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Madeline Mullen

University of New Hampshire, Durham

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Queen Victoria and The Royal Disease

Abstract
Queen Victoria, Queen of England and Ireland and Empress of India, is the second longest ruling monarch of England, she sat on the throne for sixty-three years and was so crucial to the formation of England that there is a whole era names after her, The Victorian Era. She was loved by her people and ruled with a fair but firm hand.

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Queen Victoria and The Royal Disease

by Madeline Mullen
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Queen Victoria, Queen of England and Ireland and Empress of India, is the second longest ruling monarch of England, she sat on the throne for sixty-three years and was so crucial to the formation of England that there is a whole era names after her, The Victorian Era. She was loved by her people and ruled with a fair but firm hand.

Queen Victoria passed on generations of wealth and leadership to her offspring and, in fact, the royal family of England today is still descendant from her, more specifically from her son, Edward (Aronova-Tiuntseva 4). Together with her husband, Prince Albert, Queen Victoria had nine children (Corcos 286). She created a dynasty and as her children married into other royal families across Europe and Asia her family's dynasty only grew. However, beneath the opulence, gowns, balls and strength there lay a secret buried deep in Queen Victoria’s genes, hidden, quiet; never showing itself until she had her final son, Leopold (Aronova-Tiuntseva 4). Only then did they notice the child who should be perfectly healthy was quite sick, and would bleed uncontrollably due to even the smallest cuts, he was diagnosed with Hemophilia; a disease that would haunt the royal family for generations to follow. Queen Victoria and Prince Albert would worry about their son for the rest of their lives, but little did they know that this mutation in Victoria’s genes would last generations after she was gone, and change the very course of history.

Queen Victoria was a carrier of Hemophilia and because of this passed it down to her children and them to their children and so on for generations. This is able to happen because of something called The Founder Effect, or more specifically a genetic bottleneck. In Kurt Vonnegut book, Galapagos, an extreme genetic bottleneck occurs, and I wondered when in history other genetic bottlenecks had happened, and what the impacts were? To look into this though we must first look into the Founder Effect, which was first discovered by a man named Ernst Mayr (Coyne 1213). The Founder Effect is when a population is created based off a very small amount of genetic variation (Coyne 1213). Ernst was born in 1904 and died in 2005 at the age of 100 years old (1213).

Many called him the Darwin of the 21st century and though that may seem a like a bit of an exaggerated epitaph his contribution: to science allow us to understand evolution in a way we could not quite comprehend before his discoveries. He focused a lot through his research on genetics and how they contribute to evolution (obviously in a monumental degree), however he put into words what Darwin suggested through his research but had never quite put a title on before. He states in regard to genetic bottlenecks, “Geographically isolated populations of a single species undergo independent evolutionary divergence,” (1212). While certainly Queen Victoria’s family did no undergo separate “evolutionary divergence” they did have a significant reduced gene pool to choose from in choosing partners and having offspring. In this case Queen Victoria is our founder, and the limitation placed on the royal family to only marry pure royal blood offspring is what creates our bottleneck.

Researcher Montgomery Slatkin says the founder effect can come from an “extreme reduction in population size,” (Slatkin 1). Certainly this was true in many royal families around the world yet it is most potent in Queen Victoria's family because “A founder effect can account for an allele at an unusually high frequency in an isolated population,” (Slatkin 1). This high frequency
occurring gene is the downfall of Queen Victoria’s family. The allele that carried the Hemophilia mutation occurred far more the usual in her family and is what led to the very altering of history.

Hemophilia is inherited as a sex linked trait; more so, it is a trait due to a gene whose transmission is specifically related to the sex of a person (Corcos 286). Because of this hemophilia is found almost always solely in males. This is because hemophilia is associated with a gene mutation on the X chromosome that is the catalyst for creating blood clots (Hermans 847). Females have XX sex chromosomes, so, if the mutation is on one X chromosome they still have the other X which can produce the blood clotting enzymes (Corcos 847). However, males have XY sex chromosomes, so if their one X chromosome carries the mutation their body has no way of creating blood clots (Corcos 847). So, even though hemophilia is a recessive trait, males will always express it, while females can be carriers but not express the trait (Corcos 847).

Specifically, hemophilia is a mutation of the gene coding for F8 or F9 gene which is present on the “long arm” of the X chromosome (Hermans 843). Of the two options, often called type A or type B, type B is less common and more severe. Of F8 and F9, F8 is quite a large protein with 26 exons, while F9 is quite a small protein with only 8 exons. Researchers and scientists were recently able to discover, by excavating the entire Romanov family in 2007, that Queen Victoria passed type B to her offspring and their descendants (Hermans 846). Type B hemophilia occurs when a substitution happens in one of the coding sites, or “exon 4,” from the F9 gene (Hermans 846). One little change at one minuscule coding site may seem insignificant. However, this gene is responsible for creating enzymes which form blood clots, therefore stopping cuts from excessively bleeding both externally and internally (Aronova-Tiuntseva 4). Essentially, for the blood to clot it goes through a process. Says scientist Susan Offner, “each reaction is catalyzed by an enzyme, and each of these enzymes is coded for by a gene” (Offner 1). Without that code it all falls apart. Again we can see why this disease only affects men, women have two X chromosomes so even if the F8 or F9 gene is affected in one X and unable to make enzymes which clot blood, they still have another which can do so. But men only have one X, so when that X has the mutation their blood is unable to clot which is almost always eventually fatal.

Queen Victoria, the matriarch of the royal disease, did not know she was a carrier of hemophilia. In fact, there is some speculation as to if she might have been “illegitimate” because her father was not affected by the disease and it appears her mother was not a carrier either, however, that mystery remains unsolved. Queen Victoria herself only had one son impacted by hemophilia, Prince Leopold; named by the public as the “bleeder prince” (Rushton 1). He died at the age of 31 from a brain hemorrhage, not before having two children though, a daughter and son; the son had hemophilia and the daughter was a carrier (Hermans 844). Surprisingly, although carriers have about a 50/50 chance of passing it on to their children none of her other sons had hemophilia (Hermans 844). Two of of her daughters however, Alice and Beatrice, were carriers (Bhadra 770). However, where we see the true historical consequences occur not from Queen Victoria’s children, but her grandchildren of whom three were carriers/affected; and great children, of whom seven were carriers/affected (Hermans 844).

These offspring married in royal families in Germany, Spain, and Russia; the disease quickly spread throughout the royal families in these countries. Queen Victoria noted herself the danger of this “bottlenecked” gene pool that came from royals only marrying other “pureblood royals,” saying to her daughter Vicky, “I can’t help thinking what dear papa said – that there is some little imperfection in the pure royal descent, that some fresh blood was infused,” (Aronova-Tiuntseva 3). How right she was, if only they had listened.

The real consequences of hemophilia can historically be seen in Spain and most importantly, Russia (Hermans 844). In Spain Alfonso XIII married Queen Victoria’s favorite granddaughter, Princess Victoria Eugenie of Battenberg in 1906 (Hermans 844). Together they had six children, two of which had hemophilia; their firstborn son, Gonzalo, and their youngest son, Alfonso (Hermans 844). King Alfonso took over the throne at a young age. His inexperience combined with the couple’s immense worry over their two very sick children, and the public’s worry about the heir to the throne being so sick, led to the abdication of the throne in 1931, (Hermans 845). His replacement eventually led to the rule of cruel dictator, Francisco Franco, who tormented the
country for many years (Hermans 845). This all stemmed from Princess Victoria passing on the gene to two of their sons, which altered the very course of Spanish history. Surprisingly however, the first royal marriage to be declined because of the potential for passing on Hemophilia was not declined until 1913, (Stevens 28).

However, the events caused by hemophilia entering into the Russian royal family had a far greater impact on their country. Czar Nikolai of Russia married Queen Victoria’s granddaughter, Princess Alix, in 1894, (Stevens 27). Together the couple had their first son, Alexis, who was quickly diagnosed with hemophilia, (Hermans 845). The couple was absolutely distraught by the pain the disease caused their son, a close friend of Princess Alix said of her, “She hardly knew a day’s happiness after she knew her boy’s fate,” (Potts 2). they turned to a local well known monk, Rasputin, (Hermans 844). Although Rasputin was basically a fancy con man he did somehow slightly ease Alexis’s pain and so Rasputin became a permanent fixture of the royal Russian family and also become intimately close with the family, particularly Princess Alix (Hermans 845). In this way he was actually able to wield influence of the affairs of the state, particularly during World War I (Hermans 845). The people of Russia quickly began to dislike Rasputin and discredit the Royal family. Many believe that it was due to this that the Romanov family fell and the Bolshevik Revolution occurred, in 1917, in which the whole government of Russia was overthrown and the whole Romanov family, including all their children, were brutally murdered. It is interesting to imagine what would have happened if hemophilia had not been in the Russian royal family, would the Bolshevik revolution have even happened? We will never know but it is clear the hemophilia did alter this country’s history.

Hemophilia today is not as fatal a diagnosis. There are measures that can easily be taken to clot blood if you do have a mutation on your F8 or F9 gene. If the mother or father is a carrier the child can immediately be tested. However, Queen Victoria’s family possessed the disease in a time when none of this was possible. She did not know she was a carrier, the family did not understand they had created a genetic bottleneck and that they were all passing the disease to each other. They certainly could have never comprehended the impact this one little mutation could have on the entire course of European history. The disease caused Queen Victoria’s families and royal families across Europe unimaginable pain. Yet through this pain it also serves as an incredible example of genetics and how the Founder Effect and genetic bottlenecks can impact a family and their offspring for generations to come.

Bibliography


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