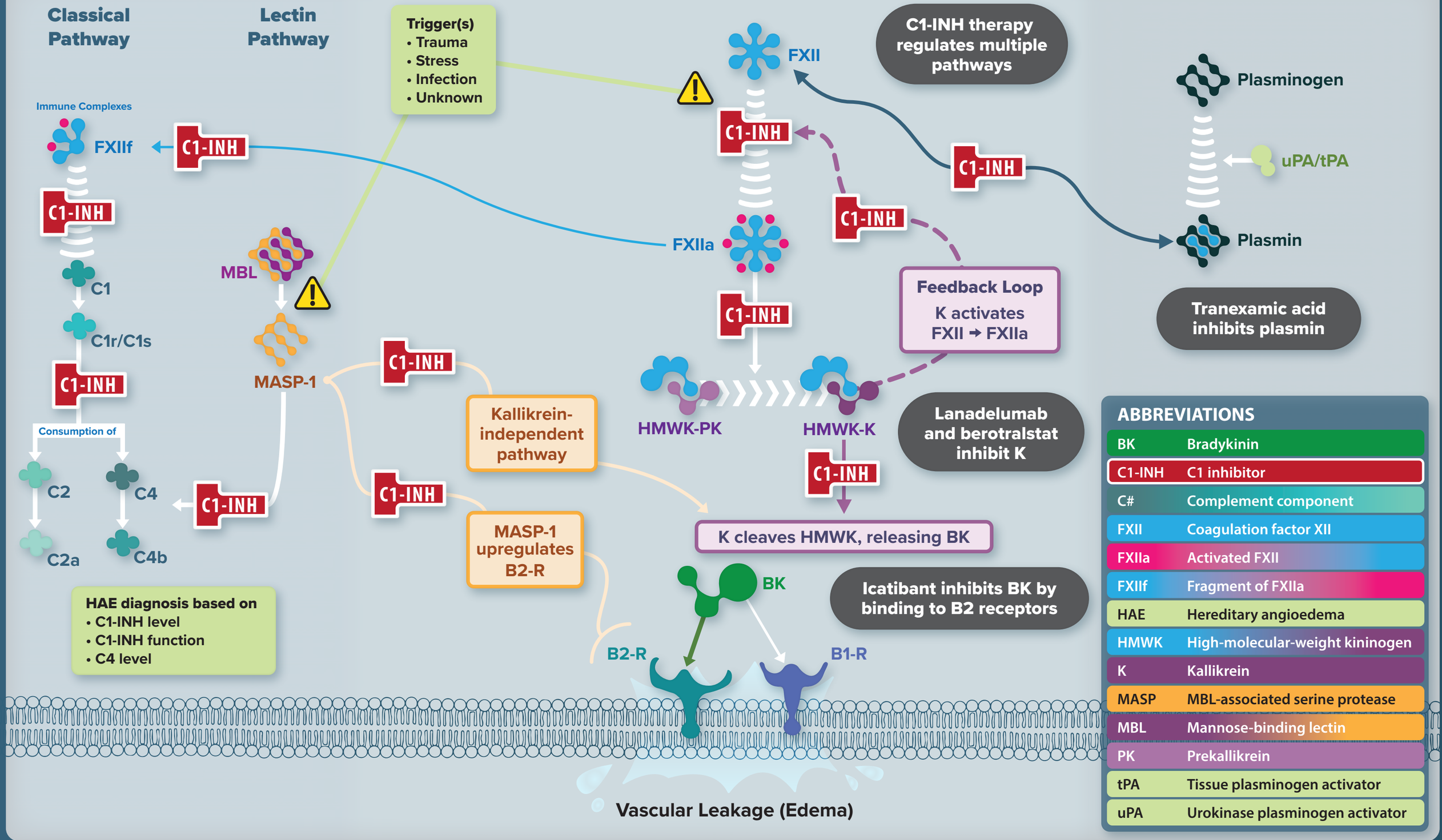
COMPLEMENT SYSTEM[1,4-6,10,11]

CONTACT ACTIVATION SYSTEM[1,6,12-14]

FIBRINOLYSIS SYSTEM[7,15,16]



ABBREVIATIONS

BK	Bradykinin
C1-INH	C1 inhibitor
C#	Complement component
FXII	Coagulation factor XII
FXIIa	Activated FXII
FXIIf	Fragment of FXIIa
HAE	Hereditary angioedema
HMWK	High-molecular-weight kininogen
K	Kallikrein
MASP	MBL-associated serine protease
MBL	Mannose-binding lectin
PK	Prekallikrein
tPA	Tissue plasminogen activator
uPA	Urokinase plasminogen activator

Complement System

- Participates in the elimination of invading microorganisms[17]
- Low C1-INH levels fail to block MASP-1 from cleaving HMWK, which may augment bradykinin production[18]
- Low levels of C1-INH lead to activation and consumption of C4 and C2 during acute HAE attacks[19]

Contact Activation System

- Represents a group of plasma proteins that promote inflammation upon activation[20]
- Low C1-INH levels fail to block FXIIa and Kallikrein, leading to an increase in bradykinin production[19]

Fibrinolysis System

- Regulates the dissolution of clots as wounds heal by degrading fibrin, the netting that clots blood[21]
- Plasmin activates FXII to FXIIa (and FXIIf)[7]

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Pharming

The Importance of C1-INH for HAE[1-9]

Created in partnership with Dr. Allen Kaplan