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INTRODUCTION AND OBJECTIVES

Primary prophylaxis is recommended as the first choice of treatment for people with severe hemophilia. It prevents bleeding and joint destruction, and can preserve normal musculoskeletal function. Delayed prophylaxis is able to decrease frequency of bleeding and may slow progression of joint disease and improve quality of life, but it does not reverse established joint damage. We describe the experience of tertiary prophylaxis started in adults in our service.

METHODS

Adults with severe hemophilia, without inhibitors, who had bled frequently or had severe bleeding episode and thus were switched from on-demand to prophylactic treatment (for at least 45 week/y) after the age of 18 years were assessed. The period of time the patient received prophylaxis was compared to the same amount of time previous to the initiation of prophylactic treatment (~15 UI kg⁻¹ thrice weekly). Differences in annual number of total and joint bleeds (ABR and AJBR, respectively), number of life-threatening bleeding and factor utilization (UI/Kg/y) were assessed comparing the period before and after tertiary prophylaxis, in each patient.

RESULTS

Twenty-five patients with hemophilia A (n=23) and B (n=2) were evaluated. The median age at start of prophylaxis was 26 y (18 – 57 y) and the median duration of prophylaxis was 33 months (13 – 56 mo). Twenty-four patients (96%) had arthropathy in at least one joint before prophylaxis.

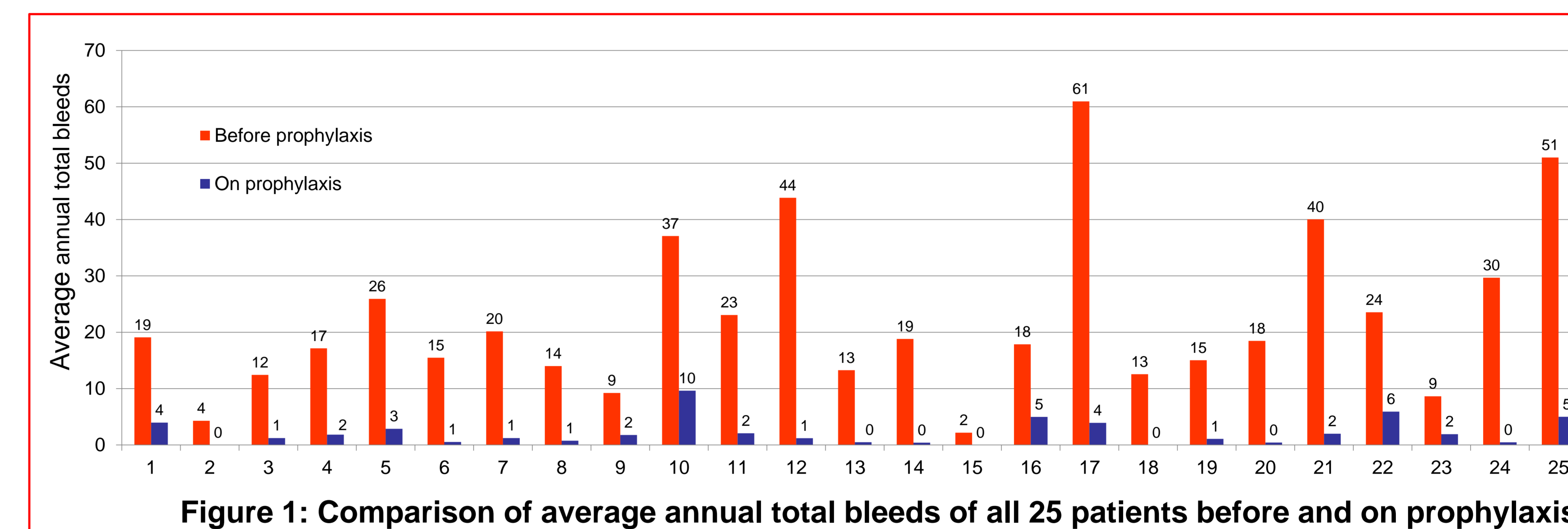
Tertiary prophylaxis reduced the mean ABR and AJBR ($P < 0.0001$). *Table 1, Fig. 1.* Eight episodes of life-threatening bleeds occurred in 6 patients before prophylaxis and none after. On the other hand, there was higher factor concentrate consumption during prophylaxis ($P < 0.0001$). *Table 1*

Many patients were treated with short-term prophylaxis during the period before tertiary prophylaxis.

Table 1: Comparison of bleeding episodes and factor usage before and on prophylaxis in 25 patients

	Before	On-prophylaxis	P value
Mean ABR* (range)	22.1	2.1	$P < 0.0001$
Mean AJBR' (range)	15.3	1.2	$P < 0.0001$
Total number of life-threatening bleeding	8	0	
Mean annual factor used (UI/kg/y)	1.240	3.043	$P < 0.0001$

* ABR = annual number of total bleeds ; 'AJBR = annual number of joint bleeds



CONCLUSIONS

In our country, primary prophylaxis only started in 2011 and many of our adult patients have joint sequelae. Long-term prophylaxis starting in adulthood has been associated with a reduction of more than 90% of bleeding episodes with an increase of 2.5 times of factor consumption. Despite the challenges, tertiary prophylaxis should be considered for those adults with active bleeding patterns while receiving on-demand treatment.

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