Evaluation of the PFA-100 system for monitoring desmopressin therapy in patients with type 1 von Willebrand's disease

The platelet function analyzer (PFA-100) is a rapid and simple in vitro system for evaluating primary hemostasis. In this report we show the efficacy of this method for monitoring the response to desmopressin therapy in 24 patients with type 1 von Willebrand's disease.

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The platelet function analyzer PFA-100 (Dade International Inc., FL, USA) is a relatively novel method for rapid in vitro global evaluation of primary hemostasis.[1] Since the PFA-100 has been shown to be very sensitive to platelet and von Willebrand factor (VWF) levels and function, it is useful mainly as a screening test for von Willebrand's disease (VWD) and several platelet disorders.[2,3] Recently, the PFA-100 has been proposed for therapeutic monitoring of patients with VWD treated with desmopressin (DDAVP) or factor VIII/VWF concentrate.[4,5] In the present study, we evaluated the PFA-100 system in 24 patients with type 1 VWD, before and 1 hour after the subcutaneous injection of DDAVP at a dose of 0.3 µg/Kg. The median age of the 24 patients was 34.2 years (range 21 to 43 years) and the male/female ratio was 1.2.

Closure time (CT) on the PFA-100 system was measured with both collagen/ADP (CT-ADP) and collagen/epinephrine (CT-Epi) cartridges. We also tested the effects of the DDAVP test on plasma concentrations of VWF antigen (VWF:Ag), VWF activity (VWF:ristocetin cofactor, RCo), coagulation factor VIII (F VIII:C) and bleeding time (BT) according to Ivy. The results are shown in Table 1. All patients had prolonged APTT and CT with both cartridges and decreased VWF:Ag and VWF:RCo levels at baseline. The baseline factor VIII levels were decreased and the bleeding time was prolonged in 10 (41.7%) and 19 (79.2%) patients, respectively. Subcutaneous DDAVP injection induced normalization of VWF:Ag and VWF:RCo in all patients with a mean increase over the baseline of 2.8 (range 1.4-3.8) and 2.9 (range 1.6-4.2) times, respectively. The mean factor VIII increase over the baseline was 2.5 (range 1.2-3.6) times. Desmopressin also shortened BT and CT with both types of cartridges in all patients. As observed in previous reports,[4,5] normalization of PFA-100 results post-DDAVP correlated well with the increase in VWF. In our study, the diagnostic sensitivity of the PFA-100 system for patients with type 1 VWD was greater than that of the BT. In fact, all 24 patients (100%) with type 1 VWD had abnormal basal CT as measured with both collagen/ADP and collagen/epinephrine cartridges, whereas a prolonged BT was recorded in 19 patients (79.2%, p <0.001). Therefore, our results substantially agree with earlier ones[4,5] and confirm that the PFA-100 system might be a useful tool for diagnosis and therapeutic monitoring of patients with VWD. Moreover, since the PFA-100 system appears to be more sensitive and less invasive, we believe that it could represent a valid and reliable alternative to BT as a screening test for type 1 von Willebrand's disease.[6]

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Table 1. Mean values and ranges of hemostatic parameters at baseline and 1 hour after s.c. injection of DDAVP (0.3 µg/Kg) in 24 patients with type 1 von Willebrand's disease.

Parameter	*Baseline*	*1 hour after DDAVP*
APTT (sec)	41.1 ± 4.3 (36.3-48.7)	32.5 ± 3.6 (26.1-36.0)
VWF:Ag (IU/dL)	42.7±8.1 (29-57)	121.2 ± 26.8 (89-230)
VWF:RCo (IU/dL)	39.6 ± 7.6 (26-49)	116.7 ± 21.3 (81-205)
F VIII:C (IU/dL)	62.1 ± 12.4 (44-83)	153.2 ± 17.8 (114-270)
BT (min)	9.2 ± 2.8 (5-13)	4.8 ± 1.9 (3-6)
PFA-100 (sec)		
Collagen-ADP	136.8 ± 26.7 (112-193)	67.4 ± 25.7 (42-93)
Collagen-Epi	168.1 ± 33.3 (148-250)	88.0 ± 19.6 (51-124)

Values are given as mean ± SD (range). Normal reference values: APTT = 24.2-36.0 sec; FVIII:C = 50-150 UI/dL; VWF:Ag = 60-150 UI/dL; VWF:RCo = 50-150 UI/dL; BT = 2-6 min; CT-ADP = <110 sec; CT-Epi = <140 sec.

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