

Hematologické imitátory sepsy

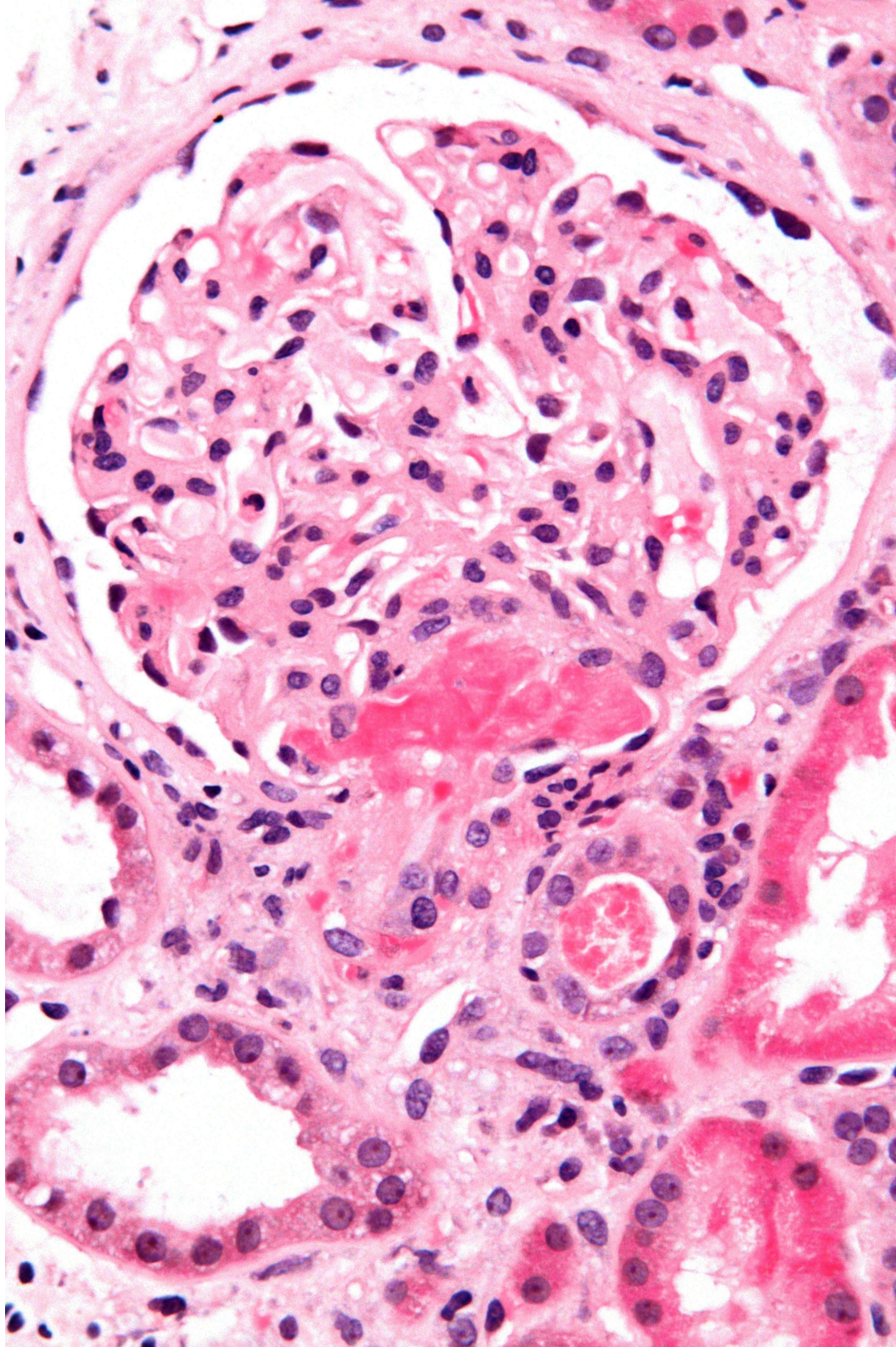
Jan Vydra

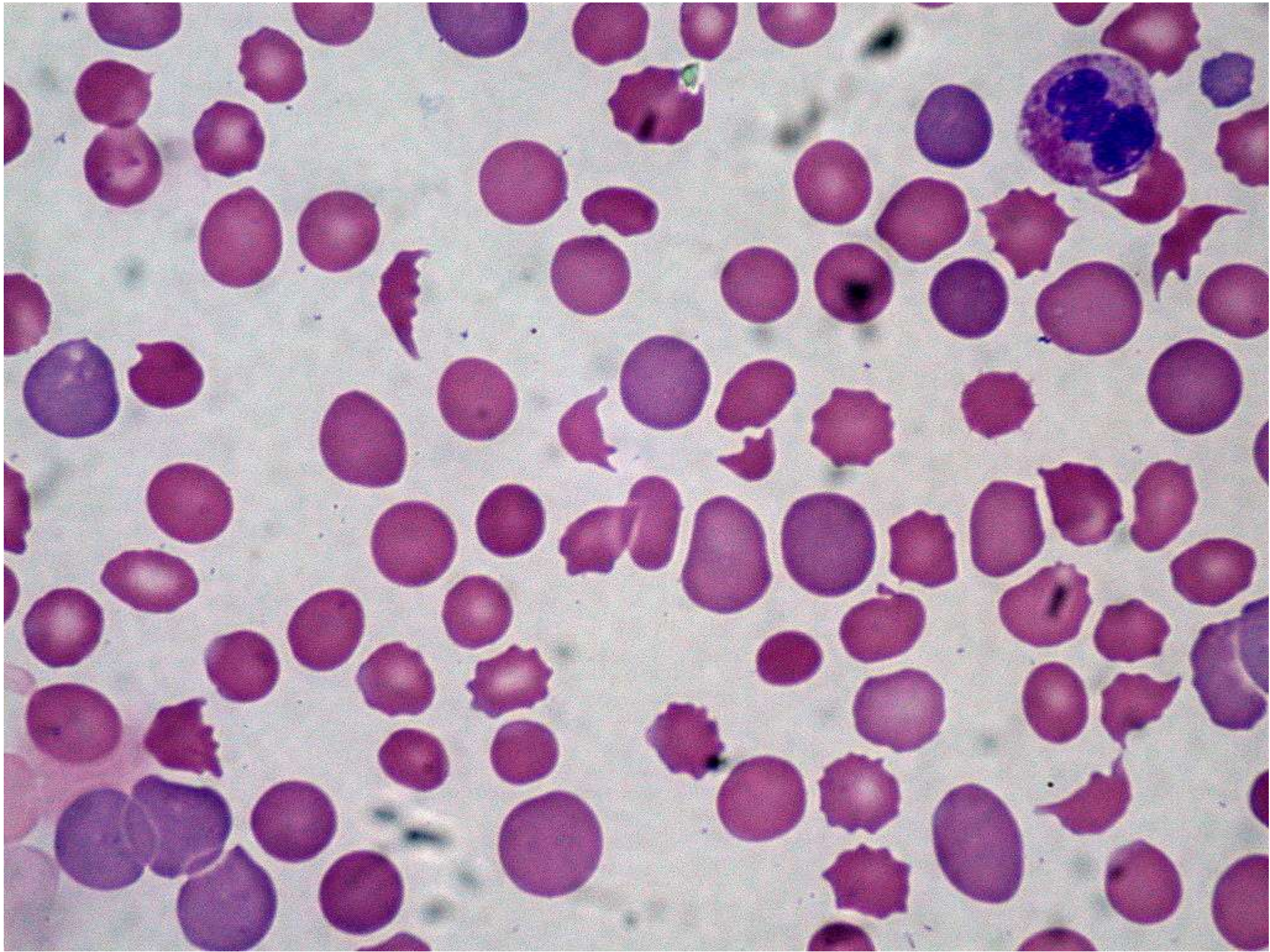
Ústav hematologie a krevní transfuze

Praha

Hematologická onemocnění imitující některé znaky sepse

- Trombotické mikroangiopatie
- Hemofagocytující lymfohistiocytóza
- Akutní leukémie
- Tumor lysis syndrome
- "Cytokine release syndrome"

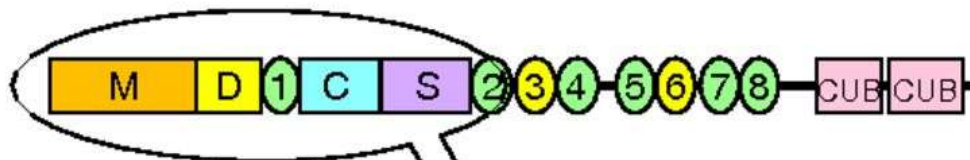




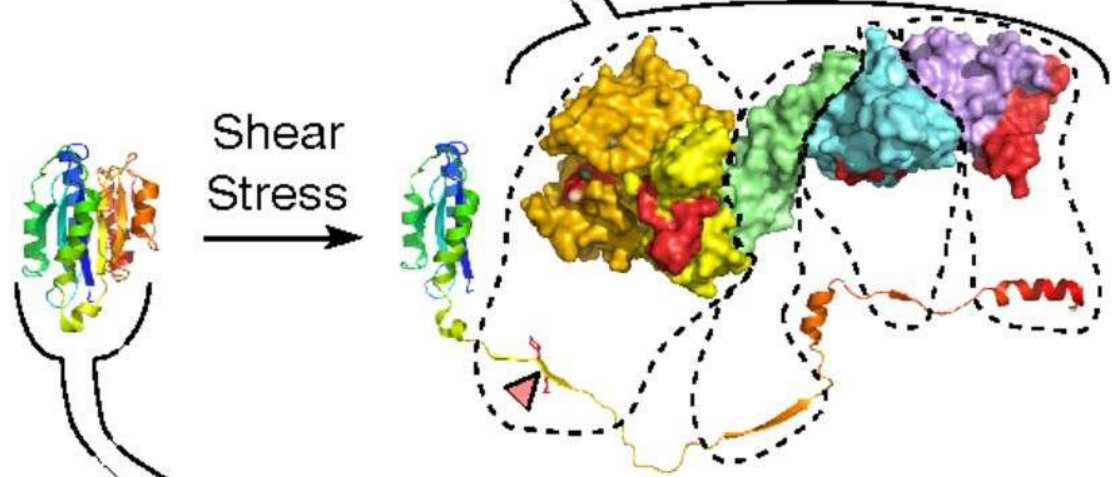
Trombotické mikroangiopatie

- Onemocnění, u kterých dochází k obstrukci mikrocirkulace a současně k projevům mikroangopatické hemolýzy
- Primární TMA syndromy
 - TTP
 - atypický hemolyticko uremický syndrom - komplementem podmíněné TMA, hereditární a získané
 - těhotenstvím indukovaný aHUS
 - shiga toxin HUS
 - Pneumokokový HUS
 - polékové TMA
 - hereditární poruchy metabolismu B12, plasminogenu a další
- Systémová onemocnění s MAHA a trombocytopenií
 - preeklampsie / HELLP syndrom
 - hypertenzní krize
 - systémové infekce, DIC
 - diseminované malignity
 - autoimunity - SLE
 - komplikace transplantací (HSCT nebo SOT)

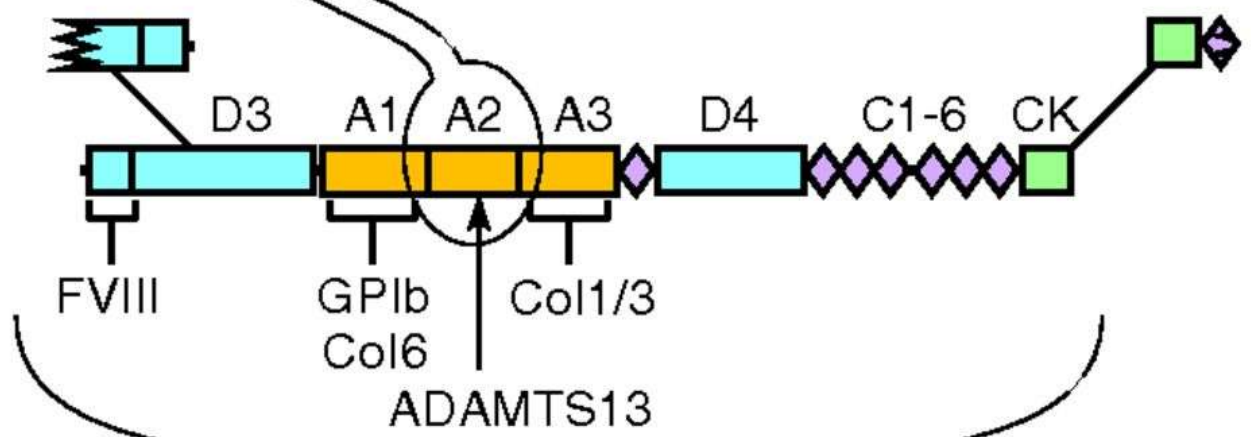
ADAMTS13



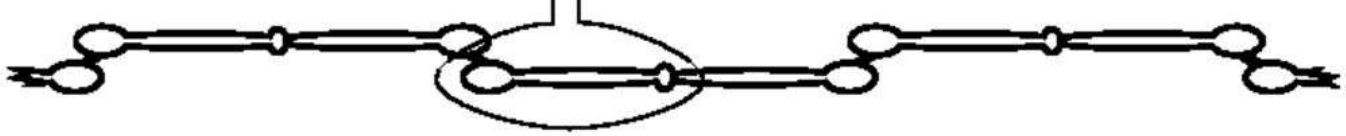
Domain A2

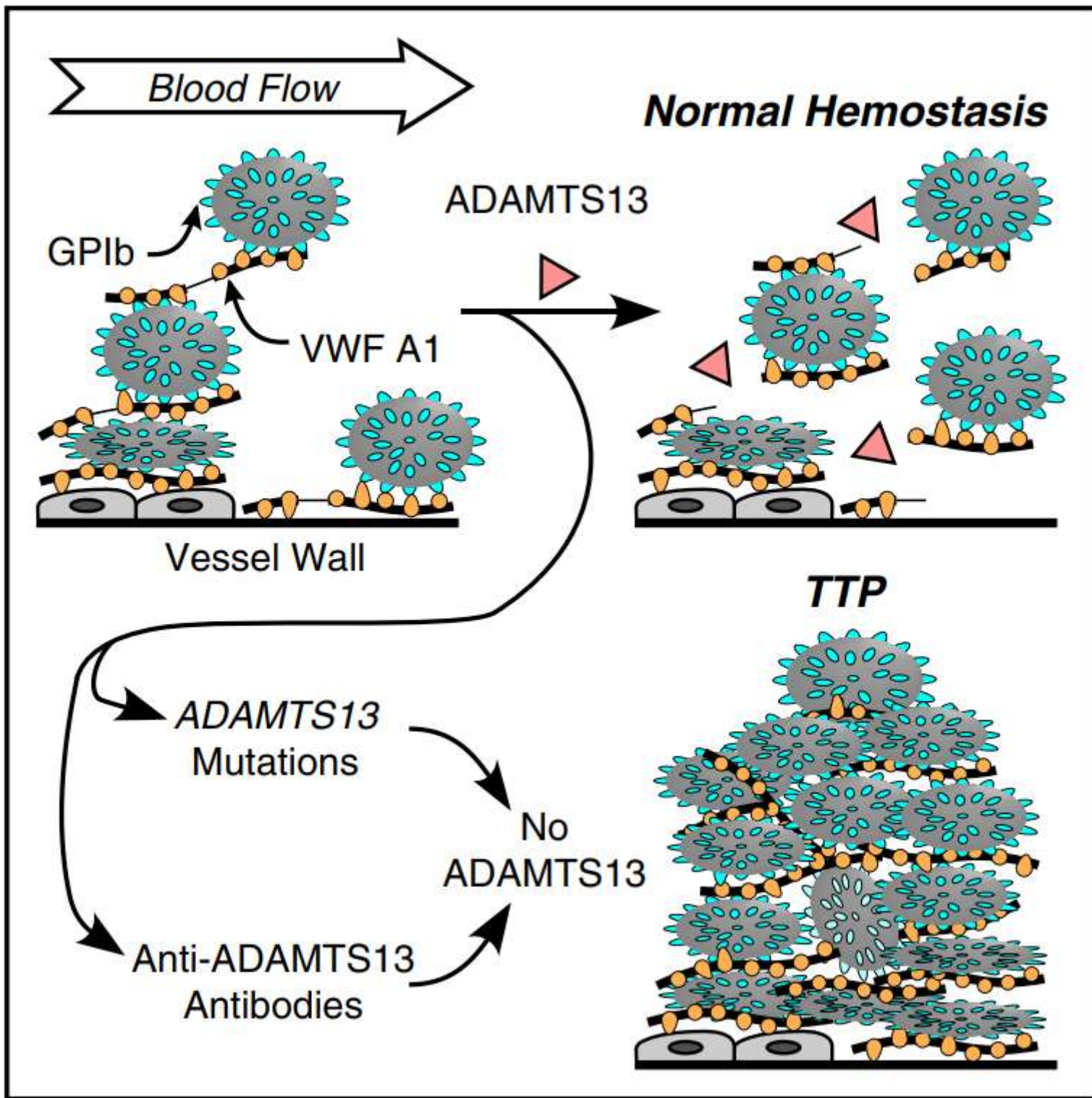


Subunit

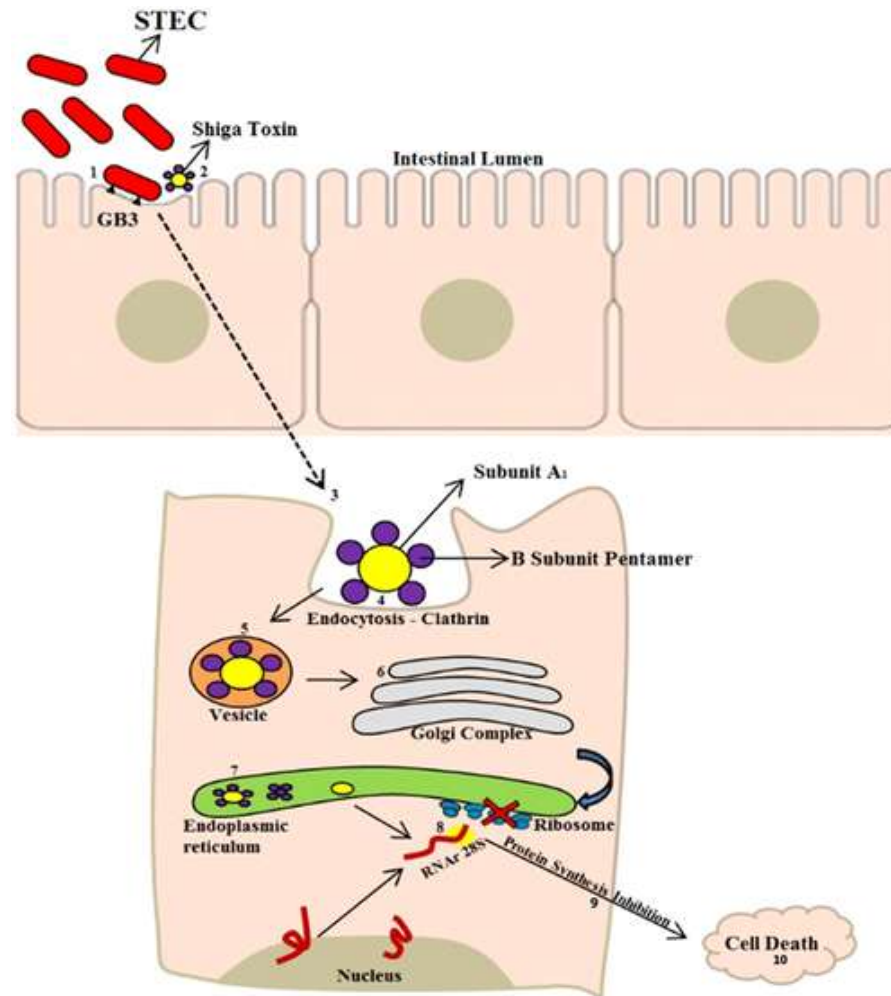


VWF Multimer

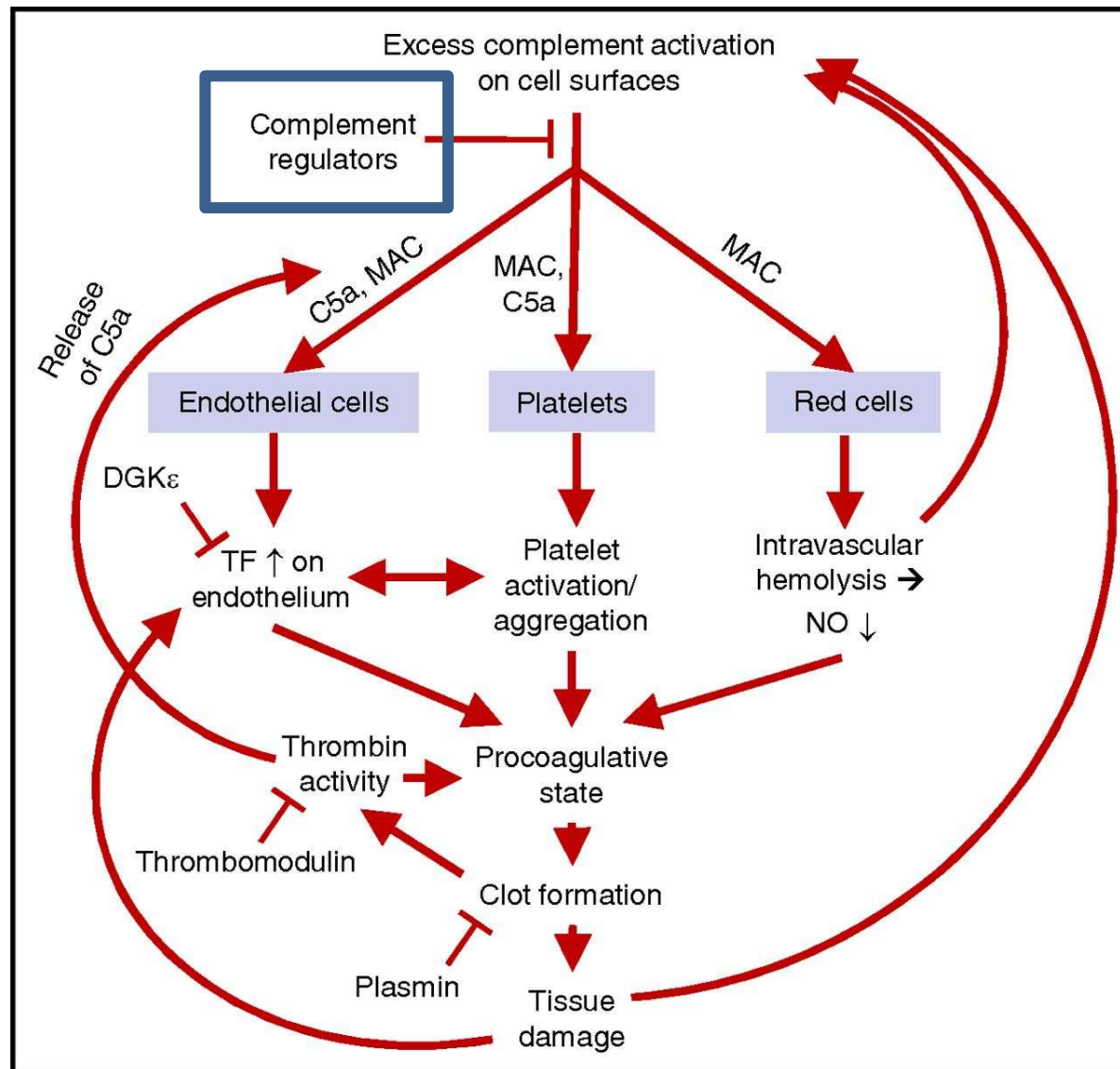




Shiga-toxin Producing *Escherichia coli*: Pathogenicity, Supershedding, Diagnostic Methods, Occurrence, and Foodborne Outbreaks

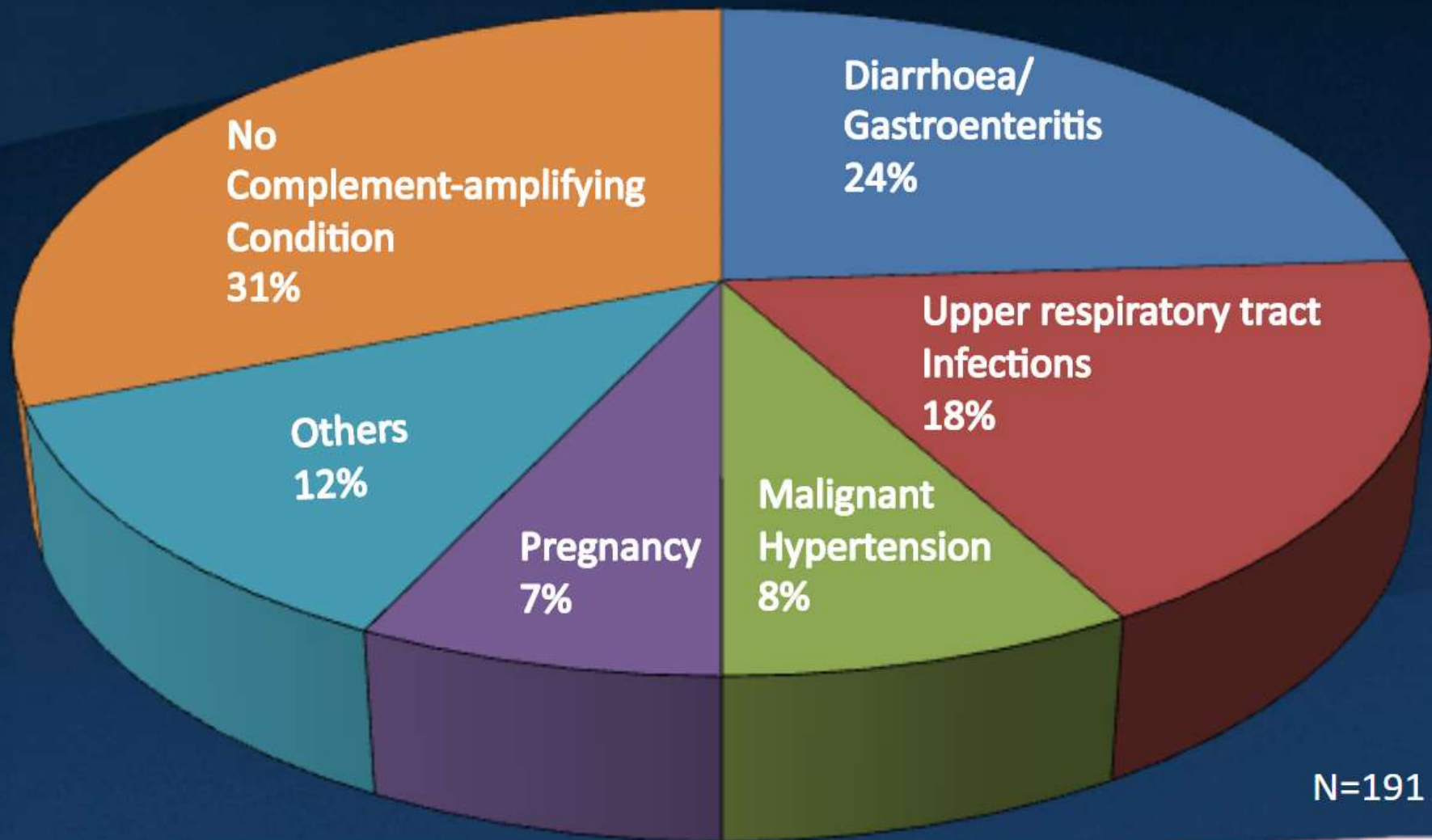


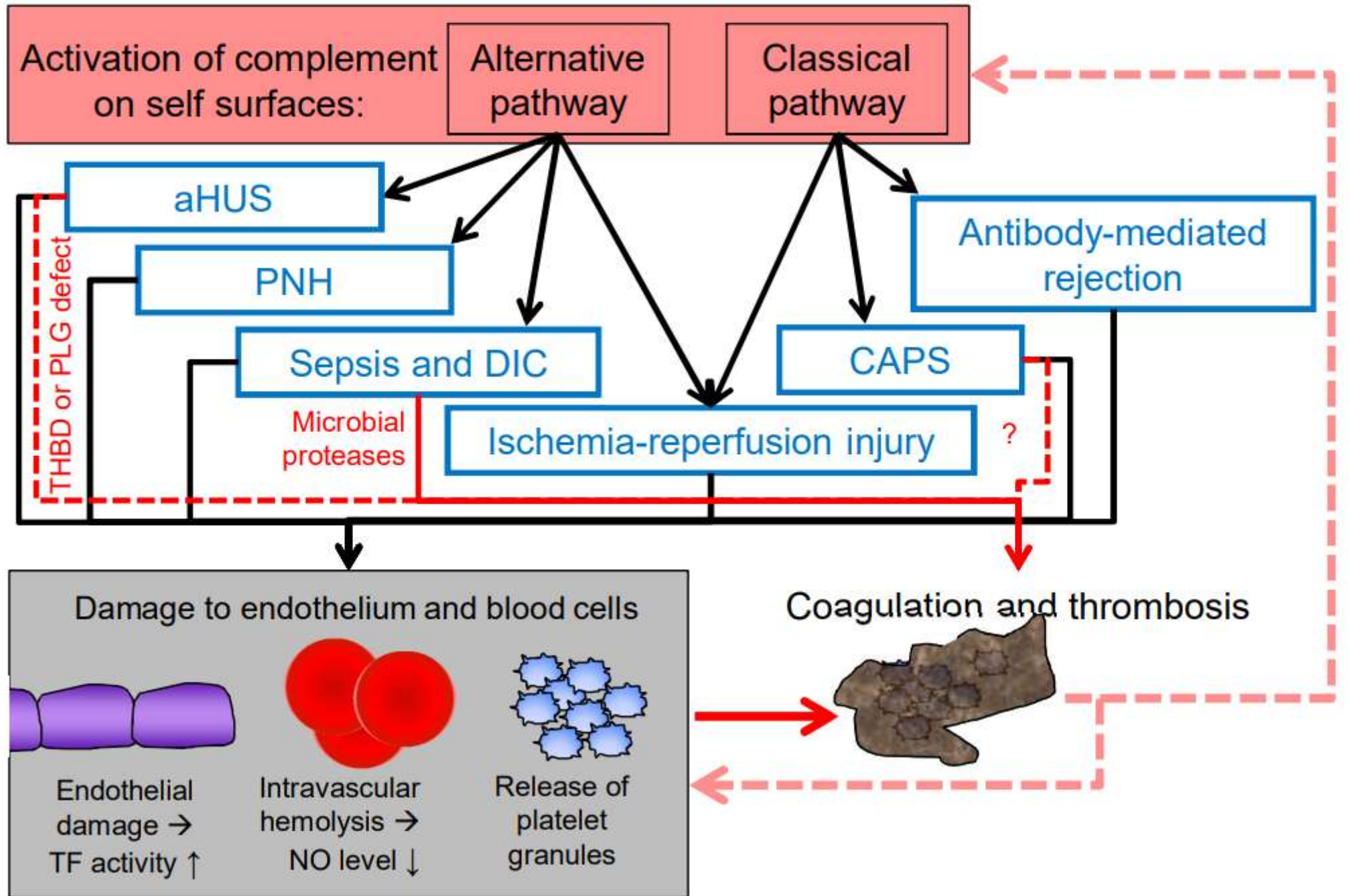
A model of the parallel pathogenic processes in HUS. Excess complement activation on endothelial cell, platelet, and red cell surfaces leads to C5a release and membrane attack complex (MAC) formation.



T. Sakari Jokiranta Blood 2017;129:2847-2856

69% of Patients With aHUS Showed Their First Clinical Manifestation while experiencing One of the Following Complement Amplifying Conditions ¹



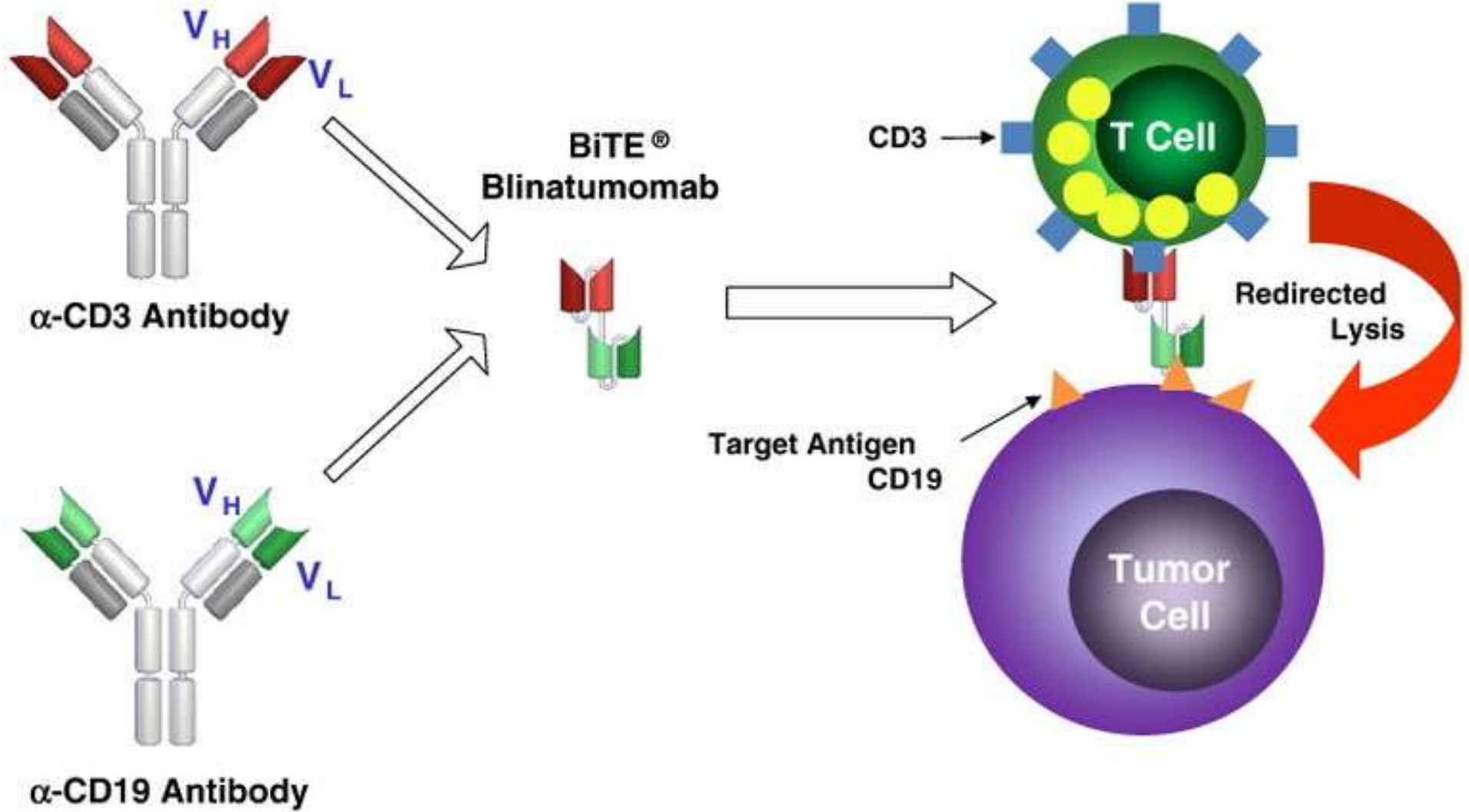


CYTOKINE RELEASE SYNDROME

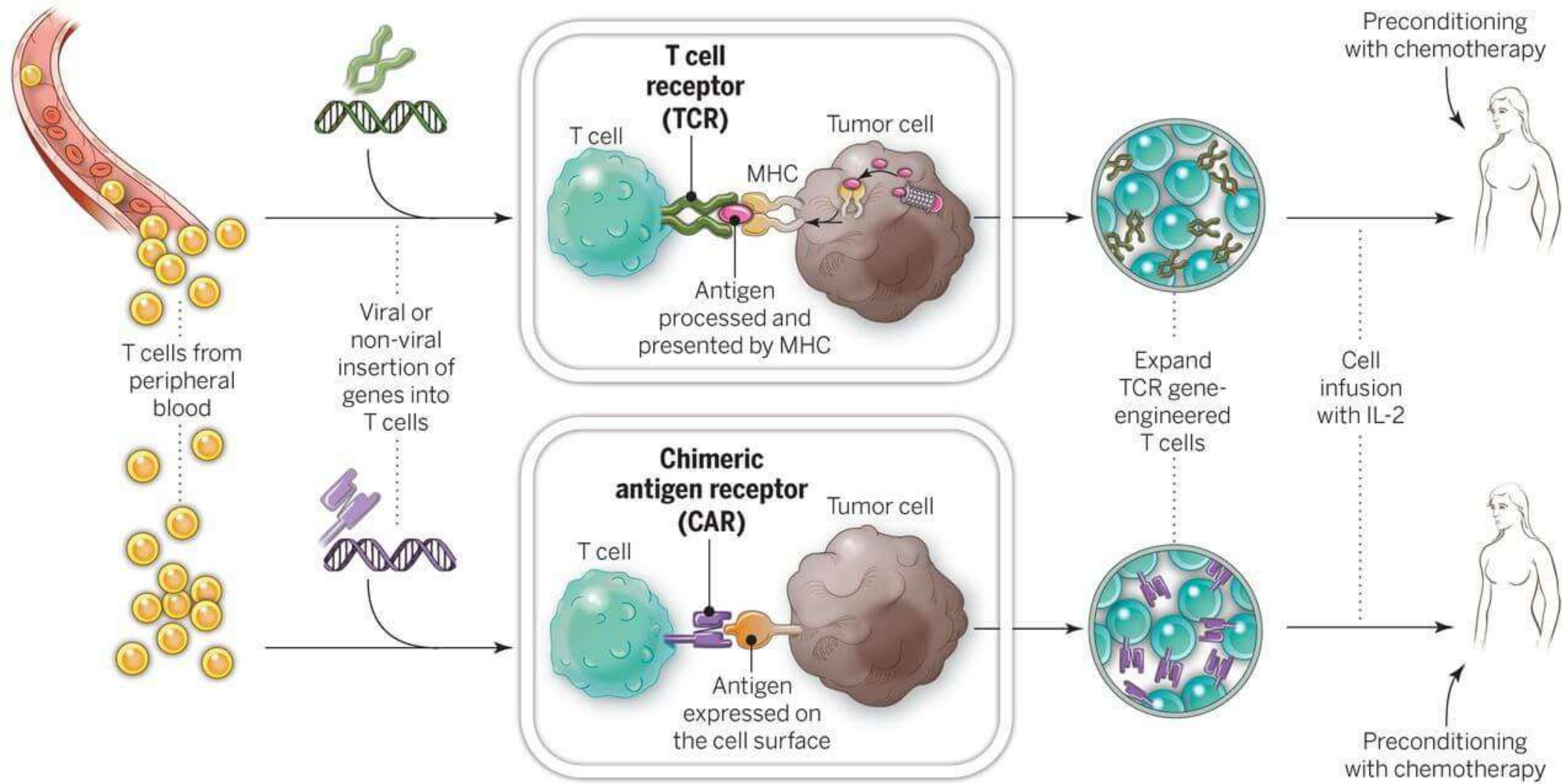
Cytokine release syndrome

- Komplikace imunoterapie
 - T cell engager - blinatumomab
 - CAR T cells (94% CRS, 27% sCRS)
 - PD1 inhibitory
- Transplantace krvetvorby od haploidentických dárců bez T deplece

Blinatumomab



CAR T cells



tisagenlecleucel

 **KYMRIAH™**

NDC 0078-0846-19

Human T cells Rx only

Suspension for IV infusion

Cultured, genetically modified

For autologous use only

Target Total Volume 10mL-50mL per bag

Dispense with Medication Guide

Dosage: See prescribing information.

Contains 2×10^6 to 2.5×10^6 CAR-positive viable T cells

Cryopreserved in: 31.25% (v/v) of Plasma-Lyte A, 31.25% (v/v) of 5% Dextrose/0.45% sodium chloride, 20% (v/v) of 25% HSA, 10% (v/v) of 10% Dextran 40 (LMD)/5% Dextrose and 7.5% (v/v) DMSO

Store at $\leq -120^\circ\text{C}$; vapor phase of liquid nitrogen

Name: John Doe

DOB: 01-JAN-2000

DIN: W1234 17 123456

Expiry: 01-JAN-2018

Batch: 12345678

Properly identify intended recipient and product

Do not use leukocyte depleting filter

Do not irradiate


Not evaluated for infectious substances

Mfd. by: Novartis Pharmaceuticals Corporation
Morris Plains, NJ 07950

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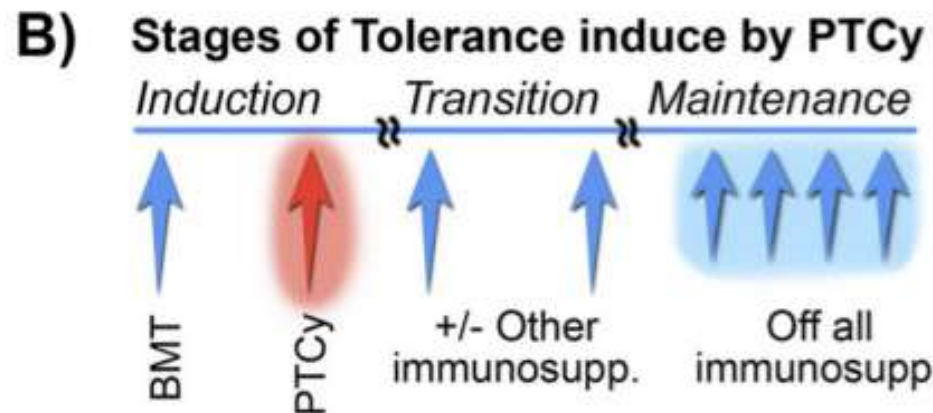
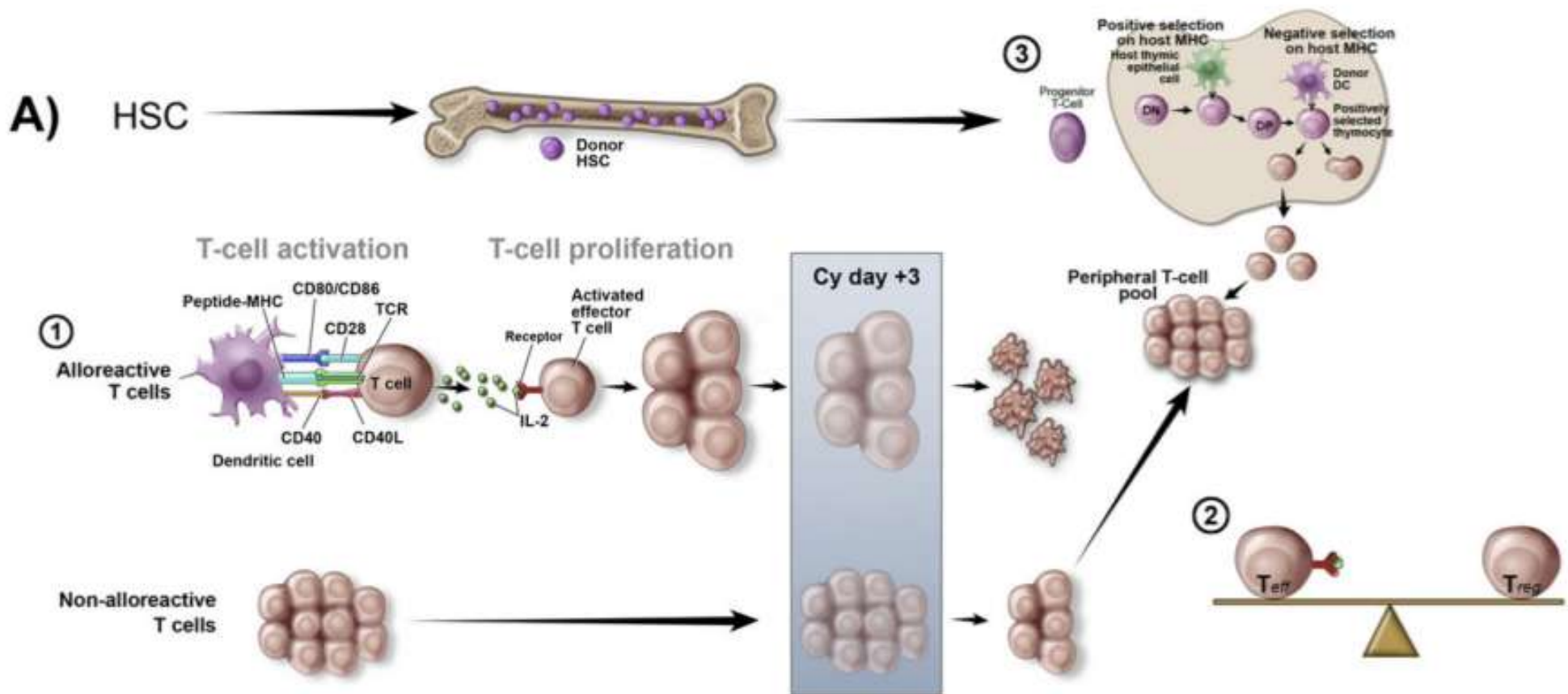
PP Material No. 8123456

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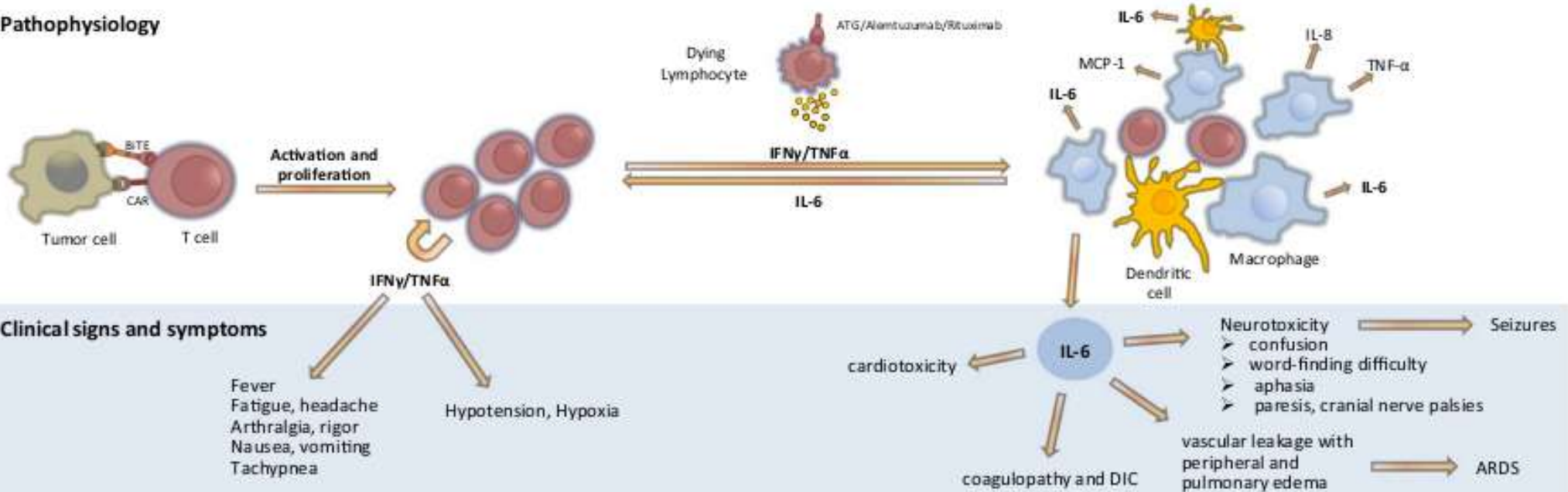
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Pathophysiology



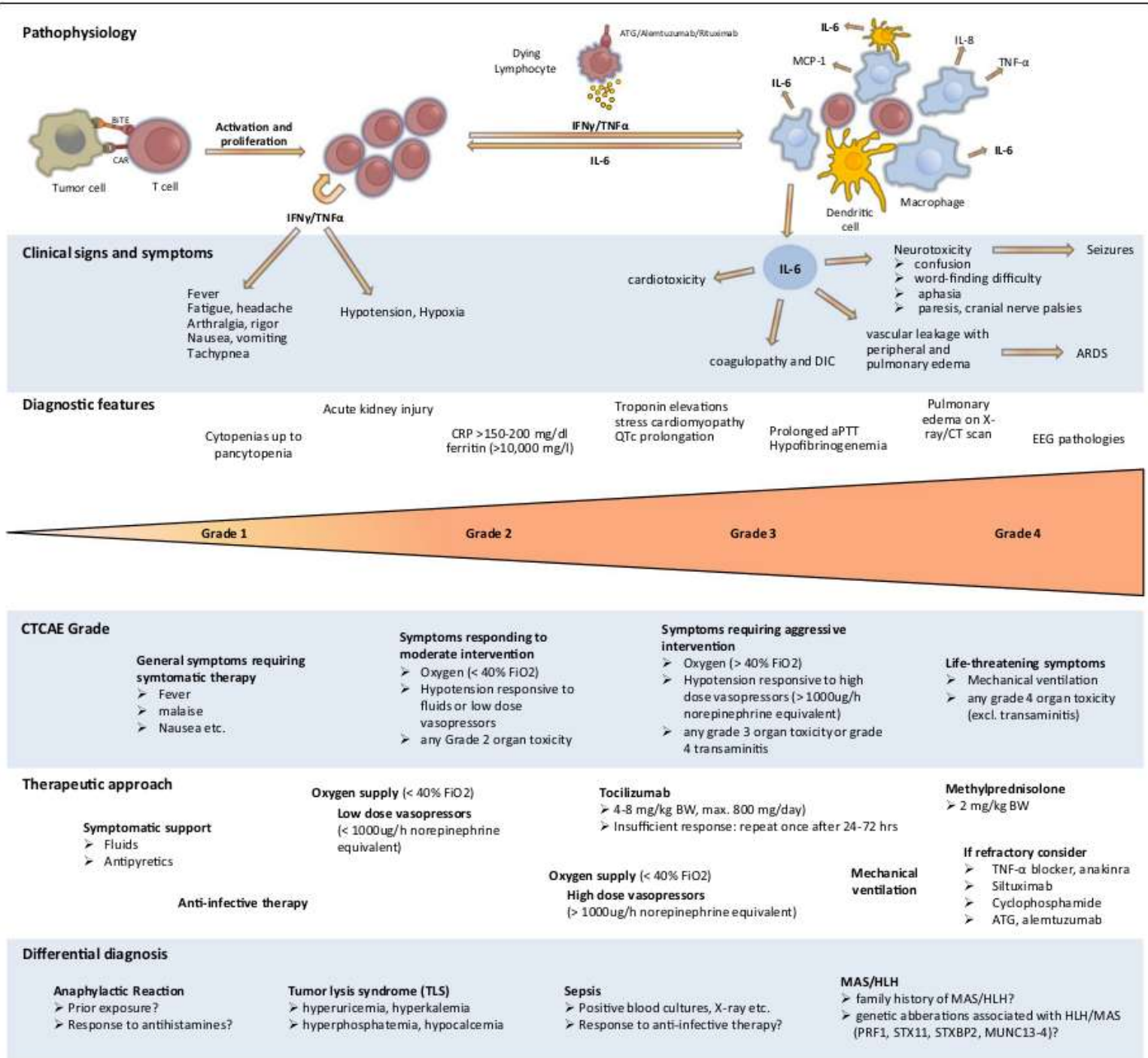


Fig. 1 Pathogenesis and management of CRS. ATG anti-thymocyte globulin, BiTE bispecific T cell engager, CAR chimeric antigen receptor, IFNγ interferon gamma, TNFα tumor necrosis factor alpha, IL-6 interleukin 6, MCP-1 monocyte chemoattractant protein 1, DIC disseminated intravascular coagulation, ARDS acute respiratory distress syndrome, FiO2 fraction of inspired oxygen, BW body weight, MAS macrophage activation syndrome, HLH hemophagocytic lymphohistiocytosis