

# A ROYAL DISEASE: CAN A HEMOPHILIAC BE CIRCUMCISED?

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**D**uring the nineteenth and twentieth centuries, European royalty suffered from a sex-linked genetic disease that prevents blood clotting, known as hemophilia. It is believed that Queen Victoria passed on the X-linked mutation to her two daughters, Princess Alice and Princess Beatrice. The disease had a calamitous effect upon the British royal family. The nineteenth century Duke of Albany, for example, slipped, fell, and died of blood loss in the year 1884. Queen Victoria's grandson, Friedrich, died from exsanguination, fatal blood loss, at the young age of two years old. Similarly, Leopold and Maurice, two other grandsons, died at the ages of thirty-two and twenty-three, respectively. Many of Queen Victoria's heirs inherited the disease, and as they married into different royal families, the pathology was dubbed the "royal disease" and spread through the royal families of Britain, Spain, Russia, and Germany. Through DNA analysis on the bones of the Russian royal family it has been discovered that Russian royalty, the Romanovs, suffered from a rare subtype of the blood clotting disorder, hemophilia B [1].

Hemophilia is an X chromosome-linked disorder that may be inherited from either the mother or the father; however, hemophilia is nearly exclusively manifested in males. Males, said to be hemizygous, inherit one X chromosome from their mothers and thus have a single copy of all X-linked genes. Women, on the other hand, inherit two X chromosomes, giving them a "double dose" of sex-linked genes. Although the trait for hemophilia is recessive, since males inherit a Y chromosome from their father, which lacks the gene for blood clotting, they are unable to mask a deleterious gene on their X chromosome. Females may be carriers for X-linked diseases if they carry the deleterious gene on only one X chromosome, giving them a fifty percent chance of producing affected sons [2].

As mentioned previously, hemophilia is a disorder that prevents blood from clotting normally in the event of a wound. For a hemophiliac, even a minor cut can bleed for a long period of time. When one bleeds, the body launches a series of reactions, called the coagulation cascade, to clot the blood [2]. The coagulation cascade involves proteins known as coagulation factors. One has a higher chance of bleeding when one of these coagulation factors

is missing. Hemophilia A is characterized by a lack of the blood clotting factor VIII. Without sufficient factor VIII, the blood cannot clot properly to stop bleeding. Hemophilia B is characterized by a deficiency of blood clotting factor IX. Similarly to hemophilia A, one who is affected by hemophilia B lacks the ability to clot normally in order to control bleeding [3].

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Records of medical literature indicate that hemophilia was first discovered in 1803 by John Conrad Otto [3]. However, long before Otto, a passage from the Talmud conveyed that the genetics behind the sex-linked blood clotting disorder was understood. Rabbi Judah and Rabbi Simeon hold differing beliefs in the amount of repetitive events necessary to establish a pattern for the transmission of hemophilia. On the one hand, Rabbi Simeon believed one should wait after the passing of three sons who died as a consequence of circumcision in order not to circumcise the fourth child; however, Rabbi Judah believed the third child should not be circumcised. If the mother found that circumcising her first two sons resulted in their death, she should not circumcise her third son. The twelfth century Biblical commentator and physician, Maimonides, detailed in his work the *Mishneh Torah, Sefer Abavah, Hilchot Milah* 1:18, that hemophilia was undoubtedly transmitted from the mother. Maimonides explained that if a woman's first son died as a result of circumcision, which "enfeebled his strength," and her second son also died as a result of circumcision, regardless of whether her second son was from her first husband or second husband, she should not circumcise her third son. Further extrapolating on Maimonides' work, Rabbi Joseph Karo explained that there are families "in which the blood

is weak;” furthermore, like Maimonides, Rabbi Karo explained that the mother should not circumcise her third son in the event that her first two sons died after circumcision [5]. Evidently the scholars of the Talmud, as described by Maimonides and Rabbi Joseph Karo, recognized that hemophilia is transmitted maternally, and that it causes exsanguinations, leading to the death of the circumcised child.

While performing surgery on hemophiliacs could pose many problems because of their inability to clot, modern medical and technological advancements have made many surgeries possible, including protocols that make it feasible to circumcise hemophiliacs. This procedure relies upon factor concentrate replacement therapy followed by continuous post-surgery replacement therapy for diverse periods of time. Haemostatic agents, substances that promote the stop of bleeding, such as fibrin glue, are used during the procedure. Fibrin glue is a topical adhesive that imitates the function of clotting factors in that it emulates last stages of coagulation. The haemostatic agent has proven to be successful in controlling bleeding in other realms of surgery, such as in neurosurgery and cardiovascular surgery. In the event that the missing clotting factor is provided to the hemophiliac baby to safely perform circumcision, some Rabbis prohibit the circumcision on the Sabbath if the missing clotting factor would have to be ad-

ministered intravenously [6]. Rabbi J. David Bleich considered the aforementioned procedure; however, he also explored an alternative route in the pursuit of circumcising a hemophiliac: laser surgery. Rabbi Bleich stipulated that laser circumcision can satisfy the requirement of circumcision to be a *koah adam*, a human act, because the already existing laser rays are brought to bear upon the foreskin by a direct human act [7]. On September 5<sup>th</sup>, 1998, Judy Siegel, a writer for the Jerusalem Post, reported that a two-month-old baby of Israeli immigrants had undergone laser surgery. Dr. Shlomo Wallfish, who had initially performed laser circumcision a decade earlier, performed the laser circumcision on the young child [8].

Circumcision has been an ongoing practice for the Jews for thousands of years. Even during the midst of persecution when circumcision was forbidden, the Jewish people resorted to daring measures to ensure the continuation of the ritual. However, the *halakhic* ramifications of performing the traditional procedure on a hemophiliac have since evolved. Medicine has greatly advanced since the times of our Talmudic sages who stipulated that if hemophilia was a confirmed genetic disease in the family it should not be performed on a hemophiliac. Today, the use of haemostatic agents and laser surgery allows a hemophiliac to take part in the sanctified practice of circumcision. ■

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