



CORONARY ARTERY BYPASS SURGERY IN PATIENTS WITH HAEMOPHILIA B

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Introduction and Objective:

- The life expectancy of patients with haemophilia B (PWH-B) has increased after the induction of factor-IX concentrates (FIXc).
- Age-related conditions including cardiovascular disease have become even more common.
- Despite the hypocoagulable state it is generally accepted that PWH-B are not protected against atherosclerosis.
- Similarly to the recommended approach in the general population, coronary artery bypass (CABG) surgery should be proposed when another approach is not indicated.

Materials and Methods:

- A retrospective analysis of the patients with PWH-B in a single haemophilia department who underwent CABG was performed and review of literature was made to identify additional cases.

Results:

- The analysis revealed a man with mild haemophilia B (3.7% of all PWH-B). He has rarely been treated with recombinant-FIXc (rFIXc) for bleeding episodes. He had cardiovascular risk factors including smoking and obesity and at the age of 70 years he successfully underwent a 3-vessel CABG.
- Preoperatively a bolus infusion of rFIXc was given in order to achieve FIX levels about 100%. Full dose heparinization was administered for cardiopulmonary bypass (CPB) and satisfactory haemostasis had been achieved without any supplementary rFIXc and no antifibrinolytics were given as an adjuvant treatment.
- During the postoperative period there were no complications.
- Review of the literature revealed 13 additional cases of PWH-B. The patients' characteristics and replacement therapy both preoperatively and at the postoperative period are depicted in Table-1.

Table-1. Review of reported cases of patients with haemophilia B who underwent CABG

REFERENCE	SEVERITY	AGE (years)	RISK FACTORS	F-IX TARGET LEVELS	REPLACEMENT THERAPY	COMPLICATIONS
Suzuki T et al. (2016)	Moderate	52	No data	Pre.: 100%, Post.: 80-100%	Pre.: Bolus rFIXc , Intra./Post: Continuous rFIXc for 8 days	None
Shalabi M et al. (2015)	No data	55	No data		Continuous FVIIIc	None
Bhave et al. (2015)	Mild		No data	0.8-1 IU/ml days 1-5, 0.6-0.8 days 6-10	Pre.: No replacement, Intra./Post.: Bolus rFIXc after termination CPB for 10 days	None
Varzaly J et al. (2013)	Mild	59	No data		Continuous rFIXc	None
	Female Carrier	79	No data		Pre/Intra: Continuous rFIXc, Post: Continuous rFIXc	None
Pesaro et al. (2009)	Severe	37	HIV	Pre: 100%, Post: 20-40%	Pre.: Bolus FIXc, Intra./Post.: Continuous FIXc for 12 days	Vein-graft occlusion/ PTCA
Grandmougin et al. (2005)	Severe	52	HCV, Diabetes	Pre: ≥100%	Pre.: Bolus pdFIXc , Intra./Post.: Continuous pdFIXc 450 IU/h for 7 days then bolus pdFIXc	None
Donahue et al. (1999)	Moderate	80	No data	Pre.: 100%, Post.: 50-150%	Pre./Intra. Bolus rFIXc, Post.: Bolus rFIXc Antifibrinolytics: Aminocaproic acid	Aortic dissection / Death
Palanzo et al. (1995)	Moderate	71	Diabetes	>80%	Pre.: Bolus pdFIXc, Intra./Post.: Continuous pdFIXc 50ml/h, bolus after termination CPB for 6 days, Antifibrinolytics: Aprotinin	Transient atrial fibrillation
Scharfman et al. (1993)	Moderate	71	Obesity	Pre: 100% Post: 50%	Pre.: Bolus pdFIX PCC , Post.: Bolus pdFIX PCC for 7 days Antifibrinolytics: Aminocaproic acid	None
Wilson et al. (1991)	Mild	74	Diabetes, smoking, hypertension	Pre: ≥60%	Pre/Intra: Bolus pdFIX PCC (60IU/Kg) , Post: Bolus pdFIX PCC (10IU/ Kg) every 12h for 7-10 days	None
Raish et al. (1985)	Moderate	59	No data	Pre.: 50%	Pre.: Plasmapheresis/FFP, Post: FFP	None
Tourbaf et al. (1979)	Moderate	40	Smoking, family history	Pre.: 80%	Pre: FIXc PCC, Intra: FIXc + FFP, Post: FIXc for 13days	None

CPB: Cardiopulmonary bypass, F-IX: Factor IX, FIXc: Factor IX concentrate, FFP: Fresh frozen plasma, HCV: Hepatitis C, HIV: Human immunodeficiency virus, Intra.: Intraoperative, pdFIXc: plasma-derived factor IX concentrate, PCC: Prothrombin complex concentrate, Post.: Post-operative, Pre.: Pre-operative, PTCA: Percutaneous Transluminal Coronary Angioplasty, rFIXc: recombinant factor IX concentrate

Conclusions:

- Cardiac surgery is challenging in PWH-B.
- Best results appear to be achieved with a multidisciplinary team approach in a hospital with a haemophilia unit.
- Current treatment strategies are often based on local clinical experience and adaptation of general guidelines used in the non-haemophilic population.