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A Successful Coronary Artery Bypass Operation with Intermittent Factor VIII Administration in a Hemophilia A Patient who Applied with Acute Myocardial Infarction: a rare and difficult case

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There is not a large study in the literature other than a few case report which express opinion, a few case reports and reviews about procedure of coronary artery bypass grafting (CABG) surgery planning for patients with hemophilia. An internationally accepted, definitive algorithm that recommends an approach to these patients wasn't included in the guidelines.

A 51-year-old male patient applied to emergency service with sudden and severe chest pain. He had no other medical history except Hemophilia A. His electrocardiographic findings showed ST elevation in DII, DII and aVF derivations. Troponin T: 0.19 ng/mL (0-0.1) level was accepted as positive. Patient was admitted to coronary intensive care unit with an initial diagnosis of acute inferior myocardial infarct and coronary angiography was performed urgently. Angiography revealed a moderate left ventricular ejection fraction (LVEF: 49%) with three occluded coronary arteries. Left anterior descending artery (LAD) was critically stenotic up to 80%. Right coronary artery (RCA) was stenotic up to 50%. Circumflex coronary artery (Cx) was also stenotic up to 90% (Figure). Patient received 50 U/kg (4000 U) FVIII after angiography, which he was not able to receive before angiography due to urgency of the case. Thereafter, he received 25 U/kg (2000) FVIII twice a day for 3 days. 20 U/kg (1600 U) FVIII was replaced for the following 7 days at intervals of 12 hours. No intervention was performed during angiography because of multi-vessel disease and decided on bypass operation. Among blood parameters tested during admission of the patient, activated partial thromboplastin time (aPTT) was 51,4. FVIII inhibitor identification test resulted negative. His childhood FVIII levels were 11,3, thus he was evaluated as a mild case of hemophilia A.

In the morning of by-pass operation, patient received 50 U/kg (4000 U) FVIII replacement and was taken to operation with an aPTT level of 45,6. Bypass operation was carried out with same procedures as non-hemophilia patients including standard heparinization. In order to prevent disseminated intravascular coagulation during factor replacements of the patient, heparin was not used except pump procedure. After patient was weaned from cardiopulmonary pump 50 U/kg (4000 U) bolus FVIII was administered. For the following 3 days, 25 U/kg (2000 U) FVIII was administered at intervals of 12 hours. Thereafter, 20 U/kg (1600 U) FVIII was administered for 7 days at intervals of 12 hours (Table). Patient has been

followed up for 3 years with routine controls. Within this period, he had no serious medical problem except nose-bleeds.

When the literature was analyzed, it was identified that continued infusion of FVIII was rarely administered in pre- and intra-operative period [1,2]. Similar to the literature, we didn't administer continued infusion of FVIII. Because we believe thrombosis risk is more of an issue comparing to the bleeding.

World Hemophilia Federation recommend FVIII levels between 80% and 100% before and after major operations [3]; but considering urgency and thrombosis risk in our case we brought a different approach as all disciplines responsible for the case and arrived at a consensus. We present our method in the table as a recommendation. In this method, we administered 2x50 U/kg on the day of operation (1 day), 2x25 U/kg for the following 3 days, 2x20 U/kg for the following 7 days and named it "1-3-7 protocol". Our protocol needs to be tested with further studies.

References:

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Figure: Stenosis. RCA 50%, LAD 80% and Cx 90%.

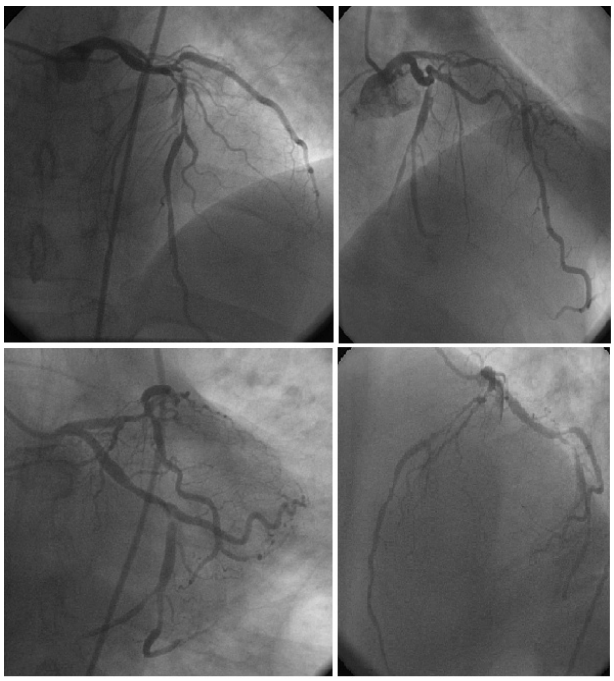

Table: Daily total dosage of FVIII, evaluation of aPTT and FVIII during the perioperative period.

Day	aPTT	FVIII	FVIII dosage (daily)
0 (angiography)	51,4		1x4000 U
1-3			2x2000 U
4	49,1	14,5	2x1600 U
5-10			2x1600 U
operation decision and surgery preparations			
25 (bypass)	45,6		2x4000 U
26-28			2x2000 U
29	40,5	29,8	2x1600 U
30-35			2x1600 U
exit from intensive care			
36 (nose-bleed)	39,9		2x2000 U
37	35,5		2x2000 U
38	34,0		2x2000 U
discharge preparations and warnings			
39	33,1		none
40	33,4		none
41 (discharge)	36,1	53,7	none