

Name: _____

Hemophilia: "The Royal Disease"

Hemophilia is an X-linked recessive disorder characterized by the inability to properly form blood clots. Until recently, hemophilia was untreatable, and only a few hemophiliacs survived to reproductive age because any small cut or minor bruise were fatal. Now hemophilia is treated with blood transfusions. However, such treatment is very expensive.

Hemophilia affects males much more frequently (1 in 10,000) than females (1 in 100,000,000). This occurs because a critical blood clotting gene is carried on the X chromosome. Since males only carry one X chromosome, if that is defective, hemophilia will immediately show up. An early death is likely.

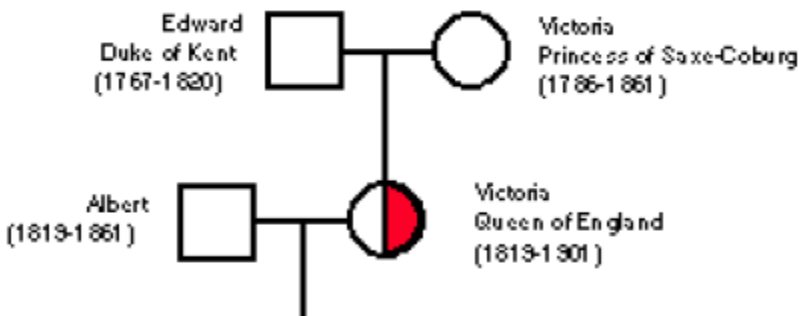
Females, on the other hand, carry two X chromosomes. If only one is defective, the other normal X chromosome can compensate. The woman will have normal blood clotting; she will simply be a carrier of the recessive defective gene. This fact will be discovered if some of her children are hemophiliacs. Naturally, women hemophiliacs are rare because it takes two defective X chromosomes in order for the condition to be seen.

The royal family in England (Prince Charles, Princess Diana, etc.) has a family history of hemophilia. It was once said that the royal family was contaminated with "bad blood". The following genetic problems will help you figure out who in the family has "bad blood".

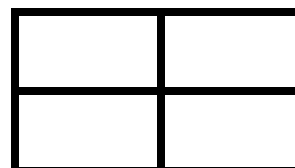


Pedigree Analysis

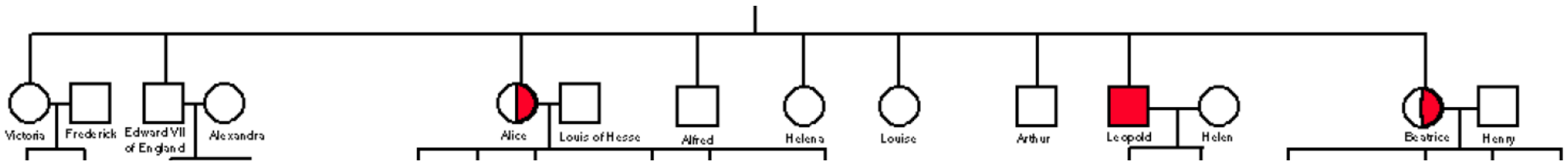
The problem begins with Queen Victoria



If Queen Victoria and Prince Albert have children, what is the probability that their children could have hemophilia? Queen Victoria is a carrier so her genotype is $X^H X^h$



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The above are the children (and spouses) of Queen Victoria and Prince Albert.

1. Identify the actual children of Queen Victoria and Prince Albert (not their spouses).

2. How many individuals have hemophilia? _____
3. How many individuals are carriers of hemophilia? _____
4. Does this match up with the predictions you made earlier? _____

Explain:

5. Prince Charles' great, great grandparents were Edward VII and Alexandra. What is the chance that he has hemophilia? _____

Explain: