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, menorrhagia, abnormal bleeding with trauma, surgery or dental treatments.

BLEEDING EPISODES AND ITS TREATMENT

- PLATELET AND ERYTHROCYTE TRANSFUSIONS
  - 1990:
    - 12 episodes
    - 9 dental treatments
    - 2 uterine curetage
    - 1 hematoma
  - 2000:
    - 16 episodes
    - 12 dental treatments
    - 2 oral surgeries
    - 1 eutocic delivery
    - 1 epistaxis

- DESMOPRESSIN ACETATE
  - Dose: 0.3 µg/kg
  - 10 episodes: uncomplet bleeding control
  - 2 episodes without success
  - 2 adverse events

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

- rFVIIa
  - Dose: 90-120 µg/kg
  - All patients successfully treated
  - No adverse events

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 desmopressin 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.