Hemophilia and royalty

What is hemophilia?

Hemophilia is a bleeding disorder that slows the blood clotting process. People with this condition experience prolonged bleeding or oozing following an injury, surgery, or having a tooth pulled. In severe cases of hemophilia, heavy bleeding occurs after minor trauma or even in the absence of injury (spontaneous bleeding). Serious complications can result from bleeding into the joints, muscles, brain, or other internal organs. Milder forms of hemophilia do not involve spontaneous bleeding, and the condition may not become apparent until abnormal bleeding occurs following surgery or a serious injury.

The major types of this condition are hemophilia A (also known as classic hemophilia) and hemophilia B (also known as Christmas disease). Although the two types have very similar signs and symptoms, they are caused by mutations in different genes. People with an unusual form of hemophilia B, known as hemophilia B Leyden, experience episodes of excessive bleeding in childhood but have few bleeding problems after puberty.

How common is hemophilia?

The two major forms of hemophilia occur much more commonly in males than in females. Hemophilia A is the most common type of the condition; 1 in 4,000 to 1 in 5,000 males worldwide are born with this disorder. Hemophilia B occurs in approximately 1 in 20,000 newborn males worldwide.

How do people inherit hemophilia?

Hemophilia A and hemophilia B are inherited in an X-linked recessive pattern. The genes associated with these conditions are located on the X chromosome, which is one of the two sex chromosomes. In males (who have only one X chromosome), one altered copy of the gene in each cell is sufficient to cause the condition. In females (who have two X chromosomes), a mutation would have to occur in both copies of the gene to cause the disorder. Because it is unlikely that females will have two altered copies of this gene, it is very rare for females to have hemophilia. A characteristic of X-linked inheritance is that fathers cannot pass X-linked traits to their sons.

In X-linked recessive inheritance, a female with one altered copy of the gene in each cell is called a carrier.

A ROYAL DISEASE

Hemophilia has often been called the "Royal Disease." Queen Victoria of England (1837-1901) was a carrier of the hemophilia gene and subsequently passed the disease on to several royal families. Victoria’s eighth child Leopold had hemophilia and suffered from frequent hemorrhages, which were reported in the British Medical Journal in 1868. Leopold died at the age of 31 of a brain hemorrhage. Leopold’s daughter Alice was a carrier, and her son, Viscount Trematon was born with hemophilia. Viscount died in 1928, of a brain hemorrhage similar to the one that killed his grandfather.

Alexandra, Queen Victoria's granddaughter, married Nicholas, the Tsar of Russia in the early 20th century. Alexandra was a carrier of the disease and her first son Alexei, was born with hemophilia. Nicholas and Alexandra were pre-occupied by the health problems of their son at a time when Russia was in turmoil. The monk Rasputin gained great influence in the

Russian court, partly because he was the only one able to help the young Tsarevich Alexei. He used hypnosis to relieve Alexei's pain. The illness of the heir to the Tsar's throne, the strain it placed on the Royal family, and the power wielded by the mad monk Rasputin were all factors leading to the Russian Revolution of 1917. In 1916, the 45-year old faith-healer Rasputin was assassinated in Petrograd by group of noblemen bent on ridding Russia of the monk's corrupting influence on Nicholas II and Alexandra

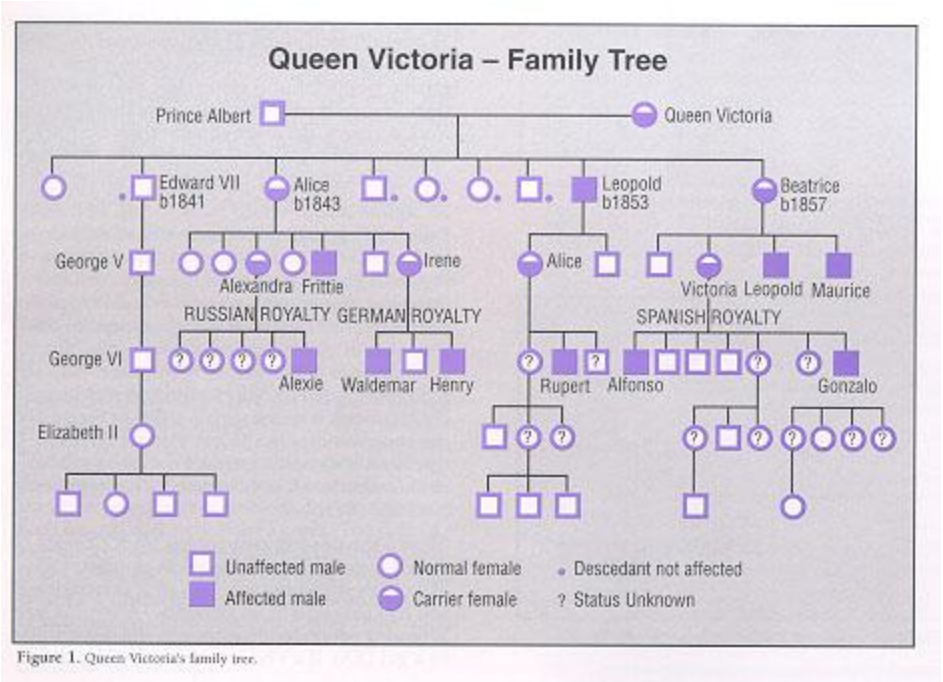

Figure 1. Queen Victoria's family tree.