

Glanzmann's Thrombastenia Treatment:

A Center's Experience

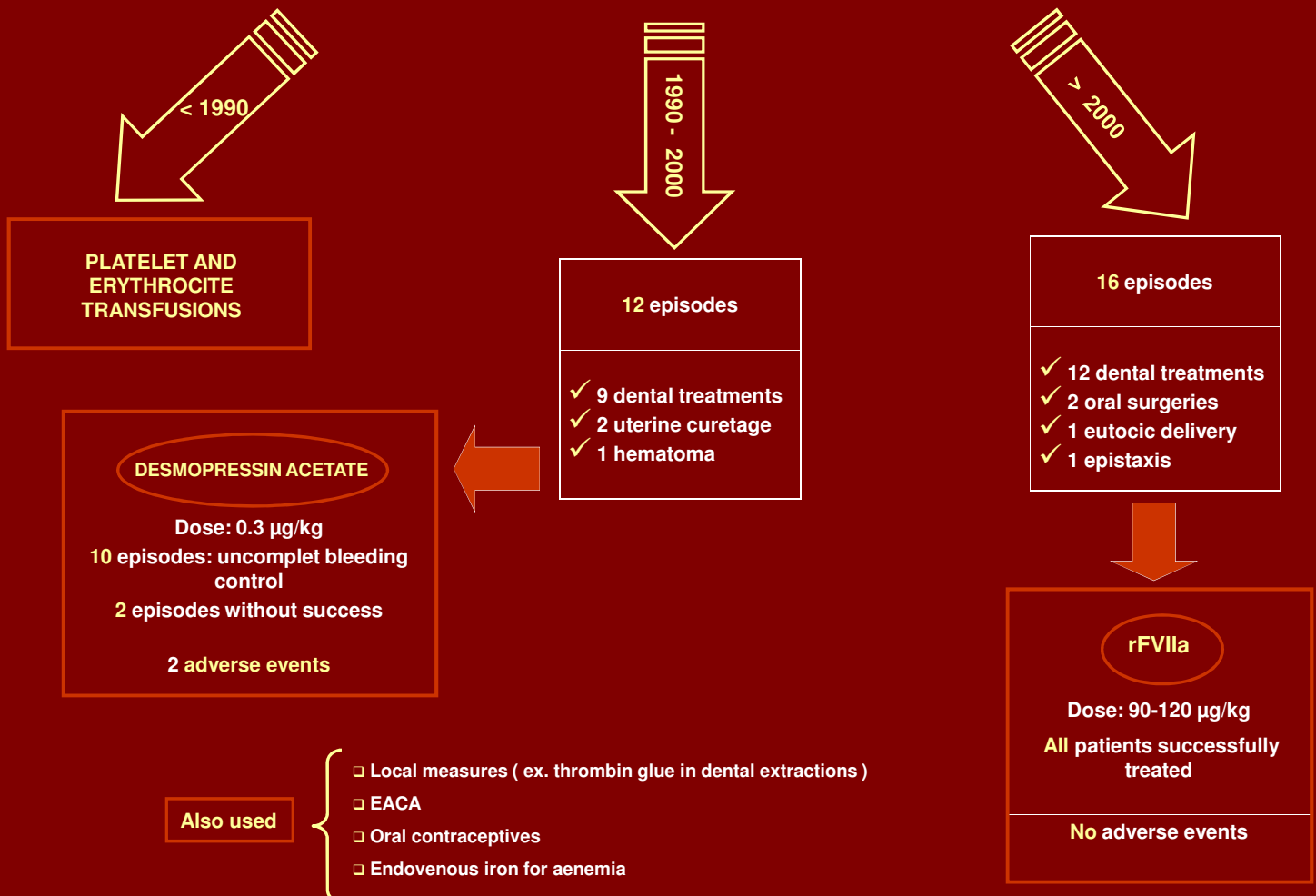
INTRODUCTION

- 🔗 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 %
- 🔗 Common clinical manifestations: ecchymosis, epistaxis, gum bleeding, menorrhagia, abnormal bleeding with trauma, surgery or dental treatments

BLEEDING EPISODES AND ITS TREATMENT



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.