Recommendation 7.3.1:

- **In hemophilia patients presenting with suspected central nervous system bleeds or bleed-related symptoms, clotting factor replacement therapy should be administered immediately before investigations are performed.** CB

- Immediately administer appropriate clotting factor replacement therapy as soon as significant trauma or symptoms occur, before any other intervention, and maintain factor level until etiology is defined. If a bleed is confirmed, maintain the appropriate factor level for 10-14 days.[22,23] (See Table 7-2.)
- Immediate medical evaluation and hospitalization are required, including a computed tomography (CT) scan or magnetic resonance imaging (MRI) of the brain and neurological consultation as soon as possible.[24,25] Ultrasound examination may be considered in children.

Recommendation 7.3.2:

- **In patients with hemophilia presenting with suspected central nervous system bleeding that could be life-threatening, clotting factor replacement therapy should be administered immediately before investigations are performed and continued until the bleed resolves.**
- **Remark: In patients with hemophilia who have been treated for central nervous system bleeding, secondary prophylaxis is recommended to prevent bleed recurrence.** CB

- Intracranial hemorrhage may be an indication for secondary prophylaxis (short-term prophylaxis for 3-6 months or even lifelong), especially where a relatively high risk of bleed recurrence has been observed (e.g., in the presence of human immunodeficiency virus [HIV] infection).[22,26,27]

7.4 | Throat and neck hemorrhage

- Bleeding into the throat or neck may be due to local pathology, trauma, or severe coughing, and may present with swelling or pain. This is a medical emergency because it can lead to airway obstruction. If indicated, gently elevate the head to help reduce airway obstruction due to the hemorrhage.
- Treat immediately with CFC to raise the patient's factor level when significant trauma or bleeding symptoms occur in the throat and neck area, without any delay that could occur while awaiting full evaluation. (See Table 7-2.)
- Immediate hospitalization and medical evaluation by a specialist otolaryngologist is required.[28]
- Protective factor levels should be maintained until symptoms resolve.[28-30] (See Table 7-2.)

Recommendation 7.4.1:

- **In hemophilia patients with throat and neck bleeding, clotting factor replacement therapy should be administered immediately and critical care evaluation sought.** CB

Recommendation 7.4.2:

- **In hemophilia patients with throat and neck bleeding, including injury of the tongue, clotting factor replacement therapy should continue until the bleeding symptoms have resolved.** CB

- To prevent oral hemorrhage in patients with severe tonsillitis, prophylaxis with CFCs, desmopressin (DDAVP; for those with mild or moderate hemophilia A), or antifibrinolytics (epsilon aminocaproic acid [EACA] and tranexamic acid) are advised in addition to bacterial culture and treatment with appropriate antibiotics.

Recommendation 7.4.3:

- **In hemophilia patients with throat and neck bleeding and local infection, antifibrinolytics should be started to treat the bleed and antibiotics to treat the infection.** CB

7.5 | Gastrointestinal/abdominal hemorrhage

- Acute gastrointestinal (GI) hemorrhage may present as hematemesis, hematochezia (rectal passage of fresh blood), or melena.
- In a patient with liver disease, the first sign of GI bleeding may be hepatic encephalopathy, as the failing liver cannot process the high protein load of GI bleeding.
- Any sign of GI bleeding and/or acute hemorrhage in the abdomen requires immediate medical evaluation. All patients with GI bleeds should be hospitalized.

Recommendation 7.5.1:

- **In hemophilia patients with gastrointestinal bleeding, factor levels should be raised immediately and the underlying etiology of the bleed identified and treated.** CB

- GI bleeds must be treated as soon as possible following injury and/or the onset of the earliest symptoms with clotting factor replacement therapy to raise the patient's factor level, with factor levels maintained until hemorrhaging has stopped and the etiology of the hemorrhage is defined.[31,32] (See Table 7-2.)

Recommendation 7.5.2:

- **Hemophilia patients with gastrointestinal bleeding should be prescribed antifibrinolytics.** CB

- Antifibrinolytics are often effective adjunctive therapy for both patients with hemophilia A and hemophilia B. Concurrent use with aPCC or prothrombin complex concentrate (PCC) may be used with caution in some patients.
- Treat the origin of the hemorrhage as indicated.
- Monitor hemoglobin levels regularly and treat anemia or shock as needed. Perform endoscopy, if clinically indicated, in any patient with dropping hemoglobin levels. In GI bleeding, the investigation of choice is endoscopy.