

**FIX Subcommittee  
Annual Report to  
AHCDC Annual Meeting May 2005**

1. One of the questions raised in the last AHCDC AGM meeting was whether hemophilia A and hemophilia B at equivalent severity as defined by clotting factor activity have similar clinical manifestations. The Hamilton group has conducted a retrospective review of bleed frequency and factor concentrate usage records of their hemophilia A and B patients. As indicated in the abstract (for ISTH Congress, 2005) below, Hemophilia B patients compared to Hemophilia A patient, appeared to have fewer bleeding episodes, although the clotting factor concentrate usage was not dissimilar. The number of patients included, particularly for Hemophilia B, is limited. Dr. Mohan Pai will present the data for discussion if the study should expand to include other centers as a FIX Subcommittee activity to study this question further.

**Abstract (ISTH Congress 2005)**

**Comparing Bleed Frequency and Factor Concentrate Use Between Haemophilia A and Haemophilia B**

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Hemorrhagic manifestations in patients with Hemophilia A and B are considered identical for comparable level of factor deficiency. We investigated whether patients with severe and moderate FVIII deficiency (HA) bleed more frequently and utilize more clotting factor than those patients with comparable severities of FIX deficiency (HB). With the computerized data collection system (Transfusion 2003;43:556) we collected data on frequency of bleeds and factor concentrate utilization over three years, from 01/2001 to 12/2003. Fifty-eight patients with severe FVIII(<0.01U/ml), 10 out of 15 moderate FVIII(<0.05U/ml), 15 out of 17 severe FIX, and 5 out of 10 moderate FIX deficient patients required treatment for episodic bleeds, postoperative haemostasis and for primary or secondary prophylaxis. A total of 21,363,409 IU of recombinant FVIII (rFVIII) was used by patients with HA (104,722 IU/patient/year) and 6,430,960 IU of recombinant factor IX, (rFIX) BeneFIX® by patients with HB (107,722 IU per patient/year). Adjusting for the difference in in-vivo recovery of factor concentrates (rFVIII has a recovery of 2.1% per IU/kg and rFIX has recovery of 0.75% per IU/kg) HB patients would have received a normalized amount of 84,745U/patient/year. HA patients bled more frequently than HB patients, 13.9 vs 9.1 bleeds/patient/year, but used similar amounts of concentrate per bleed, 1613 IU rFVIII vs 2538 IU rFIX. The difference in usage of factor concentrate is not statistically significant (p.>0.05). Thus, while HA and HB patients used similar amounts of factor concentrate, HB patients had 35% less bleeds than HA group. A decreased bleed frequency in patients with HB needs to be confirmed, particularly in young children for whom early institution of primary prophylaxis is increasingly being considered.

2. Dr. Poon's lab has developed a thrombin generation based-assay to detect FVIII activity below 1% for the purpose of studying heterogeneity of clinical manifestation in severe Hemophilia A. The development of a similar assay to detect FIX below 1% activity has now begun.