Severe Aortic Valve Stenosis: Sustained Cure of Acquired von Willebrand Syndrome After Surgical Valve Replacement

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Aortic valve stenosis (AVS) is the most common valve disease in adults. Severe forms are associated with acquired von Willebrand syndrome (aVWS) with loss of the largest von Willebrand factor (VWF) multimers. Diagnostic gold standard is the VWF multimer analysis. Valve replacement rapidly restores the VWF structure. Uncertainty exists if this effect is permanent and how functional VWF assays perform compared with multimer analysis. We studied 21 consecutive patients with severe AVS before and 6 to 18 months after valve surgery and compared them with 14 controls without valve disease referred for coronary angiography. The VWF multimers, VWF antigen (VWF:Ag), VWF collagen binding capacity (VWF:CB), VWF:CB/VWF:Ag ratio, in vitro bleeding time (PFA-100), factor VIII coagulation activity (FVIII:C), and VWF ristocetin cofactor activity (VWF:RCo) were determined. In all patients with AVS, the large VWF multimers were strongly reduced (56 ± 13% of normal plasma); all controls had normal multimers. The PFA-100 collagen/ADP closure times (coll/ADP CT) were prolonged in patients with AVS compared with the controls (175 ± 56 seconds vs 86 ± 14 seconds, P < .001). The VWF:CB/VWF:Ag ratio was pathological in 20 of the 21 patients but normal in controls. After surgery, the multimers normalized in all patients and coll/ADP CT shortened (pre 184 ± 65 seconds vs post 102 ± 22 seconds; P < .001). The VWF:CB/VWF:Ag ratio strongly improved (P < .001) and normalized in 14 of 17 patients. In conclusion, all consecutive patients with severe AVS had an aVWS. The combination of coll/ADP CT and VWF:CB/VWF:Ag ratio detected the aVWS in all patients. More than 6 months after valve replacement, the VWF multimers were still normalized in all patients indicating a permanent cure of the aVWS.

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