Glanzmann's Thrombastenia Treatment:

A Center's Experience

INTRODUCTION

- Glanzmann's Thrombastenia is a rare inherited bleeding disorder, caused by a deficiency/dysfunction in the glycoprotein GP IIb/IIIa receptor on platelets
- Patients with multiple platelet transfusions can develop antibodies against GP IIb/IIIa that may compromise the efficacy of subsequent transfusions

PATIENTS

- 5 Caucasian women; 19-45 Years-old
- Gp IIb/IIIa defect: 2 – 32 %
- Comum clinical manifestations: ecchymosis, epistaxis, gum bleeding, menorhagia, abnormal bleeding with trauma, surgery or dental treatments

BLEEDING EPISODES AND ITS TREATMENT

< 1990

PLATELET AND ERYTHROCITE TRANSFUSIONS

1990 - 2000

12 episodes

- ✓ 9 dental treatments
- ✓ 2 uterine curetage
- ✓ 1 hematoma

DESMOPRESSIN ACETATE

Dose: 0.3 µg/kg

10 episodes: uncomplet bleeding control

2 episodes without success

2 adverse events

> 2000

16 episodes

- ✓ 12 dental treatments
- ✓ 2 oral surgeries
- ✓ 1 eutocic delivery
- ✓ 1 epistaxis

rFVIIa

Dose: 90-120 µg/kg

All patients successfully treated

No adverse events

Also used

- Local measures (ex. thrombin glue in dental extractions)
- EACA
- Oral contraceptives
- Endovenous iron for aenemia

CONCLUSIONS

- In most patients the severity of blood loss was unpredictable and not related neither with the type of bleed nor with the severity of GP IIb/IIIa defect.
- Four patients developed platelet antibodies.

- In the 90's we empirically used desmopressine acetate in patients with platelet antibodies without satisfactory bleeding control.
- Today we consider rFVIIa as the treatment of choice in all patients.
- We had no adverse events using rFVIIa.

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rFVIIa: activated recombinant factor VII; EACA: acid ε-aminocaproic