CD4 & CD8 expression in haemophilia & von willebrand disease

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Aim of the work1- Study the value of CD4 % & CD8 % as a prognostic indicator ofcell mediated immunity.2- Determination of the risk factors of infection with Hepatitis C orHIV in patients with bleeding disorder.REVIEW OF LITERATURE4Definition:Hemophilia, which is derived from the Greek roots haima (blood) andphilein (to love), is a term that encompasses two congenital bleeding disorders.(Dunn&Thomas, 2004).Despite the inappropriateness of this description of excess bleeding, theword “hemophilia” continues to be used in patients with an inheritedpredisposition to blood loss. It is interesting that this inherited blood disease isnot mentioned in early Egyptian, Roman and Greek medical literature (Owen,2001).Haemophilia is inherited bleeding disorder caused by deficiency ofspecific coagulation factors. Haemophilia A is caused by coagulation factorVIII (FVIII) deficiency, Haemophilia B by deficiency of coagulation factor IX(FIX), and Haemophilia C by deficiency of coagulation factor XI. Theseclotting factor deficiencies are caused by recessive mutations of the respectiveclotting factor genes (Mahlangu & Gilham, 2008).Haemophilia may also be acquired, but this is a much rarer disease, withan incidence of 0.2 to 1.0 case per million per year. -Acquired haemophilia morecommonly affects elderly patients and is caused by auto antibodies to factorVIII (Franchini et al., 2005).After Von Willebrand’s disease, hemophilia A and B are considered thecommonest but also the most severe forms of congenital bleeding disorder, withFVIII deficiency being five times more prevalent than FIX deficiency..(Smith& Owen, 2006).REVIEW OF LITERATURE5Historical perspective:The first documented account of an inherited bleeding disorder was in the2nd century ad. The Babylonian Talmud describes the decision of Rabbi Juddahthat the son of a woman whose three previous sons had bled to death followingcircumcision be excused from the rite (Rosner, 1969).The First Physician Who Described Hemophilia:The famous physician Al -Zahrawi – Albucasis (936-1013 AD), in thesecond Essay of his medical encyclopedia “Kitab al-Tasrif”, described a diseasewhich he named ” ϡÏሕϟ□ ΔሕᏁü” or blood disease. His description corresponds withhaemophilia. The first recent descriptions of haemophilia are from the end ofthe 18th century. In 1803, Dr. John Conrad Otto (1774-1844), an Americanphysician, published an account about ”a hemorrhagic disposition existing incertain families” in the ”New York Medical Repository”. .(Kaadan & Angrini,2009).The actual name ’hemophilia’ was first recorded at the University ofZurich in 1828. Harvard doctors Patek and Taylor discovered the

antihemophilicglobulin in 1937, and a Buenos Aires doctor, Pavlosky, is creditedwith discovering hemophilia A and hemophilia B as triggers to more than oneform of hemophilia. Hemophilia was known as the royal disease due to QueenVictoria being a carrier of it, and passing the gene to a number of nobles such asAlexei, son of the Russian Tsar Nikolai.(Paul, 2004).Several decades later, it was recognized that more than one form ofhemophilia existed. The assumption that there were two forms of hemophiliawas made in 1944 when plasma samples from two different persons, whoREVIEW OF LITERATURE6presumably had the same diagnosis of hemophilia, were mixed together withnormalization of the clotting times(Dunn&Thomas, 2004).The Royal Hemophilia:Haemophilia figured prominently in the history of European royalty inthe 19th and 20th centuries. Queen Victoria, through two of her five daughters(Princess Alice and Princess Beatrice), passed the mutation to various royalhouses across the continent, including the royal families of Spain, Germany andRussia. Victoria’s son Leopold suffered from the disease. For this reason,haemophilia was once popularly called ”the royal disease”. The spread ofhemophilia in the royal families of Europe was a very important factor in thedevelopment of medical knowledge about the disease. The physicians divedinto the cases of hemophilia, trying to uncover its secrets, looking for thesuitable remedy to enjoy the favor of the royal families.Figure (