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Managing hemophilia in underprivileged population

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A disease of royals with references to second century rabbinical rulings exempting boys from circumcision if two previous brothers had died of bleeding after the procedure, an Arab Physician Khalaf Ibn Abbass in his book "*Kitab al Tasrif*" or Methods in Medicine (936-1013 AD) documented an illness localized to males of a particular village where patients profusely bled after minor trauma. By 12th century, Hebrew physician Moses Maimonides recognized that these bleeding tendencies were transmitted through mother. The term Hemophilia, originated with a German Friedrich Hopff (1882) who coined the name "haemorrhaphilia" which later got abbreviated to hemophilia. Though hemophilia is often associated with family of Nicholas 11, the Tsar of Russia and Alexandra, granddaughter of Queen Victoria, but the bitter reality is that it occurs in all ethnic and racial groups throughout world. Hemophilia became known as royal disease since Queen Victoria was a carrier and passed this carrier status to many of her progeny daughters since consanguineous marriages was the order of day. History bears testimony to fact that Tsars of Russia fell apart because of preoccupation with management of their Hemophiliac son Alexei Nikolavich. Rusputin perhaps the infamous monk goes down in history with his role in management of Hemophilia. A sex linked recessive bleeding disorder classified as Hemophilia A with incidence of 1 per 5000 male births, Hemophilia B with 1 in 30,000 and estimated number of Hemophiliacs worldwide is 400,000. Classified as severe, moderate and mild depending on factor levels which respectively are <1, 1-5, 5-50 per pooled plasma. Although 80% of hemophiliacs live in developing world and out of 191 member states of W.H.O., 143 developing countries fall within Asia, Africa and South America, yet there is a significant lack of data as data collection is improper and perhaps does not represent true epidemiology. This is further compounded by interruptions in availability of factors and more often than not components in form of FFP, Cryoprecipitate are being relied upon. Despite available robust screening there is always risk of transmission of disease and in one of our recently concluded study we found 21.1% prevalence for HCV, 0.9% for HBV, 0.4% for HIV. Studies across globe report prevalence of HCV infection in Hemophiliacs from 7.5 to more than 50% and multi transfusions being single most risk factor. Inhibitors contribute to miseries as well. So do target joint arthropathies. In nutshell Comprehensive Hemophilia care centers, availability of recombinant factor concentrate with nucleic acid testing (NAT) of blood could change outlook of Hemophilia management in underprivileged populace.

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Purification of coagulation factor VIII by liquid chromatography

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Factor VIII (FVIII) is an essential coagulation factor in the blood, which is missing or defective in patients with Hemophilia A, a life-threatening blood disorder affecting one in 5000 males. Currently, replacement therapy with FVIII concentrates constitutes the basis for hemophilia care. Because of the high costs and immunogenicity of the recombinant product, there is still a demand for the plasma derived concentrate, especially for the developing countries. Brazil has an excess of plasma generated as a byproduct of the production of red blood cell concentrates that can be used as source of fractionated plasma products. In the Process Development Laboratory at Butantan Institute, we have been working in the purification of FVIII based in a methodology that avoids the cryoprecipitate. Fresh frozen plasma bags are thawed, pooled and after adjusting pH, plasma is directly loaded to an anion exchange column, which is the method of choice to capture this protein, because it is cost effective, easily scalable and purifications result in high FVIII recovery. A second purification step for the separation of the vitamin-K dependent proteins, a family containing several coagulation factors and inhibitors has been the focus of our interest. Due to the difference in size, these proteins could be separated from FVIII on a gel filtration column. Furthermore, the higher histidine residue content on the surface of FVIII allowed the full separation on IMAC (immobilized metal affinity chromatography) column with good yield.

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