

From
Biotechnology
to Diagnosis

Diagnosing quantitative platelet glycoprotein abnormalities

PLT Gp/Receptors

Ref. # 7004

RESTING

ACTIVATED

Image courtesy of John W. Weisel, PhD, and Chandrasekaran Nagaswami, University of Pennsylvania School of Medicine

✓ **Platelet glycoprotein disorders :**

Glanzmann Thrombasthenia ⁽¹⁾
Bernard Soulier Syndrome
Fechtner Syndrome ⁽¹⁾
X-Linked Thrombocytopenia ⁽²⁾
Gray Platelet Syndrome like ⁽¹⁾

✓ **Technology :**

Quantitative flow cytometry

✓ **Sample :**

Citrated whole blood (only 100 µL)

✓ **Biological evaluations :**

⁽¹⁾ Schlegel N. *et al.*; Blood (1998), suppl. 1, 92, abst.3367.
⁽²⁾ Schlegel N. *et al.*; Thromb. Haemost. (1999), Suppl. 797, abst. 2524.
⁽³⁾ Hézard N. *et al.*; Thromb Haemost. (2003), 90, 116-123.

CE marked.

In Europe, For In Vitro Diagnostic Use



140, Chemin de l'Armée d'Afrique - 13010 Marseille - France
RCS Marseille 347 897 985
Tél. : 04 96 12 20 40 - Fax : 04 91 47 24 71
Tél. : Int. + (33) 4 96 12 20 40 - Fax : Int. + (33) 4 91 47 24 71
E-mail : info@biocytex.fr - Website : www.biocytex.fr



Diagnosing quantitative platelet glycoprotein abnormalities :

Background :

Platelet plays a key role in maintaining the hemostatic balance. Activating signals lead to platelet morphological and biochemical modifications associated with variations in the surface glycoprotein expression. These glycoproteins constitute major complexes involved in adhesion (GpIb/IX/V, vWF receptor), in aggregation (GpIIb/IIIa, Fibrinogen receptor) or can reflect platelet activation (GMP140, P-Selectin). Thrombopathies characterized by quantitative platelet glycoprotein abnormalities have been widely reported.

Platelet glycoprotein disorders :

Glanzmann Thrombasthenia :

Quantitative or qualitative inherited deficiency in GpIIb/IIIa.

Bernard Soulier syndrome :

Inherited disorder characterized by thrombocytopenia, giant platelets and GpIb/IX/V deficiency.

Fechtner syndrome :

Thrombocytopenia associated with large/giant platelets.

X-Linked Thrombocytopenia :

Thrombocytopenia with small-sized platelets.

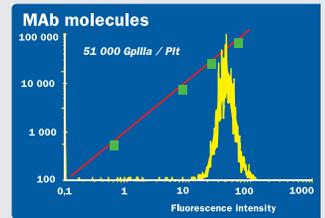
Gray Platelet Syndrome like :

α -granules defect characterised by a reduced P-selectin externalization at the activated state.



Diagnostic test :

With *PLT* Gp/Receptors kit, the whole blood sample is diluted in presence or absence of TRAP (Thrombin Receptor Agonist Peptide). After dilution, this sample is incubated with different MAb directed against human GpIIb (CD41), GpIb α (CD42b) and GMP140 (CD62P) and with a negative isotypic control. The mean fluorescence intensity (MFI) is measured with a flow cytometer after addition of a staining reagent to the test tubes and to the calibrator. Through this calibrator (coated with defined increasing numbers of MAb molecules), the MFI is converted into the absolute number of MAb molecules bound per platelet (ABC). The results are expressed in sABC (specific ABC) equivalent in our system to the number of Gp molecules per platelet.

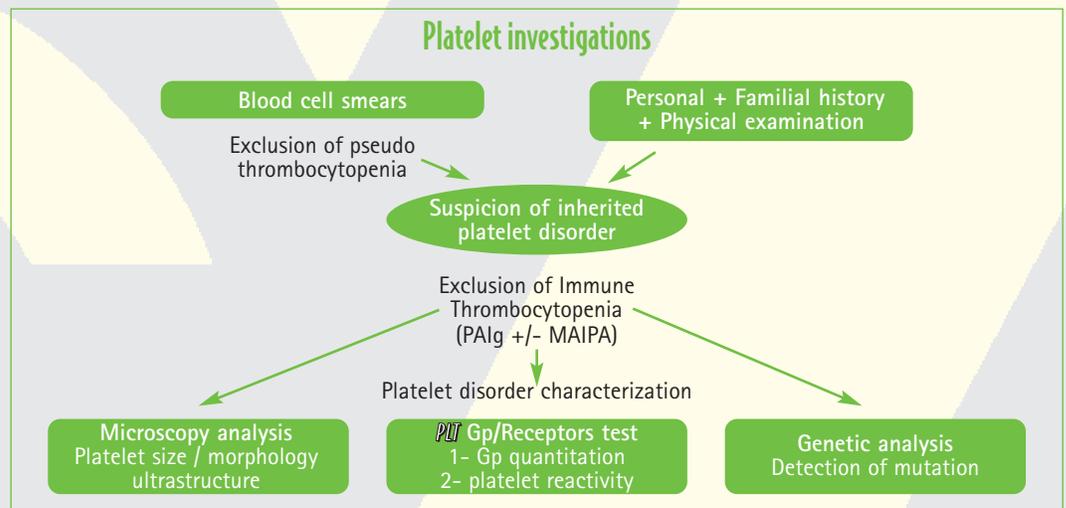
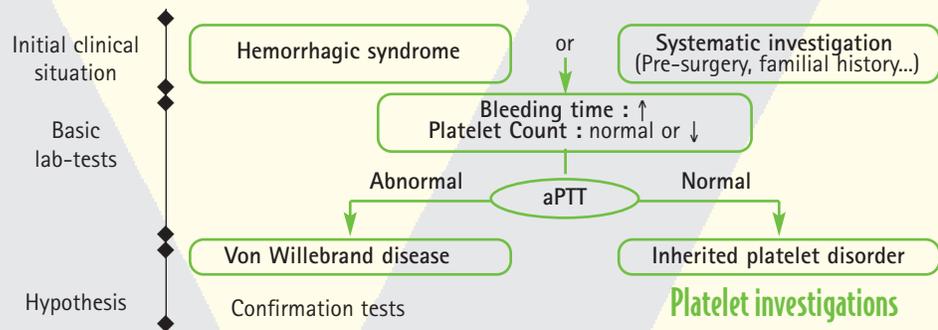


Adult normal range of platelet glycoprotein expression (n=40) :

	Basal state	TRAP activation
GMP 140 (CD62P)	< 1 000	\geq 1 000
GpIIb (CD41)	51000 +/- 14000	85000 +/- 27000
GpIb α (CD42b)	38000 +/- 11000	19000 +/- 10000

Results expressed in sABC
Normal values have to be determined according to the age⁽⁹⁾.

Diagnosis algorithm of inherited platelet disorders :



Example of inherited thrombopathy tested with *PLT* Gp/Receptors :

