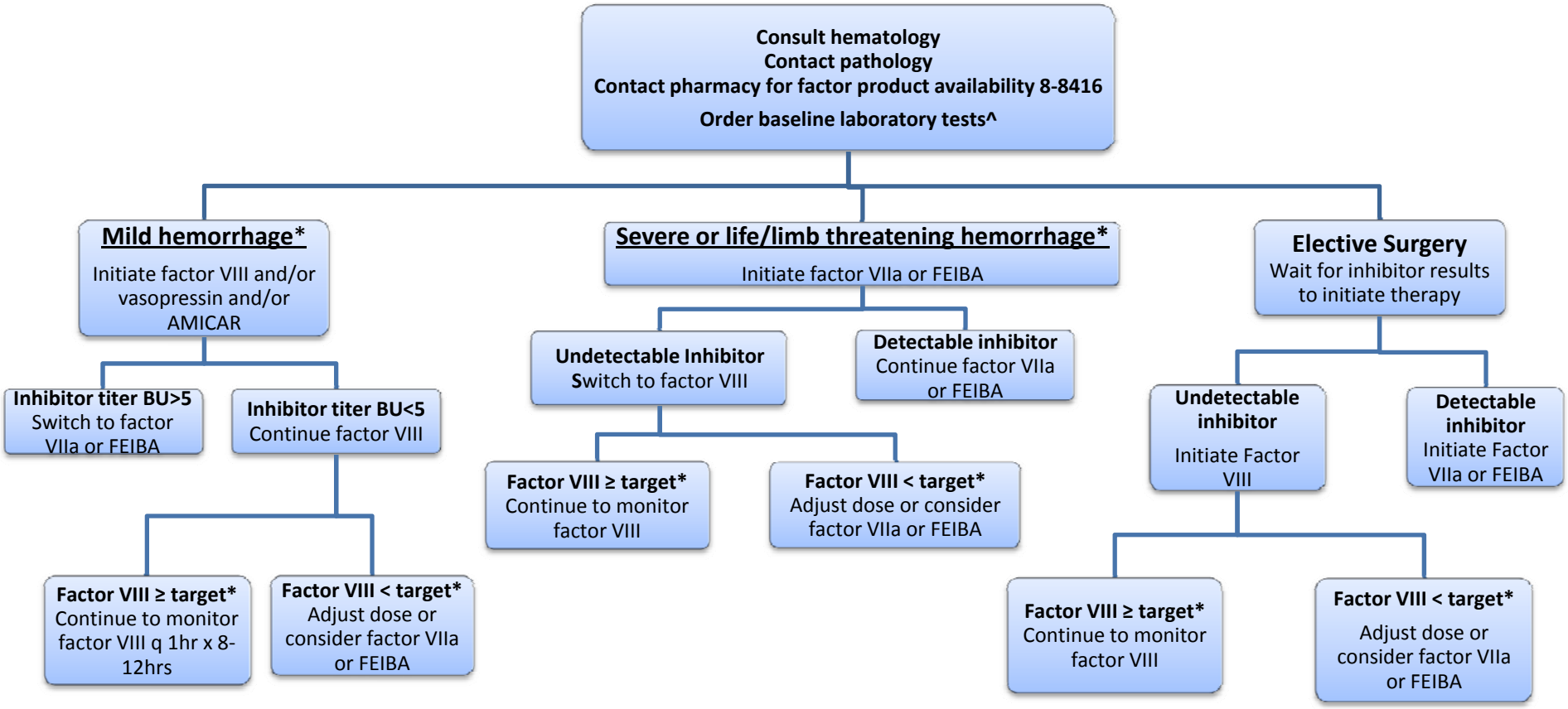
Algorithm for the Management of Bleeding in Hemophilia

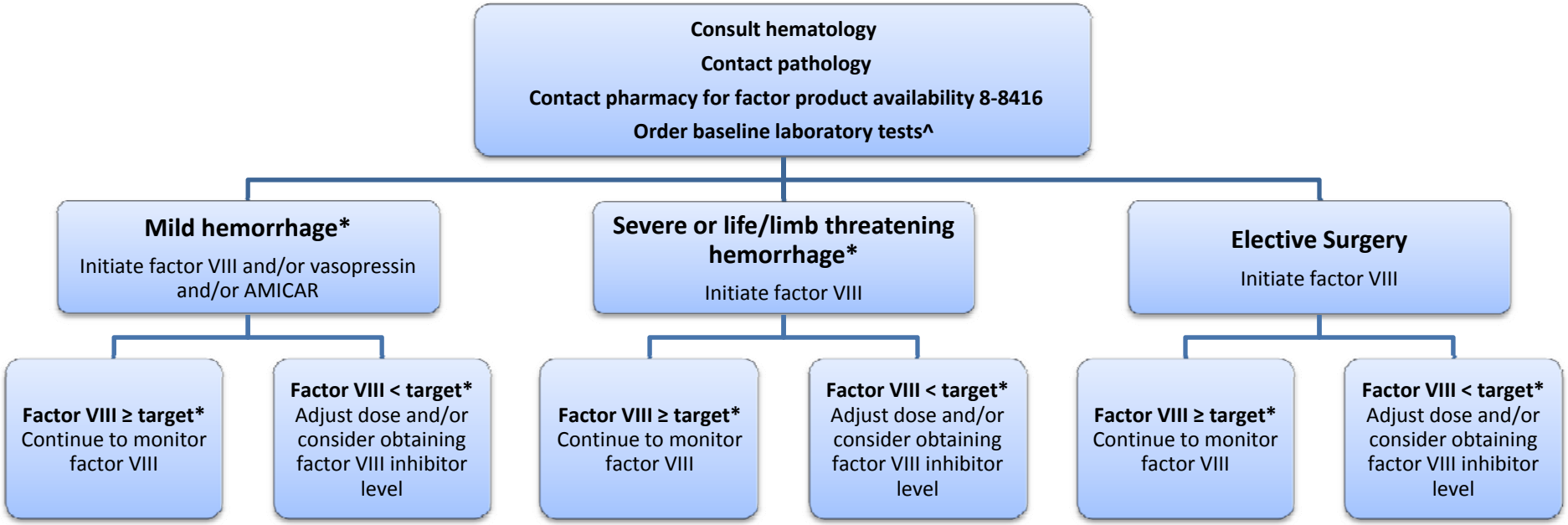
Hemophilia A with History of Inhibitor



^Initial laboratory tests include aPTT, factor VIII activity, and factor VIII inhibitor titer

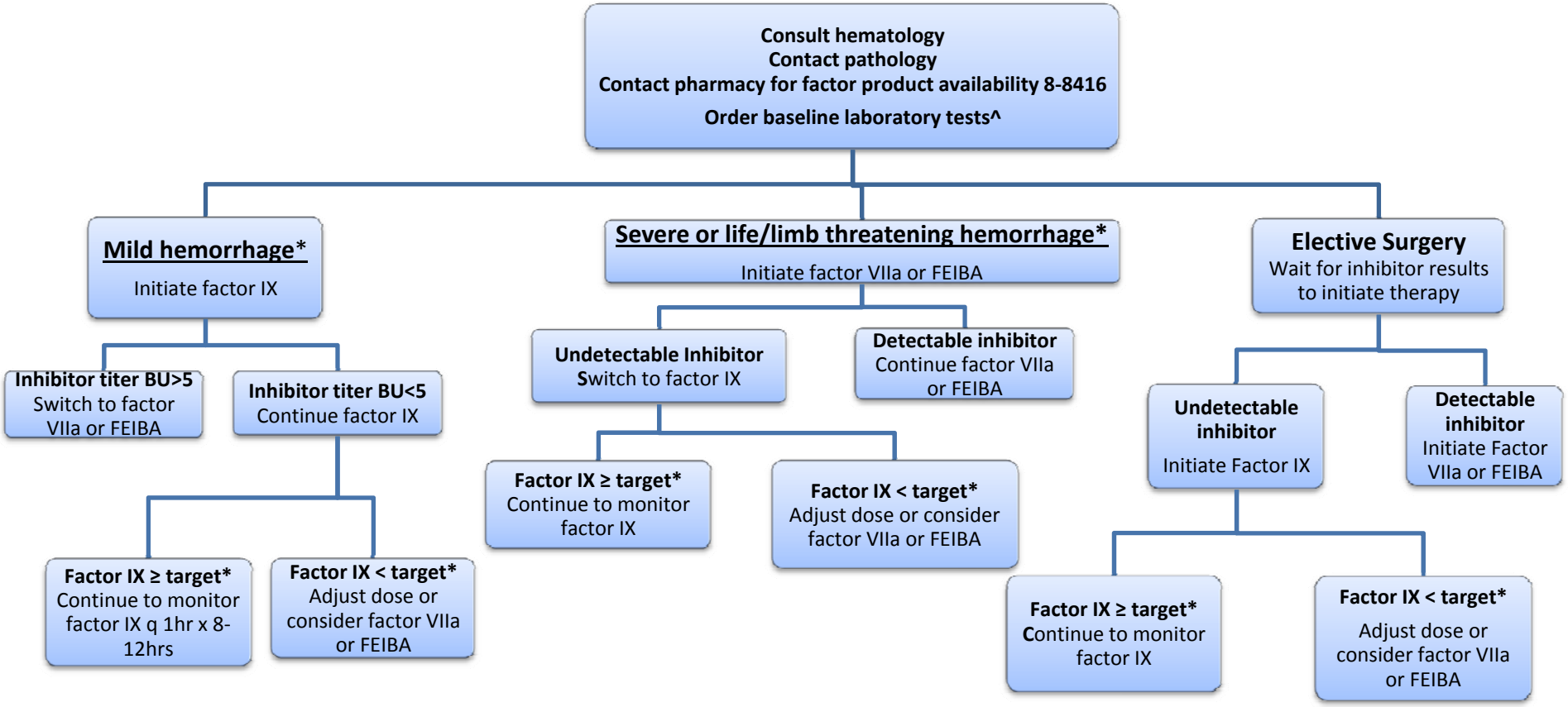
*See Table 1 to determine severity of bleed, appropriate factor VIII target levels, and Table 2 for suggested dosages

Hemophilia A without History of Inhibitor



^Initial laboratory tests include aPTT, factor VIII activity, and aPTT mixing studies
*See Table 1 to determine severity of bleed, appropriate factor VIII target levels, and Table 2 for suggested dosages

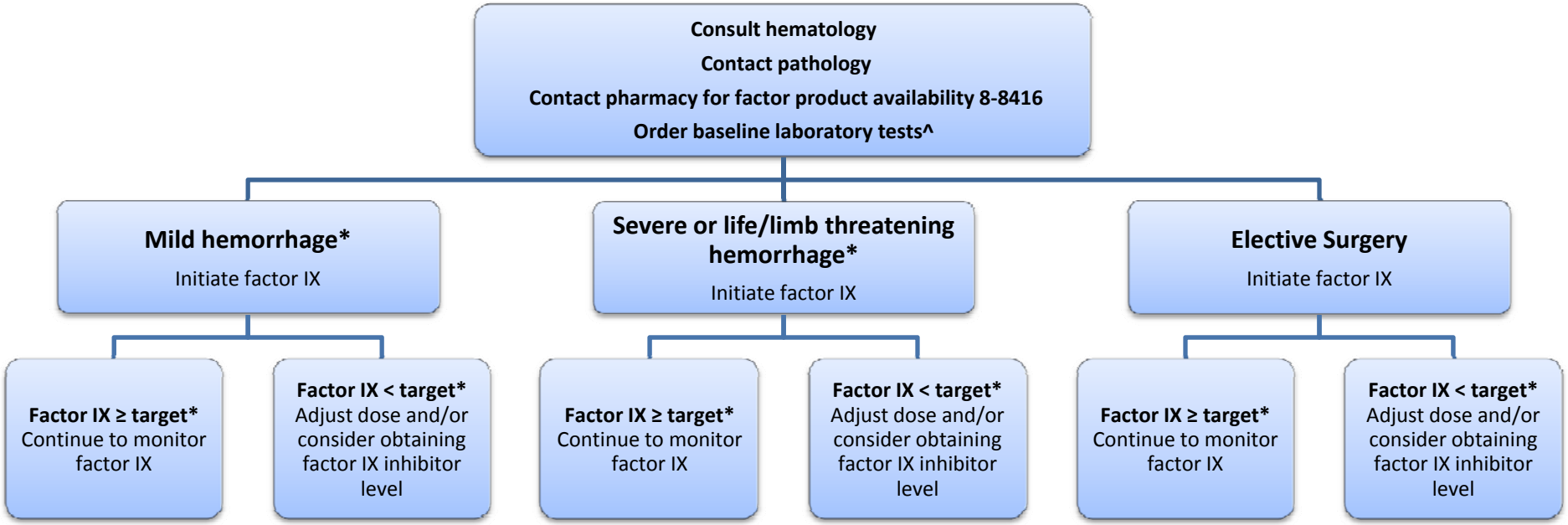
Hemophilia B with History of Inhibitor



^Initial laboratory tests include aPTT, factor IX activity, and factor IX inhibitor titer

*See Table 1 to determine severity of bleed, appropriate factor IX target levels, and Table 2 for suggested dosages

Hemophilia B <u>without</u> History of Inhibitor



^Initial laboratory tests include aPTT, factor VIII activity, and aPTT mixing studies
*See Table 1 to determine severity of bleed, appropriate factor VIII target levels, and Table 2 for suggested dosages

Table 1: Target Factor Level and Duration of Administration

Hemorrhage Type	Hemophilia A		Hemophilia B	
	Desired Level	Duration (days)	Desired Level	Duration (days)
Joint	40-60%	1-2; sometimes longer if response is inadequate	40-60%	1-2; sometimes longer if response is inadequate
Muscle (except iliopsoas)	40-60%	2-3; sometimes longer if response is inadequate	40-60%	2-3; sometimes longer if response is inadequate
Iliopsoas -Initial -Maintenance	 80-100% 30-60%	 1-2 3-5 (sometimes longer as secondary prophylaxis during physiotherapy)	 60-80% 30-60%	 1-2 3-5 (sometimes longer as secondary prophylaxis during physiotherapy)
CNS/Head -Initial -Maintenance	 80-100% 50%	 1-7 8-21	 60-80% 30%	 1-7 8-21
Throat and neck -Initial -Maintenance	 80-100% 50%	 1-7 8-14	 60-80% 30%	 1-7 8-14
Gastrointestinal -Initial -Maintenance	 80-100% 50%	 1-6 7-14	 60-80% 30%	 1-6 7-14
Renal	50%	3-5	40%	3-5
Deep laceration	50%	5-7	40%	5-7
Surgery (major) -Pre-op -Post-op	 80-100% 60-80% 40-60% 30-50%	 1-3 4-6 7-14		

Adapted from: Guidelines for the Management of Hemophilia. World Federation of Hemophilia 2005

Table 2: Product Information

Product	Dose	Comments
Vasopressin (DDAVP)	IV: 0.3mcg/kg diluted in 50-100 mL NS infused over 20-30 minutes -Peak response is seen 90 minutes after infusion	-Boosts plasma levels of FVIII by 3-6 fold -Does not effect FIX levels, so not effective in hemophilia B
Aminocaproic Acid (AMICAR)	5g in 250mL NS infused over 1 hours followed by 1g every hour x 8 hrs or until bleeding stops	-Should not be used in combination with FIX products
Factor VIII	**Factor VIII Dose (int. units) =** **weight (kg) x (desired factor % increase) x (0.5 int. unit/kg**) -Administer IV over 5-10 minutes -May repeat dose every 6-24 hours according to bleed severity and continued factor activity monitoring -Administration by continuous infusion has been studied	-See table 1 for desired factor activity %
Factor IX	**Factor IX Dose (int. units) =** **weight (kg) x (desired factor % increase) x (1 int. unit/kg**) -Administer IV at rate not to exceed 10 mL/minute -May repeat dose every 12-24 hours according to continued factor activity monitoring -Administration by continuous infusion has been studied	-See table 1 for desired factor activity %
Factor VIIa	90mcg/kg IV every 2-3 hours. Successful control of bleeding has been reported using doses that range between 35-120 mcg/kg. -Treatment can be tapered by increasing the time interval between doses -Administration by continuous infusion has been studied	-Bypassing agent -Reserved for patients with inhibitors to factors VIII or IX
FEIBA	**Joint or mucous membrane bleed:** Initial 50 units/kg every 6-12 hrs (May increase to 100 units/kg if hemorrhage does not stop) **Soft Tissue Hemorrhage:** 100 units/kg every 12 hrs **Other Severe Hemorrhage:** 100 units/kg every 6-12 hours Due to the risk of thromboembolic events, single doses > 100 units/kg and daily dose of > 200 units/kg should not be exceeded unless absolutely necessary to stop bleeding.	-Bypassing agent -Reserved for patients with inhibitors to factors VIII or IX -Made from human plasma

References

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