

# Successful Secondary Prophylaxis in Haemophilia with Inhibitors using rFVIIa Three Times per Week

M Martinez,\* P Do Nascimento, † G. Arbesu, ‡ S Gastaldo, ¶ S Borchichi †† and R Perez Bianco \*\*

\*Hematology Department, Hospital de Niños "Sor Maria Ludovica", La Plata, Argentina; †Department of Haemophilia, Instituto William Osler, Buenos Aires; ‡Hematology Department, Hospital Pediátrico "Humberto Notti", Mendoza; ¶Hematology Department, Hospital de Niños "Dr Hector Quintana", San Salvador de Jujuy; ††Pediatric Hemato-Oncology Department, Hospital Pediátrico "Fernando Barreyro", Posadas; and \*\*Instituto de Investigaciones Hematológicas, Academia Nacional de Medicina, Buenos Aires, **Argentina.**



## INTRODUCTION

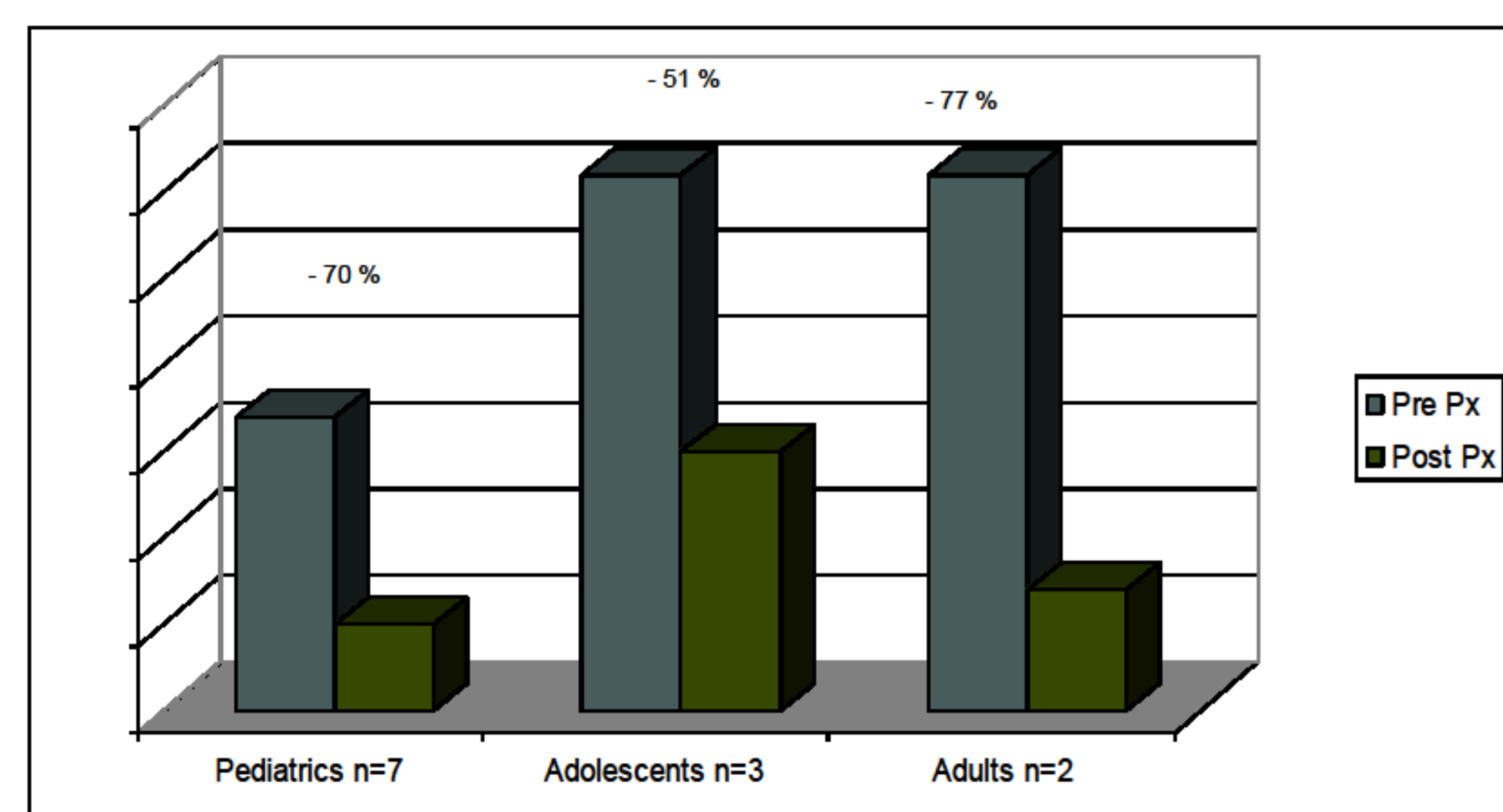
Patient with severe hemophilia develop severe bleeding complications. From the early studies (Nisson and Manco-Johnson) prophylaxis has been shown to be effective in reducing bleeding frequency and hospitalization vs. Conventional on-demand therapy. Inhibitors occur in 20-30% of patients with severe hemophilia A but in only 5% of patients with hemophilia B.

Since the introduction of prophylaxis with rFVIIa there have been numerous reports of success with different regimes. In 2010, an expert consensus from the Argentinean Haemophilia Foundation published a recommendation of 90 µg/kg three times per week as the initial dose and to escalate dosage according to the response obtained. Here, we report the experience of 5 centers with secondary prophylaxis using rFVIIa three times/week.

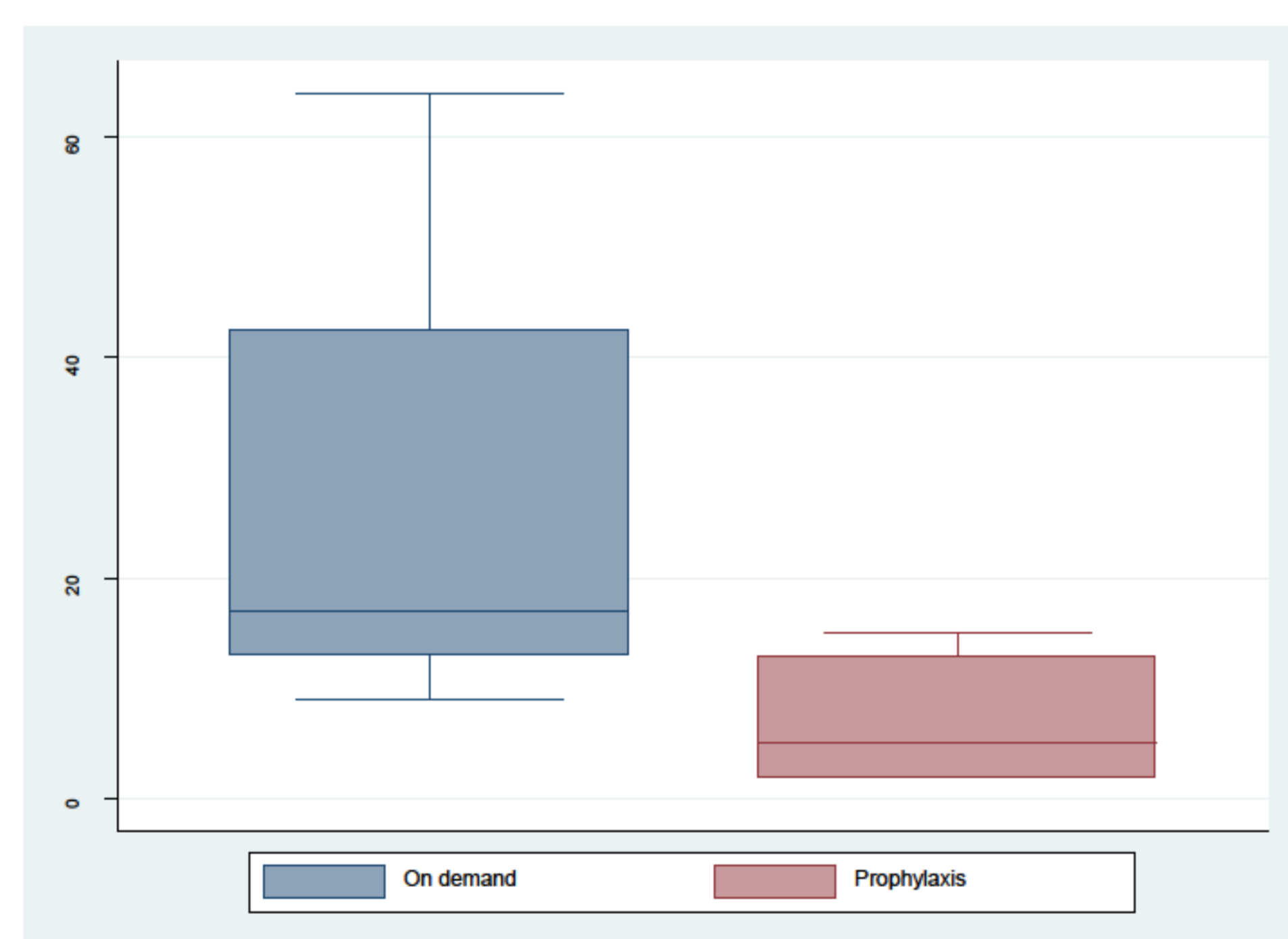
## PATIENTS AND METHODS

The study was an observational retrospective study. Twelve patients (11HA and 1 HB), mean age 12 years old (r 2- 43y), from a five different hospitals received prophylaxis therapy with rFVIIa thrice a week. The indication for prophylaxis was: recurrent bleeding episodes in 9 patients and CNS bleeding in the remaining three. All patients were initially put on 90µg/kg three times per week. 9/11 continued with this dosing during the follow up period, and 2 had the dose increased to 270 µg/kg in order to reduce the number of bleeding episodes. Patients were followed for a mean of 21 months (4-40) after the initiation of prophylactic therapy. To assess the efficacy of the thrice weekly schedules we compared the number of bleeding episodes and the admission days before and after prophylaxis was instituted.

Median number of bleeds by age



Mean number of bleeding episodes



## Inhibitors

Number of FVIII exposure days median 37.5 (r 2-73)  
Peak historical inhibitor titre (BU) mean 685 (r 4-5700)

## RESULTS

The mean number of bleeding episodes was reduced from 26 (range 9-64) for on-demand therapy to 7 (range 2-15) for prophylaxis (p: 0.003). As a group, this represented an overall reduction of 63 % of bleeding episodes. When we analyzed the need for hospitalization, the mean admission days were 6.92 (range 0-7) while on demand therapy was reduced to 0.77 days (range 0-7) on prophylaxis (p: 0.014). It is noteworthy that no thromboembolic events were detected despite the prolonged and continued exposure to rFVIIa.

## References

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## CONCLUSIONS

Our results suggest that prophylaxis with rFVIIa three times per week effectively reduces bleeding episodes in inhibitor patients. Furthermore, this reduction was accompanied by a significant reduction in admission days which can be considered an improvement in the quality of life for these patients.

