PIASTRINOPENIA
Gravità/Frequenza delle Manifestazioni Cliniche

- Conta piastrinica

  rare se >$50x10^9$/L

  relativamente frequenti se <10-$20x10^9$/L
- Tipo di piastrinopenia

  più rare in quelle da aumentata distruzione periferica
- Concomitanza di altri fattori di rischio

  traumi, febbre, infezioni, anemia, rapidità di insorgenza, età

Subgroup Analysis of Odds of Hemorrhage in 117 Patients With ITP

Age (y)	Person-years	No. of events	Person-time incidence rates	Odds Ratio	P
<40	257	1	0.4	1.0	-
40 – 60	177	2	1.1	2.8	NS
>60	67	7	10.4	28.9	<.01

Cortelazzo et al, 1991

PIASTRINOPENIA
Classificazione

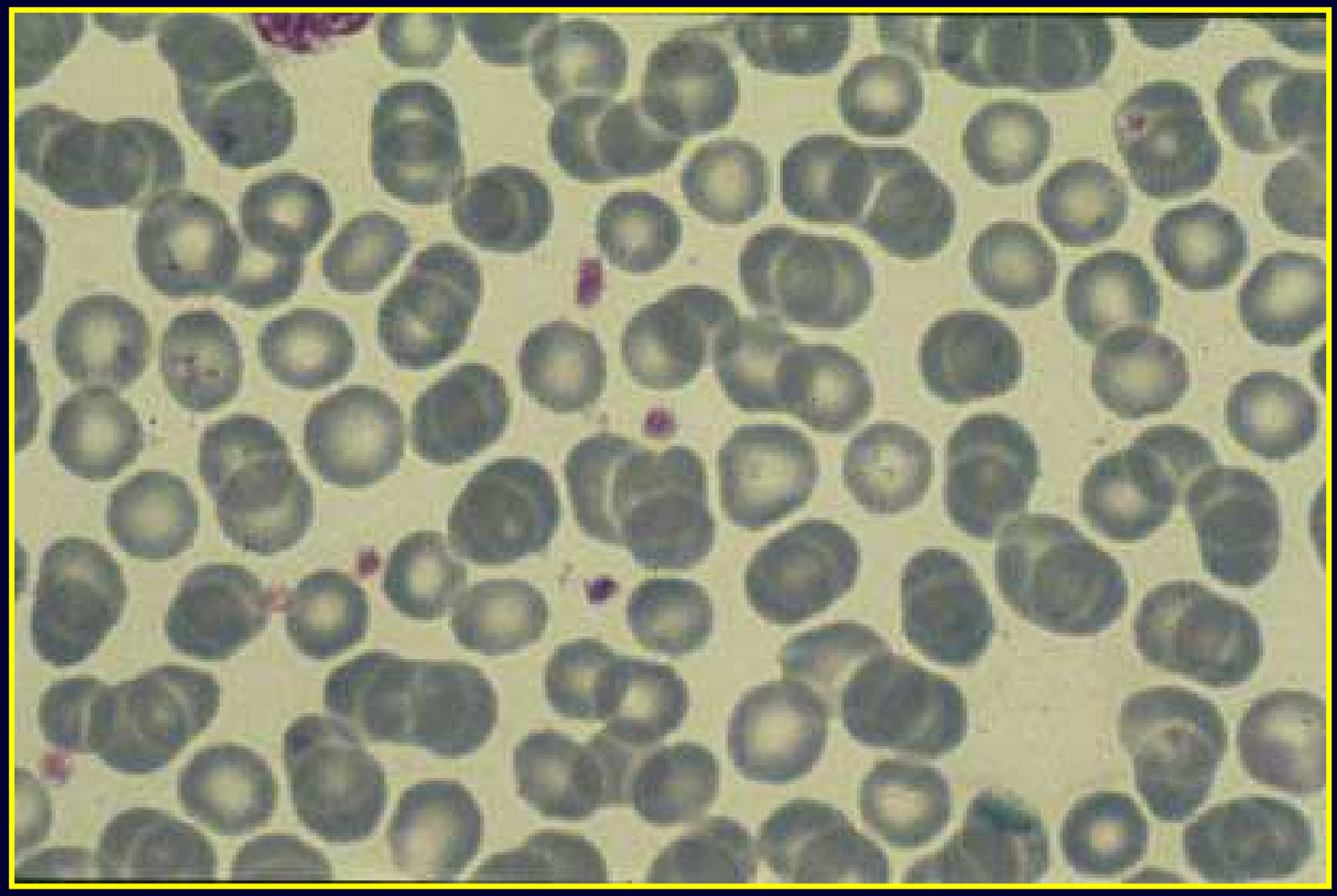
- **PIASTRINOPENIE EREDITARIE**
- **PIASTRINOPENIE ACQUISITE:**
  - Da ridotta o difettosa produzione midollare
  - Da aumentata distruzione/consumo periferico
    - su base immune
    - su base non immune
  - Da sequestro od anomalo pooling
  - Da emodiluizione
- **PSEUDOPIASTRINOPENIA !!!**

PIASTRINOPENIA

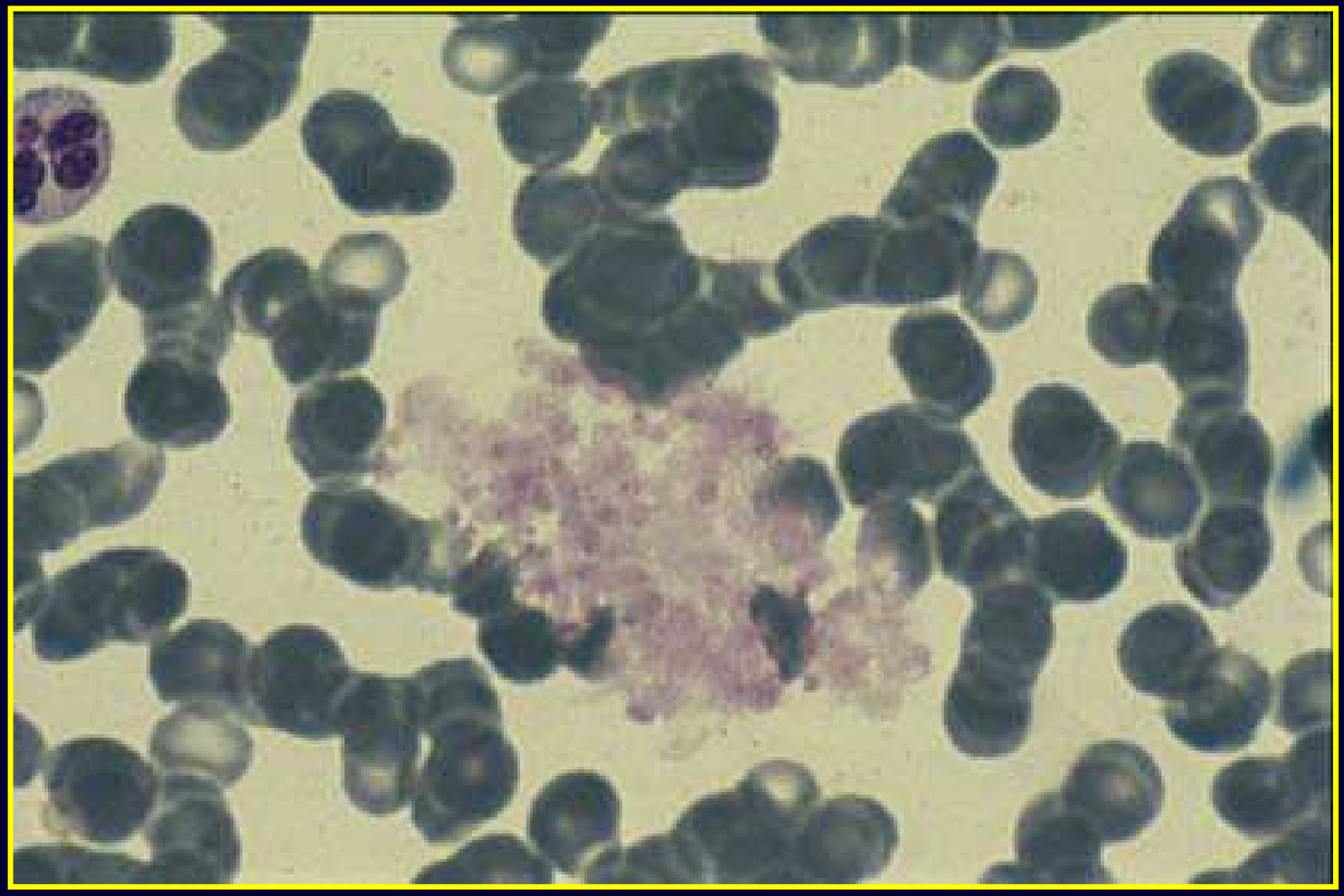
Pseudopiastrinopenia?

- Striscio di sangue periferico nativo e in EDTA

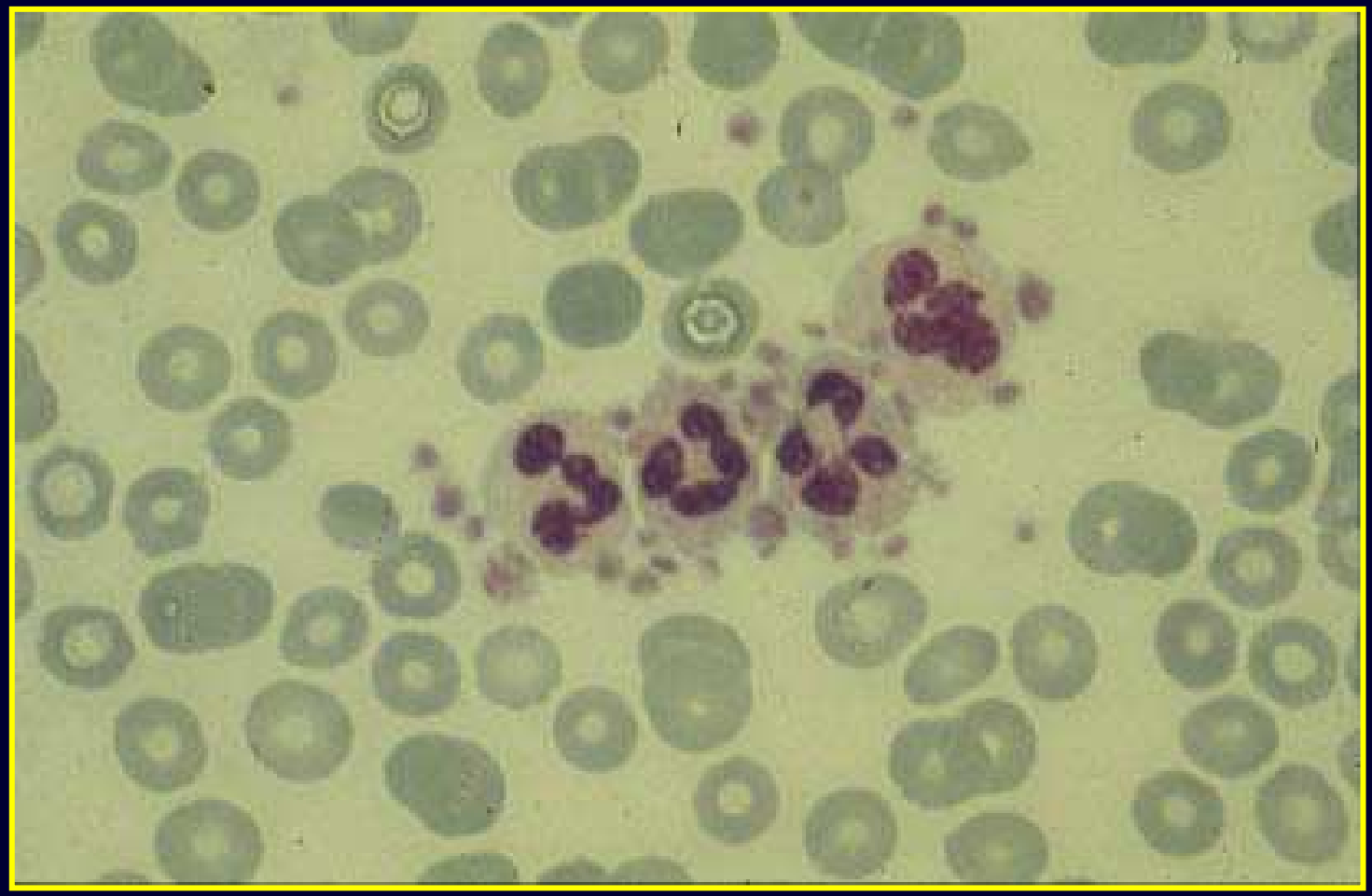
Normale – Sangue in EDTA



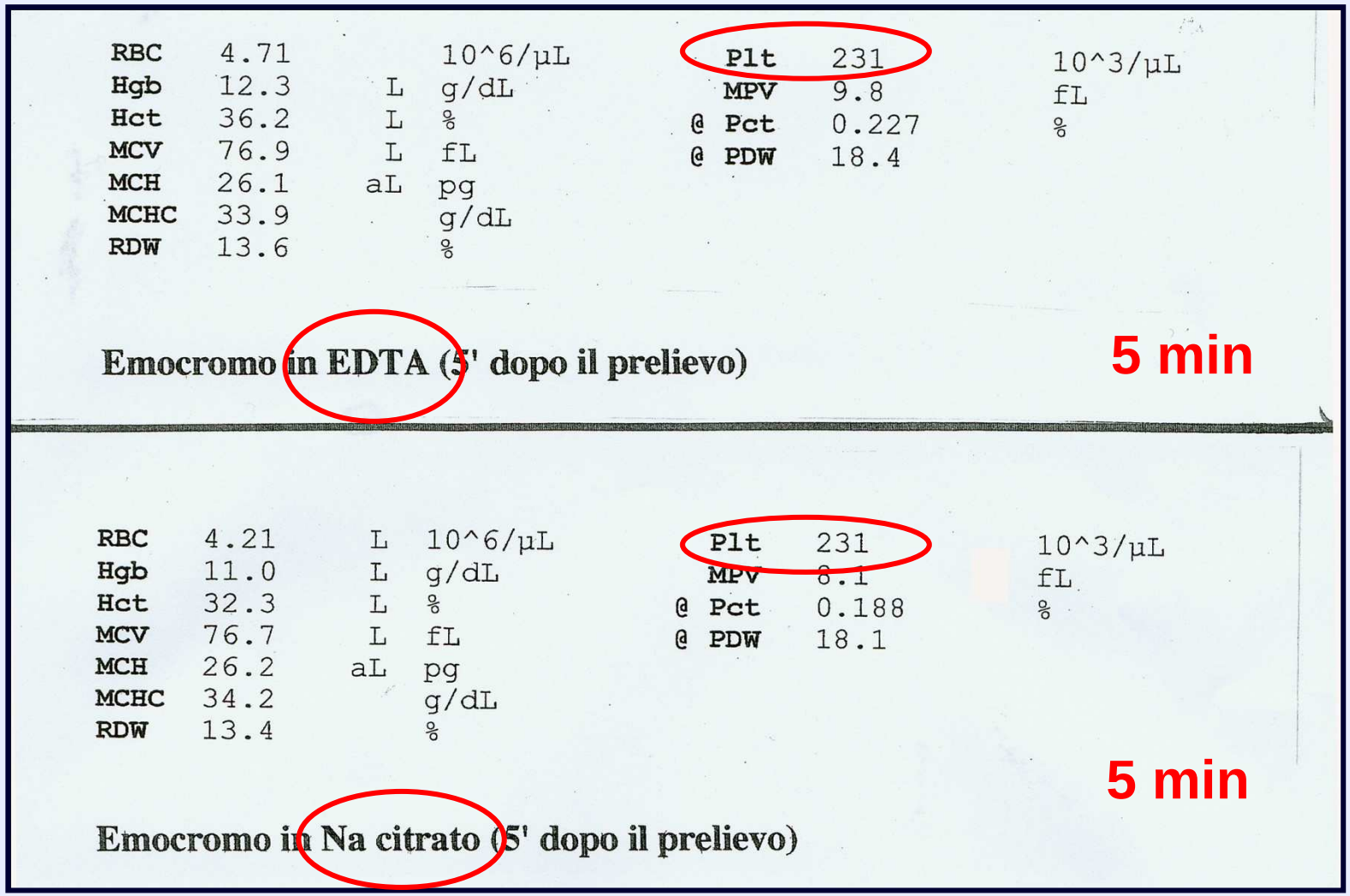
Pseudopiastrinopenia – Sangue in EDTA



Satellitismo piastrinico



PSEUDOPIASTRINOPENIA



RBC	4.71		10^6/µL
Hgb	12.3	L	g/dL
Hct	36.2	L	%
MCV	76.9	L	fL
MCH	26.1	aL	pg
MCHC	33.9		g/dL
RDW	13.6		%

Plt	231	10^3/µL
MPV	9.8	fL
@ Pct	0.227	%
@ PDW	18.4	

Emocromo in EDTA (5' dopo il prelievo)

5 min

RBC	4.21	L	10^6/µL
Hgb	11.0	L	g/dL
Hct	32.3	L	%
MCV	76.7	L	fL
MCH	26.2	aL	pg
MCHC	34.2		g/dL
RDW	13.4		%

Plt	231	10^3/µL
MPV	8.1	fL
@ Pct	0.188	%
@ PDW	18.1	

5 min

Emocromo in Na citrato (5' dopo il prelievo)

PSEUDOPIASTRINOPENIA

RBC	4.81		10^6/µL	Plt	49	10^3/µL
Hgb	12.3	L	g/dL	MPV	8.7	fL
Hct	37.2	L	%	@ Pct	0.042	%
MCV	77.1	L	fL	@ PDW	17.7	
MCH	25.4	aL	pg			
MCHC	33.0		g/dL			
RDW	13.7		%			

Emocromo in EDTA (3 ore dopo il prelievo) **3 h**

RBC	4.33	L	10^6/µL	Plt	200	10^3/µL
Hgb	11.0	L	g/dL	MPV	9.1	fL
Hct	33.6	L	%	@ Pct	0.182	%
MCV	77.6	L	fL	@ PDW	18.0	
MCH	25.4	aL	pg			
MCHC	32.8		g/dL			
RDW	13.5		%			

Emocromo in Na citrato (3 ore dopo il prelievo) **3 h**

Metodi

- **Conta piastrinica in EDTA e CPT**
  - **al tempo 0 min (max 10 min dopo il prelievo)**
  - **al tempo 90 min**

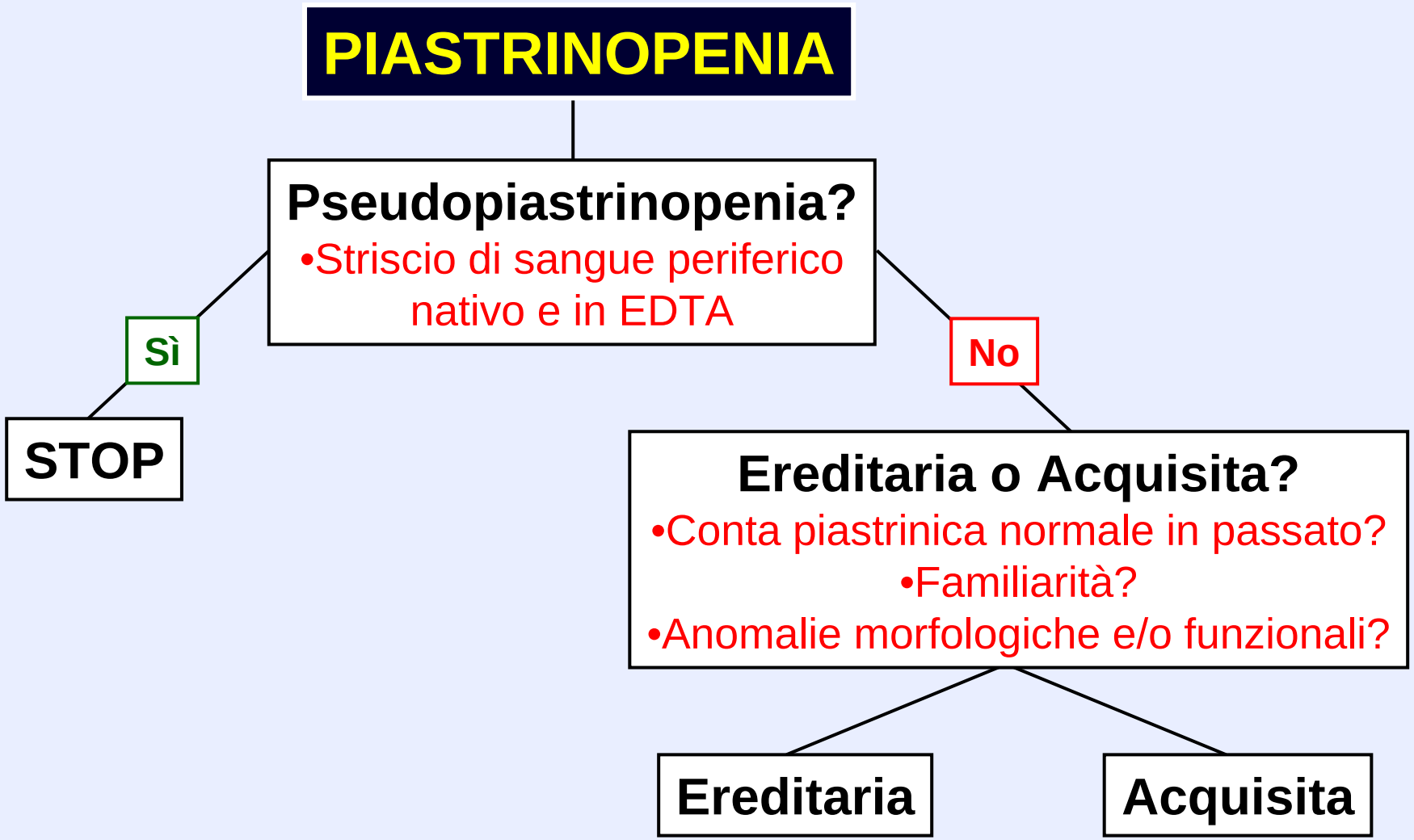
- **Striscio di sangue periferico in EDTA e CPT**
  - **al tempo 0 min (max 10 min dopo il prelievo)**
  - **al tempo 90 min**

Sangue anticoagulato EDTA

FASCE DI RISCHIO

TOT pazienti che cambiano categoria	19

Piastrine (x 10^3)	EDTA t_0 vs EDTA t_{90}
(>100) ⟶ (<100)	9/107 (8%)
≥50 ⟶ (20-49)	6/107 (6%)
≥20 ⟶ (0-19)	4/107 (4%)


PIASTRINOPENIA
Pseudopiastrinopenia?
•Striscio di sangue periferico nativo e in EDTA
Sì
STOP
No
Ereditaria o Acquisita?
•Conta piastrinica normale in passato?
•Familiarità?
•Anomalie morfologiche e/o funzionali?
Ereditaria
Acquisita

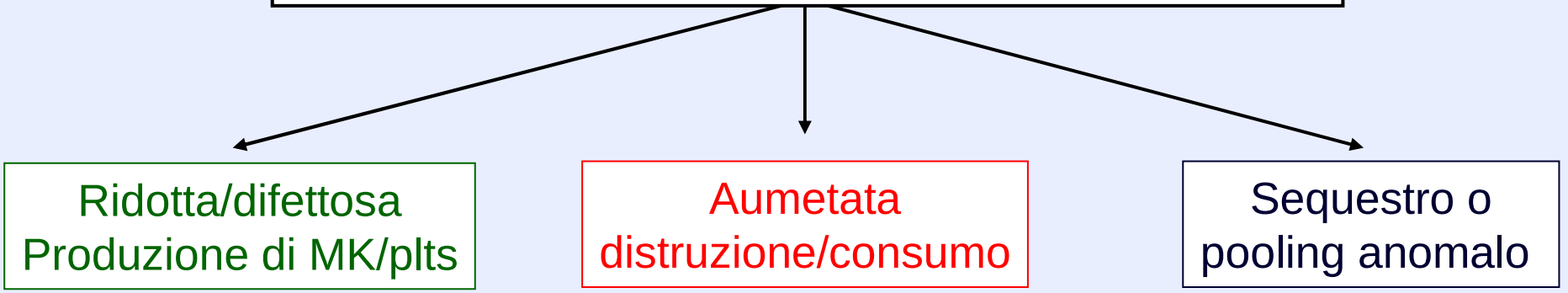
Inherited Thrombocytopenias: a Proposed Diagnostic Algorithm from the Italian *Gruppo di Studio delle Piastrine*

CL Balduini, M Cattaneo, F Fabris, P Gresele, A Iolascon, FM Pulcinelli, A Savoia, on behalf of the Italian *Gruppo di Studio delle Piastrine*

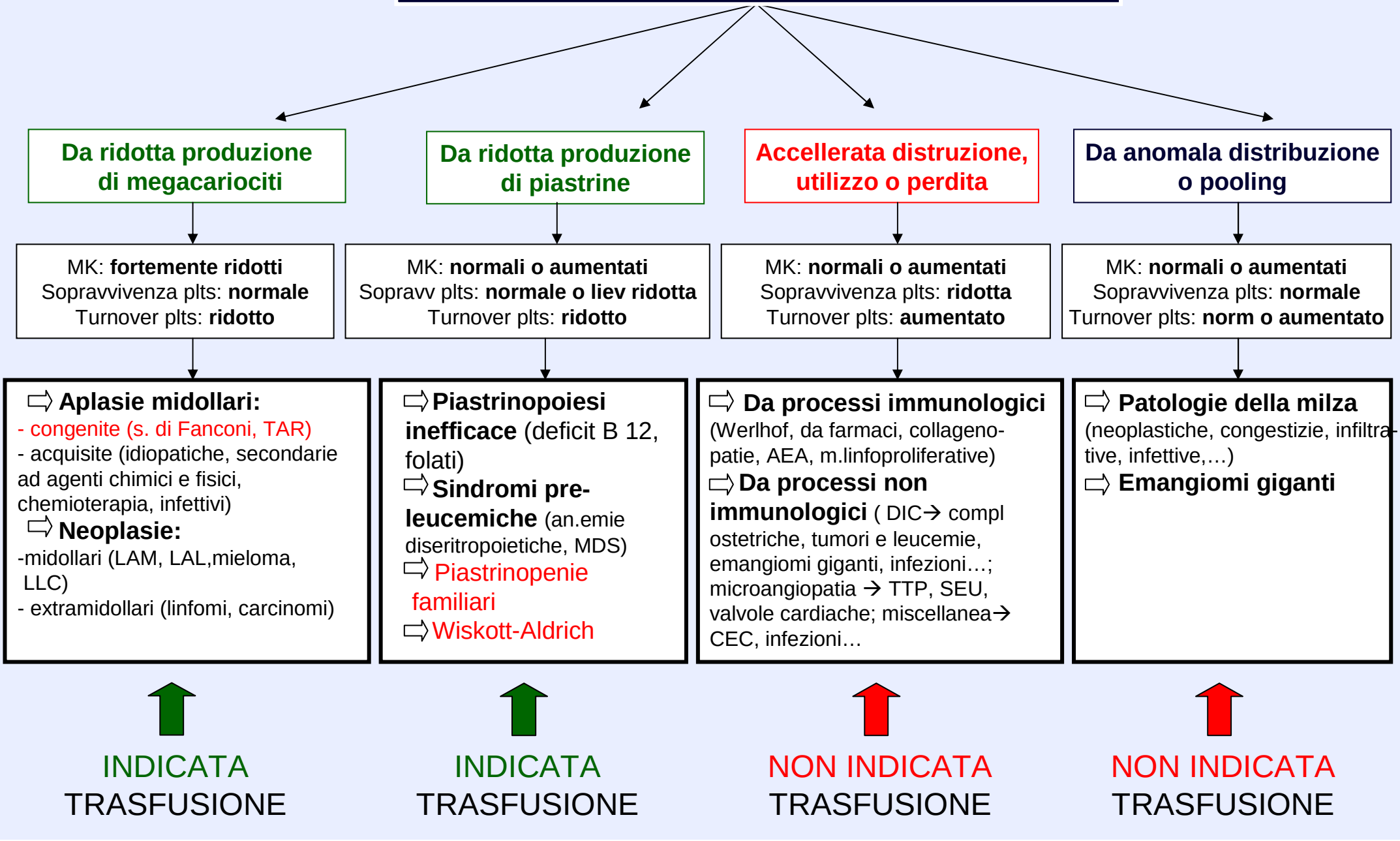
Haematologica 2003, 88: 582-592

PIASTRINOPENIA ACQUISITA

- [Emocromo, MPV]
- [Striscio di sangue periferico]
- Anamnesi farmacologica e trasfusionale
- Valutazione splenomegalia
- Markers virus epatite, Herpes, HIV
- Elettroforesi sieroproteica
- Analisi aspirato midollare (obbligatorio se >60 anni)
- Ricerca ANA
- [Sopravvivenza piastrinica]



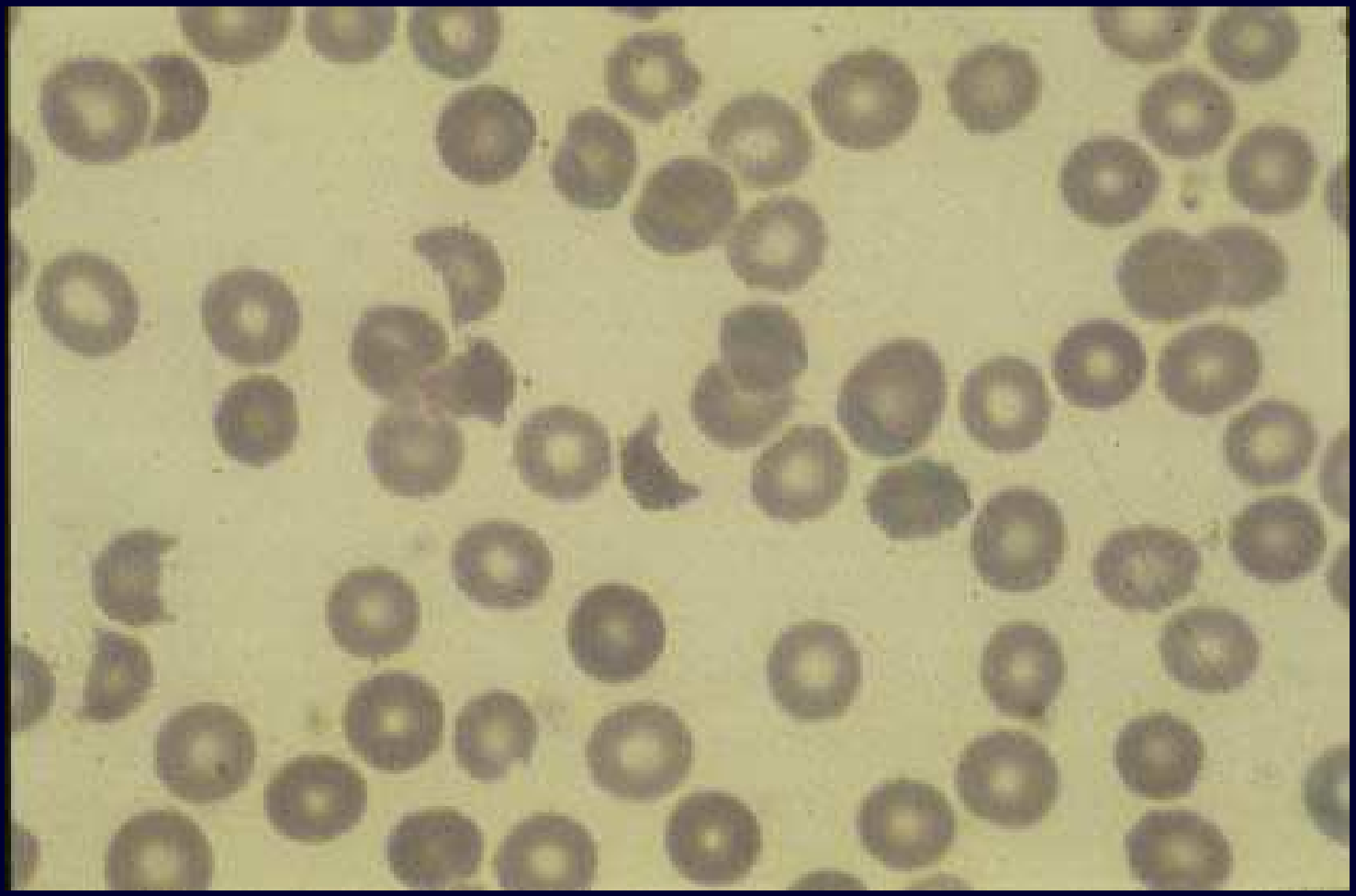
PIASTRINOPENIA ACQUISITA



Principali Farmaci Causa di Piastrinopenia Immune

• Abciximab (anche pseudopiastrinopenia)	+++
• Chinidina	+++
• Chinina	+++
• Eparina	+++
• Sali d'oro	+++
• Carbamazepina	++
• Clorotiazide	++
• Eptifibatide	++
• Rifampicina	++
• Sulfametossazolo	++
• Acido valproico	++
• Vancomicina	++

Porpora Trombotica Trombocitopenia (TTP)



CLINICAL FORMS OF THROMBOTIC MICROANGIOPATHIES

TTP

- Acute sporadic (non-familial)
- Chronic recurrent
- Familial
- Secondary: transplantation, HIV, cancer, drugs, pregnancy, estrogens, infection

HUS

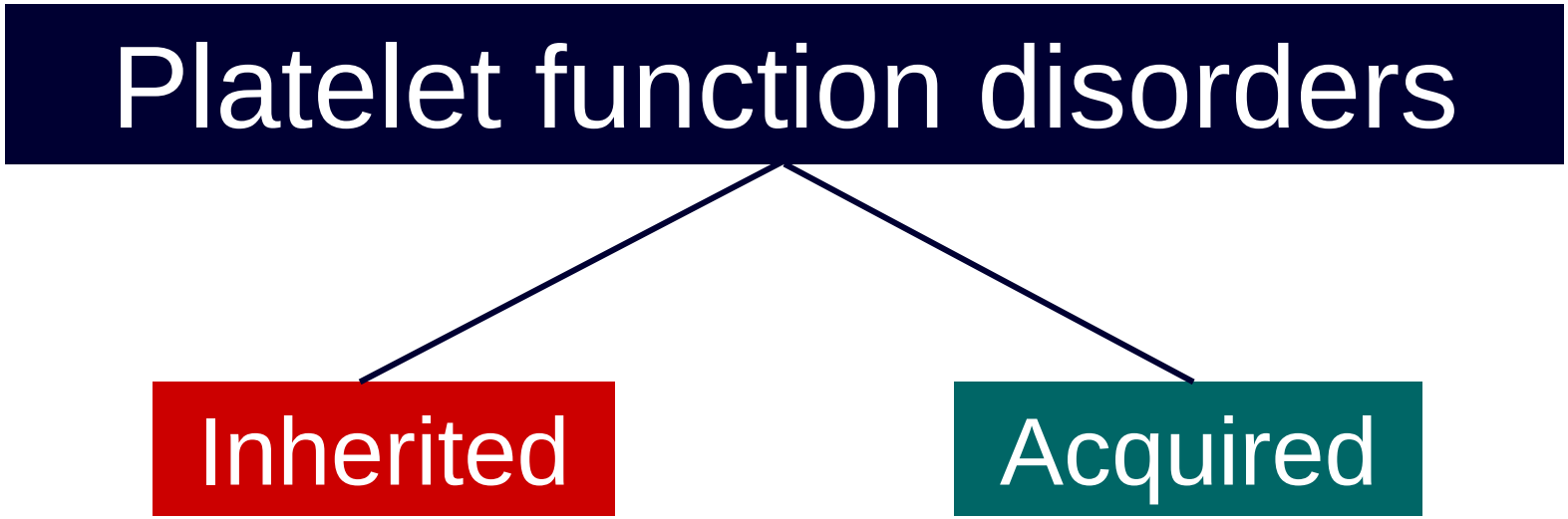
- Acute sporadic (non-familial)
- Chronic recurrent
- Familial
- Secondary: verocytotoxin, shigella toxin (diarrhea-related)

Thrombotic Thrombocytopenic Purpura (TTP)

- Thrombocytopenia
- Hemolytic anemia
- Fever
- Platelet thrombi in arterioles and capillaries
- Focal neurological symptoms

Hemolytic-Uremic Syndrome (HUS)

- Thrombocytopenia
- Hemolytic anemia
- Fever
- Platelet thrombi in arterioles and capillaries
- Renal insufficiency
- Diarrhea (in children)


Platelet function disorders
Inherited
Acquired

Acquired platelet function defects

- Drugs affecting platelet function
- Systemic disorders
  - Uremia
  - Antiplatelet antibodies
  - Cardiopulmonary bypass
  - Liver disease
  - DIC
  - Neoplasia
- Hematologic disorders
  - Chronic myeloproliferative disorders
  - Leukemias and myelodysplatstic syndromes
  - Dysproteinemias

Treatment and prophylaxis of bleedings in Platelet Function Defects
Options

- Platelet transfusions
- Recombinant Factor VIIa
- Antifibrinolytic agents (e.g., tranexamic acid)
- Desmopressin

For mild/moderate bleeding episodes, local measures (compression) can be efficacious
(e.g., epistaxis, gingival bleeding,…)

Platelet transfusions

- Should be reserved for:
  - patients with serious bleeding unresponsive to other therapies, or
  - patients with severe platelet function defects (e.g., deficient platelet membrane GPs, such as Bernard-Soulier Syndrome, Glanzmann Thrombasthenia)

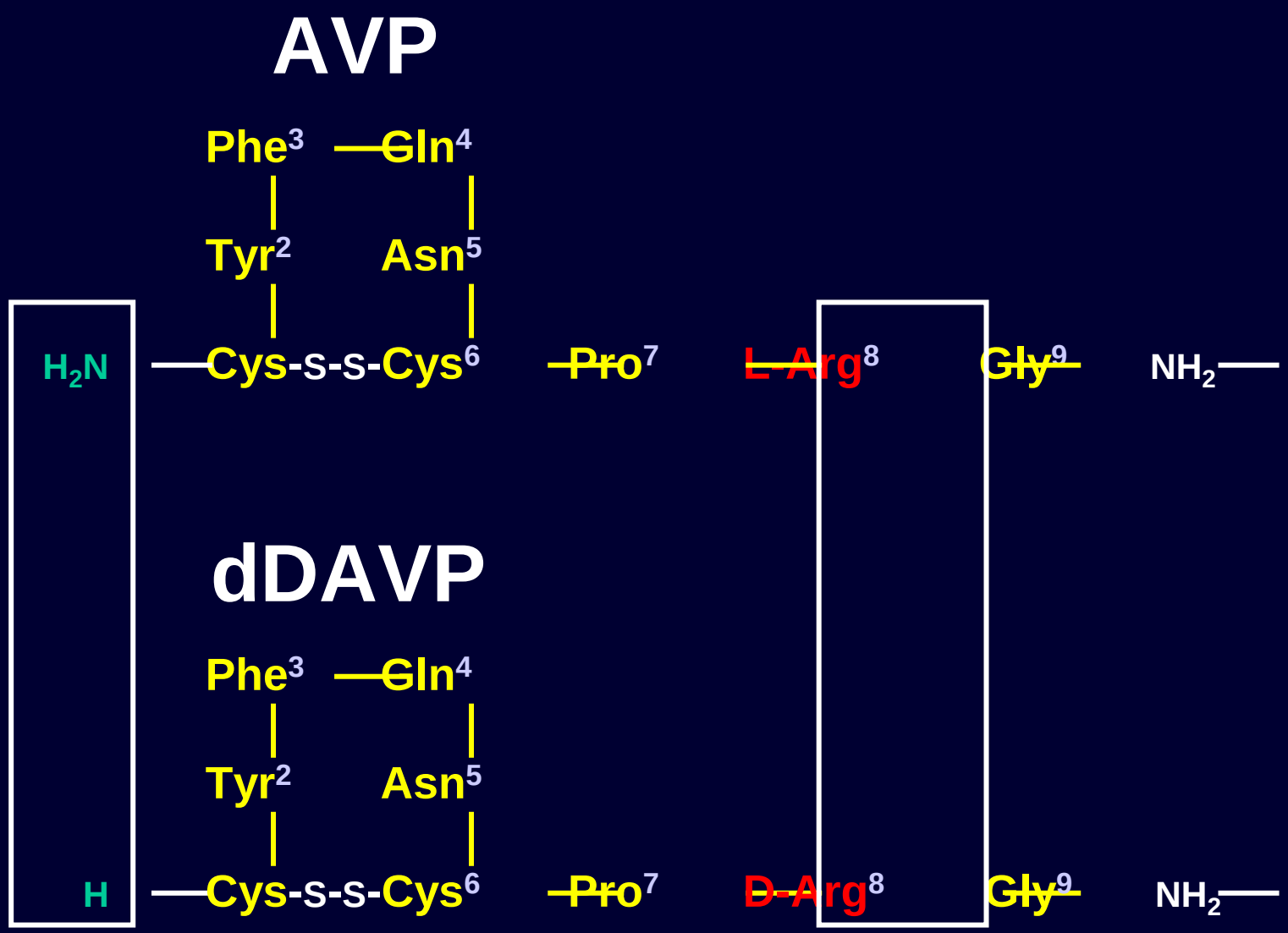
- In patients with deficient platelet membrane GPs, there is an increased risk of isoimmunization (Glanzmann Thrombasthenia > Bernard-Soulier Syndrome)

Recombinant FVIIa

Should be reserved for management of serious bleedings in patients with severe platelet function disorders, particularly in those who no longer respond to platelet transfusions because of isoimmunization

Anti-fibrinolytic agents

- Useful in adjunctive therapy for preventing and controlling bleeding with dental extraction or oral/nasal surgery
- A short coures may be useful for recurrent epistaxis
- Useful in women with menorrhagia
- MUST NOT BE USED IN PATIENTS WITH HEMATURIA
- The only therapy helpful to treat patients with the very rare Quebec platelet diroder


AVP
Phe3
Gln4
Tyr2
Asn5
H2N
Cys-S-S-Cys6
Pro7
L-Arg8
Gly9
NH2
dDAVP
Phe3
Gln4
Tyr2
Asn5
H
Cys-S-S-Cys6
Pro7
D-Arg8
Gly9
NH2