

Glanzmann's Thrombastenia Treatment:

A Center's Experience

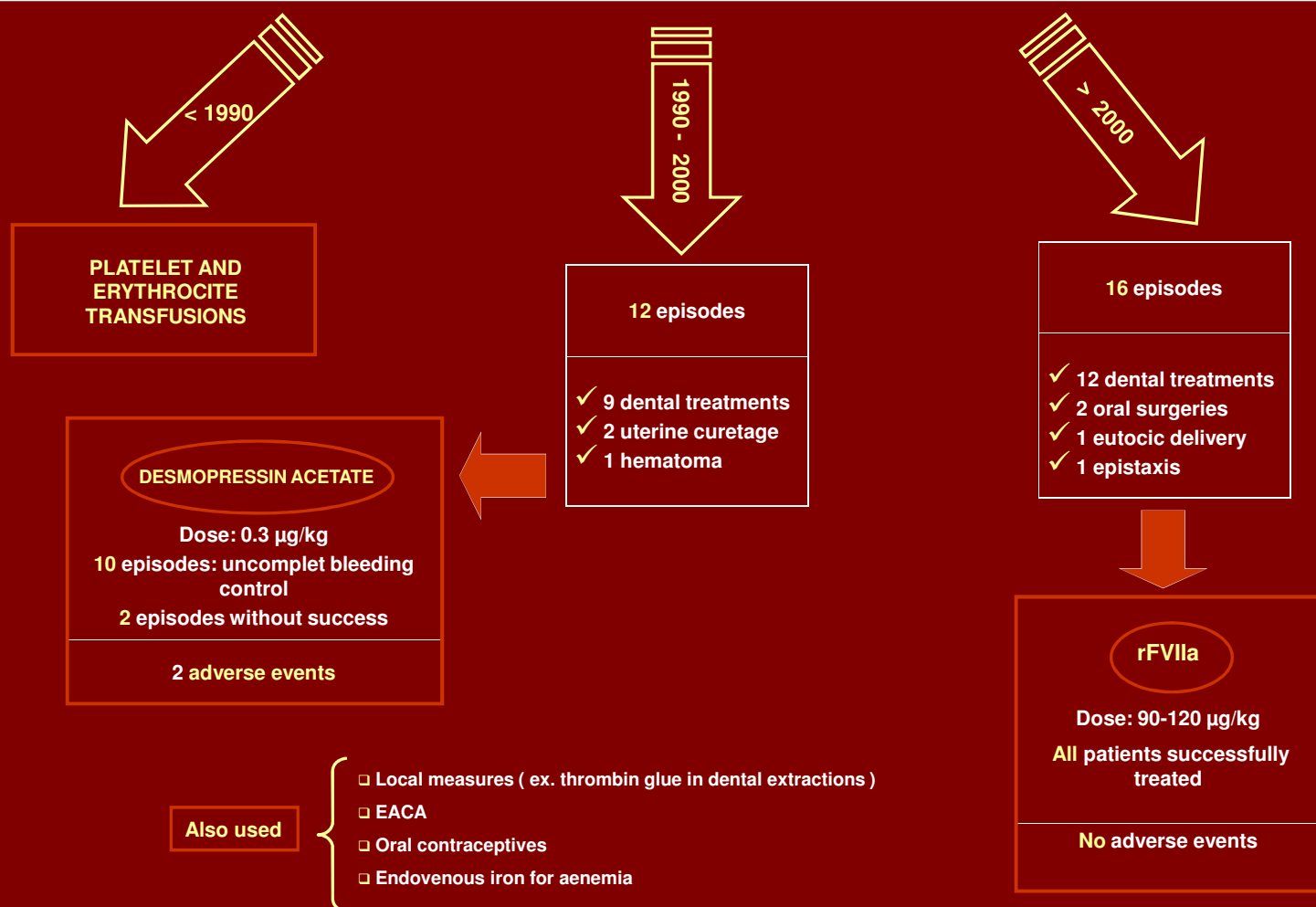
INTRODUCTION

- 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.