personal or family history of bleeding and may prove a diagnostic and therapeutic challenge, particularly if the acquired bleeding disorder is severe. One such example is acquired hemophilia A (AHA) which may occur in both men and women and may not be concomitant to any clinical condition or comorbidity. AHA is caused by autoantibodies directed against factor VIII and is therefore classified as an autoimmune disorder. AHA is most common in the elderly; the estimated median age at AHA onset is 74. Typical clinical symptoms of AHA are extensive subcutaneous hemorrhages, intramuscular hematomas, and excessive mucosal bleeding. The most characteristic laboratory AHA feature is prolongation of activated partial thromboplastin time (APTT) by usually more than 10 seconds with other hemostatic screening tests being normal (hence APTT prolongation in AHA is referred to as "isolated"). Recommendations for the management of patients with AHA are presented in separate guidelines.

The first step in approaching a profusely bleeding person at the Hospital Emergency Department or the Emergency Room, is to assess the type and severity of bleeding and promptly take the medical history. Even at this early stage, it is sometimes possible to precisely plan further management; particularly if the patient had already been diagnosed with a congenital bleeding disorder. Patients with hemophilia and related bleeding disorders often hold special identity cards with relevant information on management of their particular bleeding disorder. It is equally important to determine if the patient receives anticoagulants, the overdose of which may be the cause of excessive bleeding.

No matter what, hemostasis screening tests should always be performed. These include APTT, prothrombin time (PT), thrombin time (TT), fibrinogen levels and platelet count. APTT prolon-

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gation (by > 8 s) requires the APTT mixing test (patient's plasma mixed with an equal volume of pooled normal plasma containing all the clotting factors). The test is available in all laboratories of hemostasis. Failure to correct prolonged APTT, may indicate a circulating anticoagulant (inhibitor) in the patient's plasma. Most common are factor VIII inhibitors.

The outcome of hemostasis screening tests may also help to determine the cause of bleeding in patients who are on anticoagulant therapy. For example, if a person overdosed on vitamin K antagonists (VKA) (e.g., acenocoumarol or warfarin), his international normalized ratio (INR) derived from PT may markedly exceed the indicated therapeutic dose. For direct oral anticoagulants (DOAC), the results of screening tests are no reliable source of precise information on drug overdose. If an overdose on DOAC is suspected, appropriate tests are ordered to determine the concentration of the drug (rivaroxaban, apixaban or dabigatran) in plasma.

Patients with congenital bleeding disorders, as well as those diagnosed with AHA, find professional care at Hemophilia Treatment Centers (HTC) usually well equipped, with appropriate laboratory facilities and personnel experienced in the treatment of bleeding disorders (both congenital and acquired). As already mentioned, patients diagnosed with hemophilia and related bleeding disorders hold special ID card which — apart from general information on therapy — include contact details of the HTC.

It is noteworthy that clotting factor concentrates are provided free of charge by the Regional Blood Transfusion Centers to patients with congenital bleeding disorders and AHA as part of the National Program for the Treatment of Patients with Hemophilia and Related Hemorrhagic Diseases for 2019–2023.

Figure 1 presents the approach to patients with profuse bleeding suggestive of a hemorrhagic disorder. Scanning the QR code gives access to guidelines for the management of AHA in elderly patients.

Conflict of interest: none declared

References

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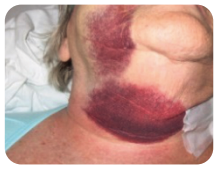
Algorithm for management of patients suspected of bleeding disorders — Hospital Emergency Department (HED)



When to suspect a bleeding disorder at the Hospital Emergency Department?

Major bleeding

- Gastrointestinal bleeding (coffee-ground emesis, tarry stools)
- Subcutaneous and/or intramuscular hematomas
- Hematuria
- Heavy vaginal bleeding
- Excessive bleeding inadequate to injury

Interview!

- Diagnosed/congenital hemostatic disorders
- Drug intake, including anticoagulant and antiplatelet drugs
- Recent invasive procedure or trauma/injury
- Severe concomittant diseases

When bleeding is life-threatening*

- **Discontinue anticoagulants!**
- **Collect blood samples for** coagulation tests
- **Avoid any non-life saving invasive procedures,** that may intensify bleeding
- **Administer RBCc and/or hemostatic drugs,** eg. coagulation factor concentrates#

- **Screening tests of hemostasis****
  - Platelet count
  - Coagulation tests: APTT, PT, TT and fibrinogen concentration
  - APTT mixing test with normal plasma (if APTT prolonged by > 8 sec.)
  - Factor VIII activity+

Anticoagulation drugs

Yes → **Determine the type and dose of medication**

No data → Confirm with specific laboratory tests

No → **Abnormal screeing tests*****

Congenital bleeding disorder

No → **Abnormal screeing tests*****

Yes → **Management according to diagnosis**
- Consult HTC$
- Verify data from patient's ID
- Order coagulation factors in regional blood center#

Abnormal screeing tests***

Yes → **Refer to a hospital ward!**
- **Consult with** HTC$
- **Differentiate bleeding disorders based on** further laboratory assays
- **Start anti-hemorrhagic treatment** eg. coagulation factor concentrates#

No → **Differentiate !**
- **Bleeding unrelated to coagulation disorders** (following injury, invasive procedure)
- **Previously unrecognized congenital bleeding disorders** with no impact on coagulation screening tests (eg. trombocytopathies, factor XIII and other factor deficiencies).

Guidelines for the management of acquired hemophilia A in elderly patients

*Life-threatening bleeds due to massive blood loss or locus (e.g. intracranial, neck)
**Interpretation of laboratory tests should include: type of anticoagulant, discontinuation date and hemostatic drugs
***Deviations may be related to conditions other than bleeding disorder, e.g. factor XII (Hageman) deficiency, high-molecular-weight kininogen or prekallikrein deficiency, lupus anticoagulant
+Common acquired clotting factor deficiency
#Drugs available at the regional blood establishments (BEs) within the National Program for the Treatment of Patients with Hemophilia and Related Hemorrhagic Diseases for 2019–2023 (listed in QRCode)
$HTC — Hemophilia Treatment Centers (listed in the National Program for the Treatment of Patients with Hemophilia and Related Hemorrhagic Diseases at gov.pl)
AHA — acquired hemophilia A; APTT — activated partial thromboplastin time, PT — prothrombin time; TT — thrombin time; RBCC — red blood cell concentrate/packed red blood cells

Figure 1.