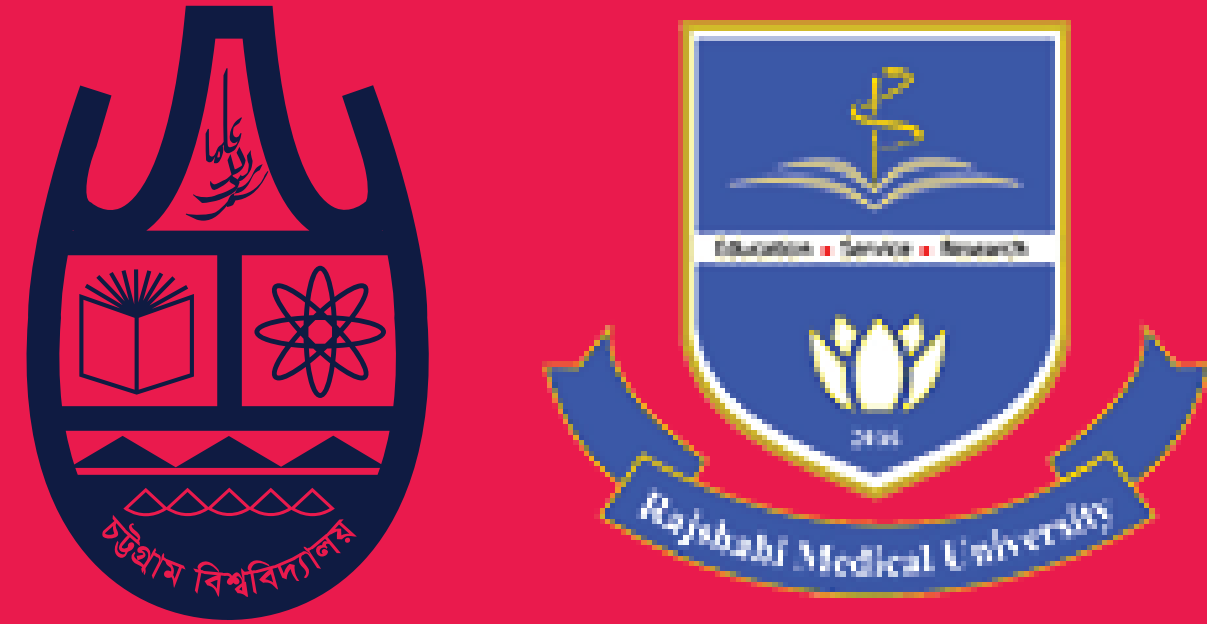


FFP INFUSION RISKS, LOW HAEM-A-QOL AND HEMOPHILICARTHROPATHY IN 32 HEMOPHILIA B PATIENTS WITH/WITHOUT INHIBITORS IN BANGLADESH: TOWARDS BETTER OUTCOMES

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BACKGROUND & AIMS

Background: Hemophilia B is a rare, inherited X-linked bleeding disorder caused by deficiency of clotting factor IX (FIX), resulting in recurrent bleeding and progressive joint damage [1,2]. In Bangladesh, recombinant FIX is largely unavailable, and most patients rely exclusively on Fresh Frozen Plasma (FFP) for on-demand therapy. Prophylaxis is virtually absent, and repeated plasma transfusions carry risks including allergic reactions, circulatory overload, and potential viral transmission [3,4].

Methods: We retrospectively analyzed 32 male patients with hemophilia B from across Bangladesh over a two-year period using structured questionnaires and registry review. Informed written consent was obtained from patients or their guardians where appropriate. Data collected included clinical severity, Annualized Bleeding Rate (ABR), Hemophilia Joint Health Score (HJHS), Quality of Life (QOL) [5], therapy type, inhibitor status, transfusion complications, viral exposure, and socioeconomic indicators. All procedures were conducted according to the Declaration of Helsinki [13].

Results: The median age was 10 years (range 3–45), with 59.4% severe, 28.1% moderate, and 12.5% mild disease. Median household income was 17,000 BDT (~150 USD), with 90.6% below the national median. FFP was the only on-demand therapy, with rare access to recombinant FIX via donations; none received prophylaxis. The mean ABR was 17.8, and inhibitor positivity (3.1%) was associated with higher bleeding rates. Target joints were present in 72%, mostly knees, and 61% had arthropathy (mean HJHS 12.4 ± 4.2), suggesting early and progressive joint deterioration. Functional impairment significantly impacted QOL, with 56% discontinuing school or employment. Transfusion-related complications occurred in 25% (mild allergic 12%, febrile 6%, severe anaphylaxis 3%, TACO 3%) [12]. No confirmed viral infections were reported, though risk remains high due to plasma dependence. Comparisons with other resource-limited settings demonstrate similar burdens [6–11], while therapeutic innovations elsewhere show potential for improved outcomes [7,14].

Conclusion: This largest dataset of FIX deficiency in Bangladesh highlights heavy reliance on FFP, frequent bleeding, progressive joint disease, and poor QOL, particularly among inhibitor-positive patients. The findings underscore an immediate need for structured hemophilia care—including prophylaxis, joint monitoring, inhibitor screening, patient education, and psychosocial support. Limited data and resource constraints call for investment in national registries, reliable recombinant FIX supply through HSB, and supportive infrastructure to reduce morbidity, prevent disability, and improve survival, aligning with SDG 3 (Good Health and Wellbeing) for better health and quality of life.

