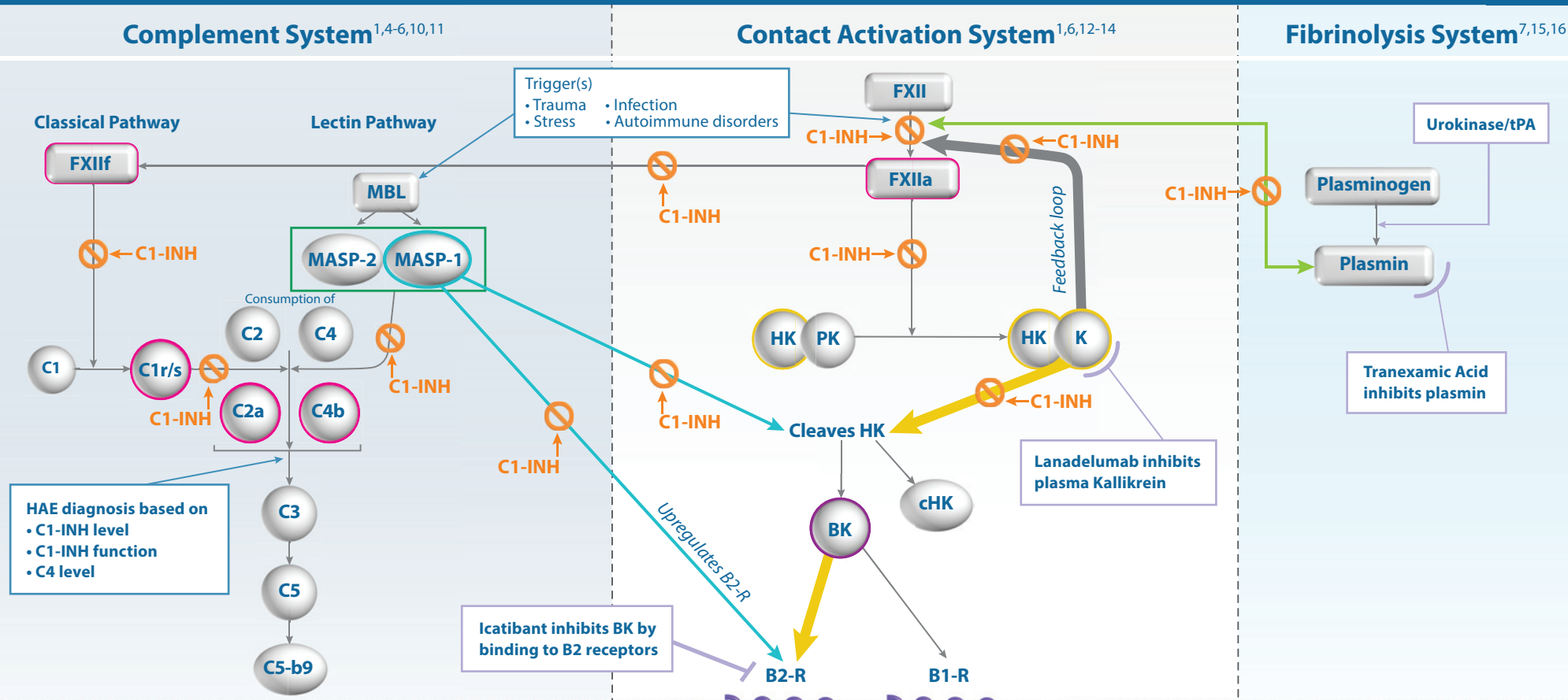
The Importance of Comprehensive C1-Inhibition for HAE[1-9]

Developed in partnership with Dr. Allen Kaplan



Vascular Leakage (Edema)

- Responsible for the elimination of invading microorganisms[17]
- Low C1-INH levels fail to block MASP-1 from cleaving HK, which may augment Bradykinin production[18]
- Low levels of C1-INH leads to activation and consumption of C4 and, during attacks of angioedema, activation and consumption of C2.[19]

- 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]

- 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]

LEGEND

- C1-Inhibitor
- FXII — Coagulation factors
- FXIIa/f — Activated coagulation factors
- PK — Prekallikrein
- cHK — Cleaved HK (inactive)
- BK — Bradykinin
- C# — Complement
- C# — Activated complement
- K — Kallikrein
- HK — High molecular weight kininogen
- MBL — Mannan-binding lectin
- MASPs — Mannose-binding lectin associated serine proteases

This is a current scientific understanding of the cascades. Clinical implications are unknown.

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OTH-US-2019-0021
Printed in USA.

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