



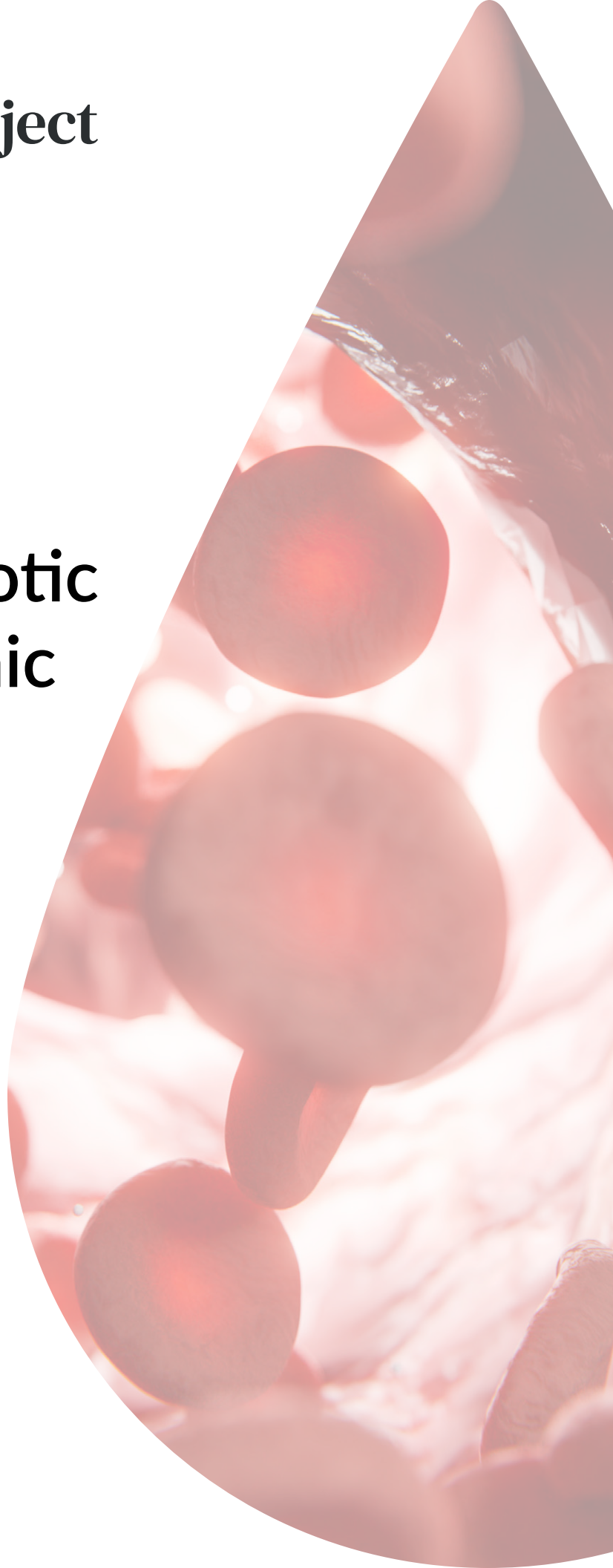
The **Blood** Project

Pocket Resource

# Diagnosis and Management of Immune Thrombotic Thrombocytopenic Purpura (TTP)

A Pocket Resource for Clinicians

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# Diagnosis

**CONSIDER A DIAGNOSIS** of immune TTP in any adult with:

- ◆ Microangiopathic hemolytic anemia (MAHA)
- ◆ Thrombocytopenia

**CONFIRM DIAGNOSIS** of immune TTP by demonstrating:

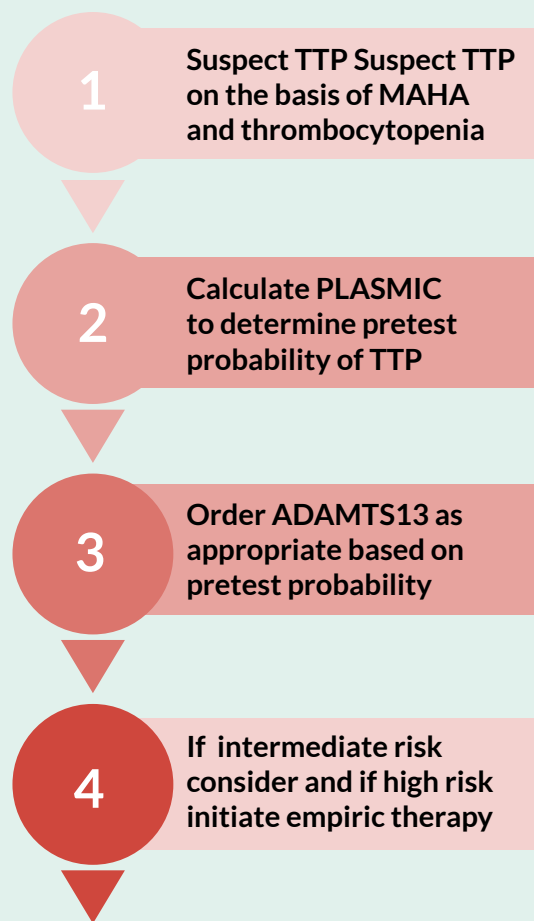
- ◆ Reduced plasma ADAMTS13 activity (< 10%)
- ◆ Presence of a functional inhibitor of ADAMTS13

ADAMTS13 activity levels usually take several days to come back. Thus, **CONDITIONAL DIAGNOSIS** (to initiate appropriate treatment) is based on clinical scoring system such as the **PLASMIC score**:

Plasmic Score	Parameter	Result	Points
	Platelet count	< 30	+1
	Hemolysis	Indirect bilirubin > 2 mg/dL or retics > 2.5% or undetectable haptoglobin	+1
	Creatinine	< 2.0 mg/dL	+1
	No active cancer in past year		+1
	No history of Solid organ or stem cell transplantation		+1
	INR	< 1.5	+1
	MCV	< 90 fL	+1
<b>Plasmic score</b>	<b>Pretest Probability</b>	<b>Risk Group</b>	
0-4	Score 0-4 0--4%	Low	
5	Score 5 5-25%	Intermediate	
6-7	Score 6-7 62-82%	High	

MCV, mean cell volume

## Workflow



### ORDER SET ADAMTS13

#### MAHA Low PLT

- ◆ CBC
- ◆ Retic count
- ◆ Peripheral smear
- ◆ LDH
- ◆ Haptoglobin
- ◆ Indirect bilirubin

#### Organ dysfunction

- ◆ Creatinine
- ◆ Troponin
- ◆ LFTs
- ◆ EKG
- ◆ CT/MRI of brain if CNS findings

#### Secondary causes, other causes of TMA

- ◆ HIV
- ◆ HCV
- ◆ HBV
- ◆ ANA
- ◆ PT/aPTT
- ◆ B12
- ◆ DAT

PLT, platelet count; TMA, thrombotic microangiopathy; LDH, lactate dehydrogenase; LFTs, liver function tests; ANA, antinuclear antibody; B12, vitamin B12; DAT, direct antiglobulin test

# Treatment (1st acute event)

Based on 2 clinical practice guidelines: International Society of Thrombosis (ISTH) and Haemostasis and British Society of Hematology (BSH)

## ISTH

J Thromb Haemost. 2020;18:2486

J Thromb Haemost. 2020;18:2496

**If high clinical suspicion based on risk assessment method**

**While waiting for results of ADAMTS13:**

♦ Start **TPE** and **corticosteroids** *without* waiting for the results of ADAMTS13 testing (strong recommendation)

♦ Consider early administration of **caplacizumab** (conditional recommendation)

ADAMTS13 activity result

<10-20%\*

<10%

>20%



**Continue**

caplacizumab



**Consider**  
**rituximab**

(conditional  
recommendation)

**Stop**

caplacizumab

\*If ADAMTS13 10%-20%,  
use clinical judgment

In general, prophylactic platelet transfusions are avoided in nonbleeding TTP; may be considered if serious bleeding

TPE, therapeutic plasma exchange

## BSH

Br J Haematol. 2023;203:546

**Treat TTP as a medical emergency (1A)**

**While waiting for results of ADAMTS13:**

♦ Start *daily* TPE\* and corticosteroids\*\* *without* waiting for the results of ADAMTS13 testing (1A)

ADAMTS13 activity result

<10%

>20%



**Start**

♦ caplacizumab\*\*\* (1A)

♦ rituximab (1B)

**Provide thromboprophylaxis once platelet counts are  $\geq 50 \times 10^9/L$**

**Platelet transfusion should be avoided (1B)**

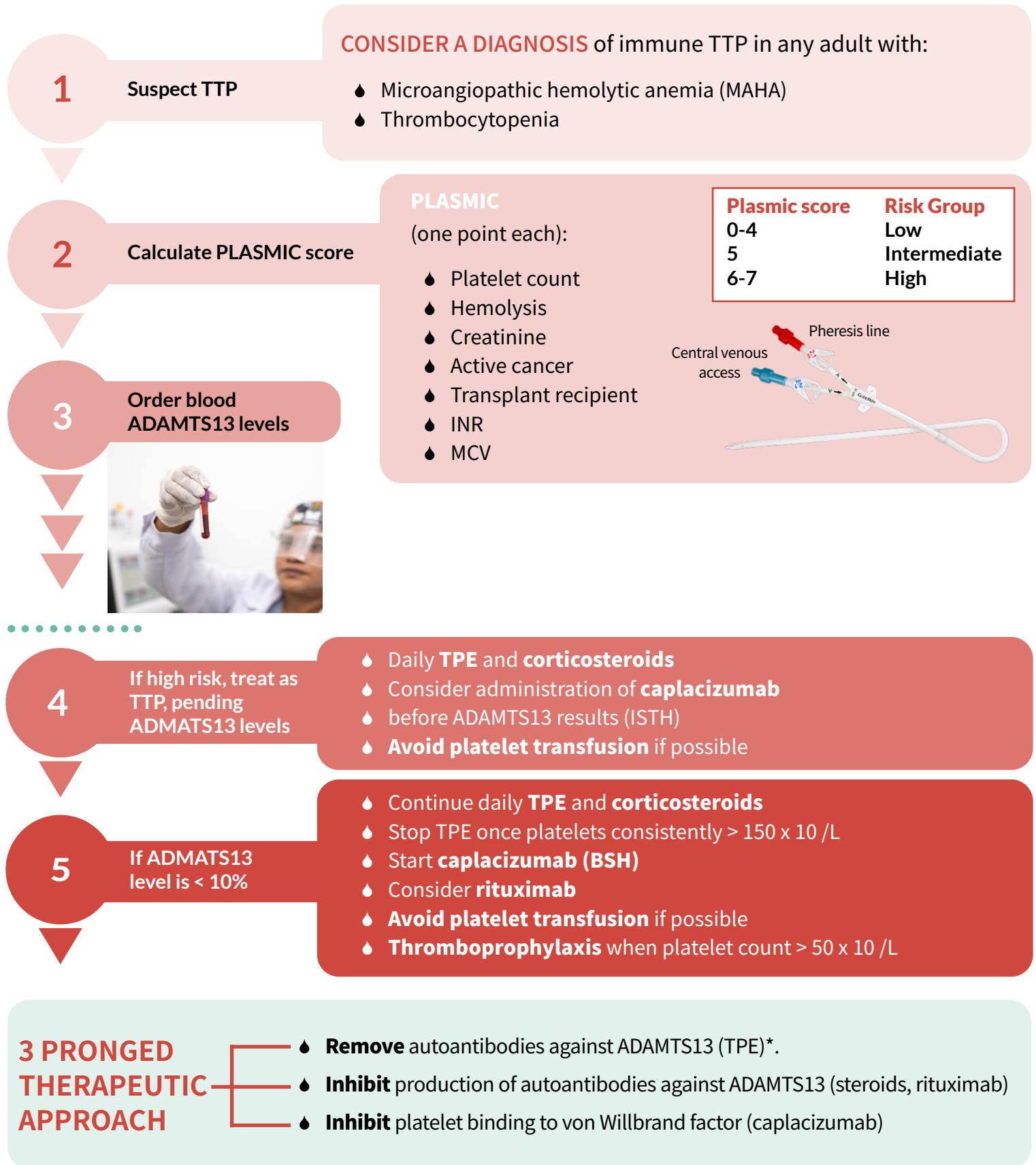
\* TPE should be initiated within four to eight hours and continued daily (1-1.5 x volume replacement). Stop TPE when sustained PLT  $>150 \times 10^9/L$ . for additional guidance see AFSA guideline on the use of therapeutic apheresis. J Clin Apher 2023;38:77

\*\*Prednisone equivalent of 1 mg/kg/day

\*\*\*IV dose of caplacizumab 10 mg is given pre-TPE. A once daily 10 mg sc is continued up to 30 days following completion of TPE. Can extend an additional 28 days beyond this based on ADAMTS13 activity < 30%.

# Treatment (1st acute event)

## Overview



\*TPE also repletes ADAMTS13 and removes ultra-long vWF multimers