

Using BeneFIX for Low-Dose Surgical Prevention In Chinese Patient with Hemophilia B: a Case Report

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Objectives:

Replacement treatment in China is usually inadequate for poor patient affordability¹. BeneFIX is the only FIX concentrates for hemophilia B in China, which has been well confirmed its effects and safety²⁻⁸. Here we report the first case of an 83-year old patient with hemophilia B who underwent roux-en-y hepaticojejunostomy managed successfully with BeneFIX for low-dose surgical prevention in China.

Methods:

Blood tests showed a prothrombin time (PT) of 13.2s and plasma level of FIX of 7%. Factor desired level was set to 40%. BeneFIX was given 45IU/Kg with frozen plasma 200mL and VK1 30mL as adjuvant therapy 3 days before and after surgical procedure (Table 1). Haemostasis response was assessed by surgeon peri- and post-operation.

Table1. Prevention regimen during roux-en-y hepaticojejunostomy operation

Days from operation	BeneFIX (IU/Kg)	Frozen plasma (mL)	VK1 (mL)
-3	45	200	30
-2	45	200	30
-1	45	200	30
0	45	200	30
+1	45	200	30
+2	45	200	30
+3	45	200	30

Results:

The surgery was successful. Peri- and post-operative estimated blood loss was similar to that expected in non-haemophilic individuals, and haemostasis was rated as excellent. No additional dose of BeneFIX or other coagulation products were used. BeneFIX was well tolerated, with no thromboembolic events and FIX inhibitor development.

Conclusions:

This case showed low dose surgical prevention with BeneFIX was effective and safe in achieving haemostasis in subjects with Hemophilia undergoing surgery in China.

Reference:

- Xue F, Zhou ZP, Yang RC. Survey of medical care and prognosis in patients with severe hemophilia A from certain cities in China [J]. Chinese Journal of Hematology, 2011, 32(7): 481-483.
- Lambert T, Recht M, Valentino LA, et al. Reformulated BeneFix: efficacy and safety in previously treated patients with moderately severe to severe haemophilia B [J]. Haemophilia, 2007, 13(3):233-243.
- Monahan PE, Liesner R, Sullivan ST, et al. Safety and efficacy of investigator- prescribed BeneFIX prophylaxis in children less than 6 years of age with severe haemophilia B [J]. Haemophilia, 2010, 16(3): 460-468.
- Shapiro AD, Di Paola J, Cohen A, et al. The safety and efficacy of recombinant human blood coagulation factor IX in previously untreated patients with severe or moderately severe hemophilia B [J]. Blood, 2005, 15, 105(2): 518-525.
- Ragni MV, Pasi KJ, White GC, et al. Use of recombinant factor IX in subjects with haemophilia B undergoing surgery [J]. Haemophilia, 2002, 8(2): 91-97.
- Windyga J, Rusen L, Gruppo R, et al. BDDrFVIII (Moroctocog alfa[AF-CC]) for surgical haemostasis in patients with Haemophilia A; results of a pivotal study [J]. Haemophilia, 2010, 1, 16(5): 731-739.
- Recht M, Nemes L, Matysiak M, et al. Clinical evaluation of moroctocog alfa (AF-CC), a new generation of B-domain deleted recombinant factor VIII (BDDrFVIII) for treatment of Haemophilia A: demonstration of safety, efficacy, and pharmacokinetic equivalence to full-length recombinant factor VIII [J]. Haemophilia, 2009, 15(4): 869-880.
- Lusher JM, Lee CA, Kessler CM, et al. The safety and efficacy of B-domain deleted recombinant factor VIII concentrate in patient with severe haemophilia A [J]. Haemophilia, 2003, 9(1): 38-49.

