

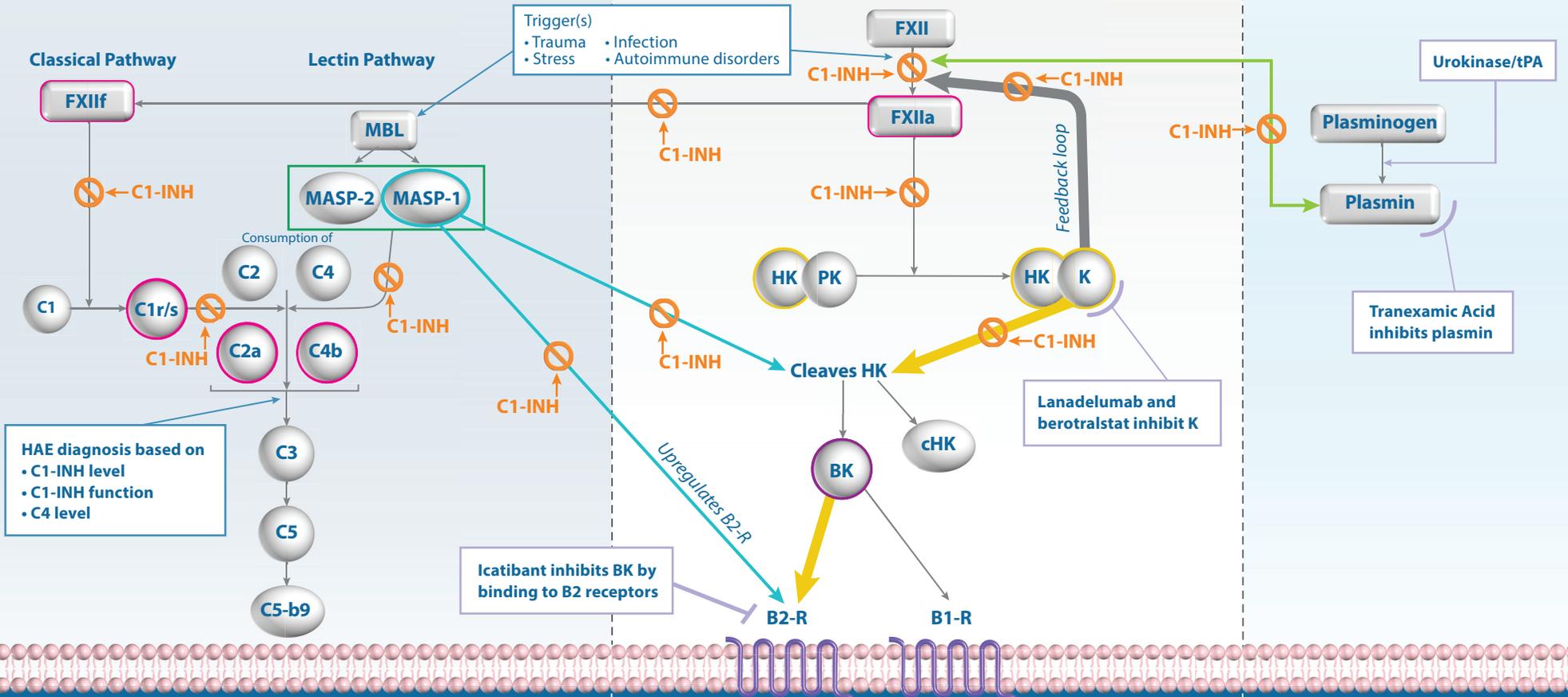
The Importance of Comprehensive C1-Inhibition for HAE¹⁻⁹

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Complement System^{1,4-6,10,11}

Contact Activation System^{1,6,12-14}

Fibrinolysis System^{7,15,16}



- Participates in the elimination of invading microorganisms¹⁷
- Low C1-INH levels fail to block MASP-1 from cleaving HK, which may augment Bradykinin production¹⁸
- Low levels of C1-INH lead to activation and consumption of C4 and C2 during acute HAE attacks¹⁹

Vascular Leakage (Edema)

- Represents a group of plasma proteins that promote inflammation upon activation²⁰
- Low C1-INH levels fail to block FXIIa and Kallikrein, leading to an increase in bradykinin production¹⁹

- Regulates the dissolution of clots as wounds heal by degrading fibrin, the netting that clots blood²¹
- Plasmin activates FXII to FXIIa (and FXIIIf)⁷

LEGEND

C1-Inhibitor

Coagulation factors

Activated coagulation factors

Prekallikrein

Cleaved HK (inactive)

Bradykinin

Complement

Activated complement

Kallikrein

High molecular weight kininogen

Mannan-binding lectin

Mannose-binding lectin associated serine proteases

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