

# Genetics, History, Genealogy and Cinema: Alexis Románov's Haemophilia

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## Summary

This paper takes us on a brief journey through history, highlighting among the many signs of anomalies related to blood-clotting, some of the key points for the updating of our current knowledge of haemophilia, both causal and clinical, as well as, and based on this, the possible keys to present and future treatments of choice.

The designation of haemophilia as a "royal disease" comes from its frequency among European royalty. Queen Victoria I of Great Britain passed it on to her son Leopold and, through her daughters, to the Spanish, German and Russian royal families, even affecting Tsarevich Alexei.

His suffering the disease and, as a result of this, his being linked to Rasputin, together with the influence of such a relationship with the royal family on family and political/social affairs and his tragic death at the hands of the Bolsheviks, add interest to this information.

We will focus our attention on two films in which Alexei and his haemophilia acquired special relevance, completing our article with the mention of archive films in which Alexei participated personally and others in which material from such archives has been combined with fictional scenes.

The text and context of this story still arouse controversy and, although there were many factors involved in the historical event, a mutation, an empress carrying haemophilia and a much-longed-for but sick successor to the throne in troubled times, are by no means lesser or marginal ingredients in such a scenario.

**Keywords:** Haemophilia, Genetics, Royal disease, Alexei Romanov.

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## About haemophilia

The origins of haemophilia can probably be traced back to the Cretaceous period. This hereditary disease appears in at least three orders of placental mammals, which differentiated about 65 million years ago: *Perissodactyla* (odd-toed ungulates), *Carnivora* and *Primates*, being specifically described in horses, dogs and humans. This alteration, lethal when uncontrolled, has probably appeared recurrently<sup>1,2</sup>.

As far as its history is concerned, there are texts written centuries before Christ reporting that the rabbis had detected that some male children who bled excessively when circumcised belonged to certain specific families. In the 2nd century A.D, Yehuda haNasi would exempt the third male child from circumcision if his two older brothers had died or suffered from severe haemorrhages following it, while Simon Ben Gamaliel exempted a male child from this practice after the male children of his three maternal aunts had died as a consequence of the disease<sup>3</sup>.

Between the 10<sup>th</sup> and 13<sup>th</sup> centuries, two doctors from Cordoba: the Arab Albucasis or Albucaasin (Abul-Qasim Khakaf ibn al Abbas al Zahravi) and the Jew Maimonides (Moshé ben Maimón o Musa ibn Maymun) wrote about haemophilia, the latter reporting in his treatises "Guide for the Perplexed" and "Mishneh Torah" that it was mothers who passed on the disease to their male descendants, thus establishing that if a woman had children with this haemorrhagic alteration and was to marry again, none of her new male descendants should be circumcised. However, Maimonides mistakenly believed that the disease disappeared with age<sup>4</sup>.

A reference to this disease in Europe can be found as early as the 16<sup>th</sup> century thanks to Alexander Benedict; descriptions of it can be found in the 18<sup>th</sup> century (Zoll in 1791, Cronsbuch in 1793 and Rave in 1796), and its first mention in relation to sex was made at the beginning of the 18<sup>th</sup> century (1803) by the American doctor John Conrad Otto, who not only reported that the problem was hereditary, that it mainly affected males and that it was transmitted by healthy women, but was also able to trace the disease back to a woman who used to live near Plymouth. Besides this, the historical horizon was extended by the contributions made towards the end of the same century by the British William Hunter, who showed that blood plasma and not red blood cells was responsible for coagulation, and by those made towards the end of the 19<sup>th</sup> century by the German orthopaedic surgeon Franz König on how the disease could affect the knees and how this used to be mistaken for arthritis or tuberculosis<sup>5</sup>.

The term haemophilia was first established in a doctoral dissertation presented in 1828 by Friedrich Hopff, a Swiss student of medicine from the University of Zurich<sup>5</sup>.

In 1937, the Americans Arthur J. Patek Jr and Floyd H. Taylor, from Harvard University, discovered anti-haemophilic globulin<sup>6</sup>, and in 1947 the Argentinean doctor Alfredo Pavlosky differentiated, *in vitro*, two types of haemophilia: A (suffered by one in 50,000 people, recessive, linked to the X chromosome and caused by the absence of factor VIII), and B (affecting 1 in 10,000 people, also linked to the X chromosome and caused by the absence of factor IX)<sup>7</sup>. Haemophilia C (not completely recessive, autosomal and linked to the absence of factor XI) was described in 1953 by Rosenthal, O.H. Dreskin and N. Rosenthal<sup>8</sup>.

To summarize, many cases of anomalies in coagulation have been recorded throughout history, but some of them have proved essential in the establishment of our current knowledge of haemophilia in its full extent, both causal and clinical, also and derived from this proving to be the keys to present and future treatments of choice.

### Presentation of the "aim": thirteen years in the life of Alexei Nikolaevich Romanov<sup>9,10</sup>

The designation of haemophilia as a "royal disease" arises from the frequency of its appearance among the European royalty. Queen Victoria I of Great Britain (1819-1901) passed it on to her son Leopold and, through her daughters, to the Spanish, German and Russian royal families.

One of those affected by this disease of the blood was Tsarevich Alexei, the youngest child of Nikolay Alexandrovich Romanov, Tsar and Autocrat of All the Russias, and of Alexandra Feodorovna Romanova (born Viktoria Helena Luise Beatrice of Hesse-Darmstadt), granddaughter of Victoria I, Queen of Great Britain and Ireland and Empress of India.

His Imperial Highness, Tsarevich and Grand Duke of Russia, his official titles, was born on the 12<sup>th</sup> of August of 1904 in Peterhof, near Saint Petersburg. He was the fifth child and only son born from the marriage between Nicholas and Alexandra and heir to the throne because of the law of imperial succession, established by Tsar Paul I in 1797 and based on the Salic law.

One month after his birth, an excessive bleeding from the umbilical area confronted the royal family with a situation that at first they denied, but which they were later forced to accept. The family retreated into isolation and this problem was concealed from the people, since as the Tsar was their leader and head of the Church he should be free of flaws, especially in light of the fact that the people attributed all physical impairments to the intervention of divine powers. While Alexei proved a lively, energetic, mischievous and boisterous child, his disease gave him no relief, one of his worse crises taking place while on a visit in Spala (Poland) at the age of 8 (Figure 1).



**Figure 1:** Empress Alexandra prays by Tsarevich Alexei's bed during his haemophilia crisis of 1912.

With the heir on the verge of death, his parents turned to the services of a well-known Siberian peasant regarded as a mystic and believed to engage in licentious behaviour, Grigori Yefimovich Rasputin, who, by suppressing the aspirin intake prescribed by doctors to ease the Tsarevich's pain (although it worsened the symptoms), managed to control the critical situation. Nicholas' and Alexandra's belief in Rasputin's supernatural powers and the conviction of the peasant himself, now also a healer, granted him the trust of the royal court where his influence was not only "therapeutic", but also extended to family and political affairs (\*).

In order to prepare him for a doubtful future as a Tsar, Nicholas allowed Alexei to attend government meetings, and during WW1 he took him to Stavka, one of the general headquarters of the Supreme Commander of the Russian armed forces, and to Mongoleiv, where Nicholas became commander-in-chief of the Russian Army.

Meanwhile, Rasputin's growing power and licentious way of life were brought to a halt in 1916 by Prince Felix Yusupov who, together with some comrades and even a British agent, after poisoning and shooting him repeatedly without success, ended up throwing him wounded as he was into the Neva River, where he drowned.

In March 1917, when the Russian Revolution took place, Nicholas abdicated in favour of Alexei, although he later retracted in favour of Grand Duke Michael, the Tsar's brother, since Alexei was expected to live at most six years more. Nevertheless, Michael refused to accept.

The royal family was moved to Tobolsk, Siberia, and later to Yekaterinburg, where they were all murdered during the early hours of the 17th of July of 1918, which, as we know, has been duly established. Alexei, who was on the Tsar's lap during the execution due to a serious accident that had affected his right knee, leaving him an invalid who had already been forced to rest for days, was finished off by a shot in the head by his father's executioner, Yakov Yurovsky.

All the bodies were exhumed between 1991

(\*) The first of the Surgeon General Catalogues of the USA to include a section on the treatment of haemophilia was a volume published in 1901. As possible therapeutic measures, it mentions the administration of calcium and nitrogen, the inhalation of oxygen, the use of thyroid gland and bone marrow, and the use of hydrogen peroxide and gelatine<sup>11</sup>. However, we have found nothing of this in the therapy undergone by the Tsarevich, except for a reference to aspirin and the "healing prayers" of Rasputin, more magical in those days than in our times if we take noetics into account.

and 2007. Today they remain buried at the Peter and Paul Cathedral in St Petersburg. The Tsarevich, Nicholas, Alexandra, Olga, Tatiana, Maria and Anastasia were all canonized as martyrs by the Orthodox Church in 2000.

Today we have proof that all the children of the royal couple had the same mitochondrial DNA as their mother, and that the haemophilia suffered by the Romanov family, in common with that of the other royal families, was of type B<sup>12,13</sup>.

Alexei Romanov's suffering, his relationship with Rasputin, the influences of this relationship on family and political affairs, his tragic murder by the Bolsheviks and its current assertion make him a particularly interesting character.

### Alexei Nikolayevich Romanov as seen in cinema, TV and the global network

While we have not found any films exclusively dedicated to the figure of Alexei, his presence in the history of cinema can be perceived through films directly and indirectly inspired in the tragic life and death of the Russian royal family using archive footage, played by different actors and even by one actress: Jessica Sportelli, in *The Romanovs' Last Photograph* (2007) by Catherine Faris King, an American black-and-white production that mixes documentary and fiction<sup>14-15</sup>.

Leaving aside films from the time of silent (1917-1929) and talking movies (1966-2008), fictional films, documentaries and archive films combining documentary and fiction where Alexei's disease was not the central topic, or where it was mentioned only incidentally without having any relevance to the plot, this paper deals with two films in which Alexei and his haemophilia play an important part in the development of the plot.

### *Rasputin and the Empress* (1932) by Richard Boleslawsky

#### Technical Details

**Title:** *Rasputin and the Empress*.

**Country:** USA.

**Year:** 1932.

**Director:** Richard Boleslawsky.

**Music:** Herbert Stothart.

**Photography:** William H Daniels (as William Daniels).

**Film editor:** Tom Held.

**Screenwriter:** Charles Mac Arthur, Lenore J. Coffee (unaccredited), Ben Hecht (unaccredited), Robert E. Sherwood (unaccredited) and Mercedes de Acosta (unaccredited).

**Cast:** John Barrymore (Prince Paul Chegodieff), Ethel Barrymore (Tsarina Alexandra), Lionel Barrymore (Grigori Rasputin), Ralph Morgan (Tsar Nicholas II), Diana Wynyard (Princess Natasha), Tad Alexander (Tsarevich Alexei 'Aloysha'), C. Henry Gordon (Grand Duke Igor) Edward Arnold (Dr A. Remezov)...

**Color:** Black and white.

**Runtime:** 121 minutes.

**Genre:** Drama and History.

**Production company:** Metro Goldwyn Mayer (MGM).

**Synopsis:** The film recounts the last years of the Romanov Empire and shows Rasputin's influence on the court and on life in Russia.

**Awards:** Nominated for Oscar for Best Original Screenplay (1934).

<http://www.imdb.com/title/tt0023374>

[Trailer 1](#)

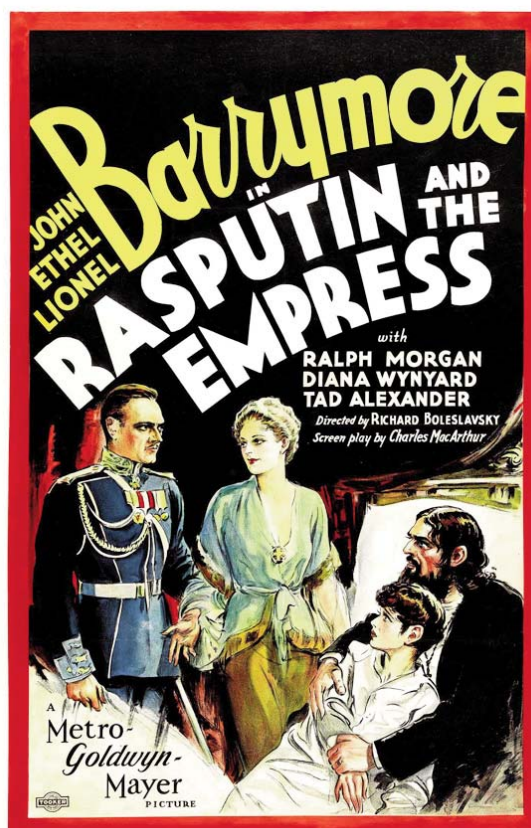
[Trailer 2](#)

The plot develops between 1913 and 1918 and it tells the story of the last years in the life of the Russian royal family. It begins with a scene of the celebration of the tercentenary of the Romanov dynasty and the social agitation that was to destroy it. Months later, Tsarevich Alexei (Tad Alexander), who is a haemophiliac, has an uncontrollable haemorrhage as the result of a fall. Since Dr A. Remezov (Edward Arnold) is unable to control the bleeding, Tsarina Alexandra (Ethel Barrymore) agrees, on the indications of Princess Natasha (Diana Wynyard), her lady-in-waiting and fiancée of Prince Paul Chegodieff (John Barrymore), to let her son be attended by Grigori Rasputin (Lionel Barrymore), a Siberian monk known for his skills as a healer. He manages to control Alexei's haemophilia crisis through hypnosis. He wins the royal family's trust and thus his influence over them begins, while he also gains control over the population and his surroundings. Time goes by and one night Rasputin attempts to rape Princess Maria (Jean Parker), the Tsar's daughter. Following this, he loses the favour of Natasha and the Tsarina, and Prince Paul, who has married Natasha, ends up killing him. The royal family, in the midst of a revolutionary situation, is likewise murdered by the Bolsheviks.

The film reports that both the uncle and the grandfather of the Tsarina died of haemophilia, a disease that is considered a legacy of the house. There is also mention of the fact that the disease does not affect everyone, that women only pass it on to their male children, and that it is due to the lack of a certain element in blood which prevents it from coagulating, such that the lightest injury may lead the person who suffers from it to bleed to death (Figure 2).

Several curious aspects appear in this film. On

the one hand, three of the main characters - John, Ethel and Lionel Barrymore - belonged to the same family and played leading roles in this film, the only one where they ever performed together. They were the great-uncles and great-aunt of present-day actress Drew Barrymore. On the other hand, a lawsuit was filed against the film because the characters of Prince Paul and Princess Natasha were based on Prince Felix Yusupov and Princess Irina Yusupov, and with regard to the latter, the film implied that she had been raped by Rasputin. In face of such a lawsuit based on invasion of privacy and libel, MGM was made to pay compensation and cut the scene. Since that moment, the following disclaimer began to be incorporated in the credits: "This film is a work of fiction and any resemblance to real persons, living or dead, is purely coincidental"<sup>16</sup>.



### *Nicholas and Alexandra* (1971) by Franklin J Schaffner

#### Technical Details

**Original Title:** *Nicholas and Alexandra*.

**Country:** UK.

**Year:** 1971.

**Director:** Franklin J Schaffner.

**Music:** Richard Rodney Bennett.

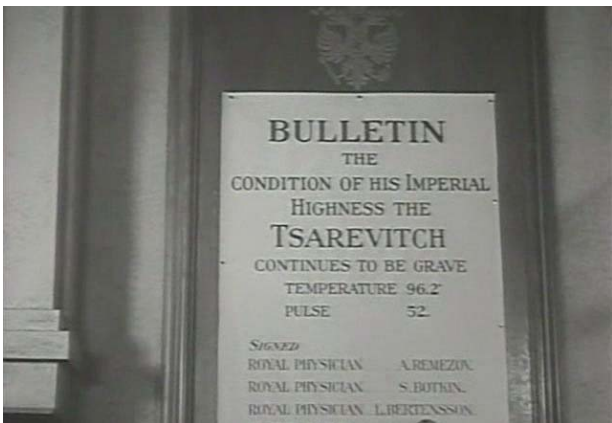


Figure 2: Tsarevich Alexei's haemophilia and Rasputin in *Rasputin and the Empress*.

**Photography:** Freddie Young.

**Film editor:** Ernest Walter.

**Screenwriter:** James Goldman and Edward Bond based on the homonymous biography by Robert K. Massie.

**Cast:** Michael Jayston (Nicholas II), Janet Suzman (Alexandra), Roderic Noble (Alexei), Ania Marson (Olga), Lynne Frederick (Tatiana), Candace Glendenning (Maria), Fiona Fullerton (Anastasia), Harry Andrews (Grand Duke Nicholas), Irene Worth (Queen Mother Maria Feodorovna), Tom Baker (Rasputin), Jack Hawkins (Count Fredericks), Timothy West (Dr Botkin), Katherine Schofield (Tegleva), Jean-Claude Drouot (Gilliard), John Hallam (Nagorny)...

**Color:** Color.

**Runtime:** 183 minutes

**Genre:** Biography, Drama and History.

**Production companies:** Columbia Pictures Corporation and Horizon Pictures.

**Synopsis:** Reconstruction of the life of Nicholas II (1868-1918), the last Russian Tsar, and his wife Alexandra, during the years of corruption, oppression and poverty that culminated in the Russian Revolution of 1917 (public synopsis).

**Awards:** It won 2 Academy Awards in the categories of Best Art Direction and Best Costume Design, and was nominated in the categories of Best Actress in a Leading Role (Janet Suzman), Best Cinematography (Freddie Young), Best Original Score (Richard Rodney Bennett) and Best Picture (Pam Spiegel) (1972).

<http://www.imdb.com/title/tt0067483>

[Trailer 1](#)

[Trailer 2](#)

[Trailer 3](#)



Figure 3: Alexei in *Nicholas and Alexandra*.

Alexei's haemophilia is diagnosed shortly after his birth. Alexandra, his mother, is desperate and feels guilty for passing the disease on to her child and the only successor to the throne (Figure 3). This leads her to befriend Rasputin, a Siberian monk who persuades both her and Nicholas II himself that he can heal Alexei. The film displays the private life of the royal family, the horrors of WW1, Rasputin's adventures and how he was killed by Prince Yusupov along with other conspirators, and the joint suffering of a son due to his disease and of a father who, following the signing of the document that put an end to 300 years of the Romanov dynasty, bursts into tears. The fact that the end (exile to Siberia, vain hopes of salvation, and final execution in the basement of the Ipatiev House in Yekaterimburg) is common knowledge because it is historical does not make it any less tragic or moving.

The sequence that deals with the diagnosis begins almost 23 minutes into the film and is as follows:

- Doctor: *"The Tsarevich's wound worries me so I ordered some tests. I wasn't sure because I am no specialist but my colleagues agree".*

- Tsar: *"Can't there have been a mistake, is there no doubt?"*

- Doctor: *"The diagnosis is haemophilia".*

- Tsar: *"I see. I'm sorry but I need to get used to the idea... So, what is the treatment?"*

- Doctor: *"There isn't any Sir".*

- Tsar: *"But that can't be possible. There must be some kind of medication".*

- Doctor: *"There isn't any medication".*

- Tsar: *"At all?"*

...

- Tsarina: *"I see".*

- Doctor: *"It is transmitted by women; the mother passes it on to her children. Your mother inherited it from her mother, Queen Victoria, and passed it on to you".*

- Tsarina: *"I see".*

- Doctor: *"It affects some males, not all of them. Of Victoria's four sons, only one of them had haemophilia".*

- Tsarina: *"And only one of mine".*

- Doctor: *"Some haemophiliacs can lead a long and normal life".*

- Tsarina: *"But others can't".*

- Doctor: *"The chemical agent that intervenes in the coagulation of blood is missing. If they cut themselves, they bleed profusely".*

- Tsarina: *"Then we'll be careful so that Alexei doesn't cut himself".*

- Doctor: *"Unfortunately that's not all. A superficial wound..."*

- Tsar: *"Allow me. External wounds are not so important. If Alexei cuts his hand the bleeding can be stopped with a bandage. But if the bleeding is internal..."*

- Tsarina: *"Internal?"*

- Tsar: *"Any contusion could lead to internal bleeding. You never know. A heavy blow might not hurt him and the lightest friction might make him bleed. The blood flows very slowly and he bleeds for days. The affected area begins to swell up until the pressure is so intense that the haemorrhage stops. The danger lies in that while the area remains swollen the blood attacks the bone".*

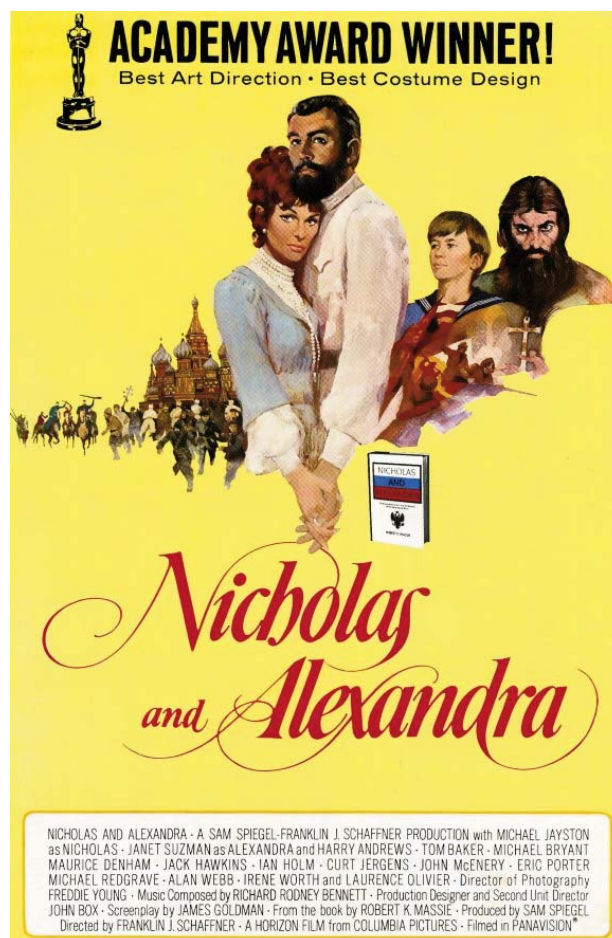
- Tsarina: *"No, Nicholas".*

- Tsar: *"The limb might become twisted and when it does nothing can be done. That's what they've told me".*

- Tsarina: *"Tell them... Tell them that they're wrong, that we'll see other doctors. Tell them to leave. My child is perfect. He will live a long life and he will become a great Tsar, like his father".*

A scene showing how Alexei is attended by doctors during one of his crises is available from the following link: <http://www.youtube.com/watch?v=-PwP6bvDA>

Robert K. Massie, author of *Nicholas and Alexandra*, on which the film was based, had a son who suffered from haemophilia. Jaime de Mora y Aragon, the brother of Queen Fabiola of Belgium plays a small part, and George Rigaud, an actor born in Argentina where he took part in several films and who later lived in Spain and France, also works in this film.



Finally, the existence of archive films, recovered from different web pages, where Alexei took part personally, is worth mentioning. In some of them he appears seated or held by others while trying to act normally under the influence of one of his haemophilia crises, in others archive footage is alternated with scenes of fiction<sup>17-19</sup>.

### Genetics and history

The Church of the Saviour on Spilled Blood, also known as the Cathedral of the Resurrection of Christ, rises in St Petersburg as a temple – a monument built on the spot where Alexander II was mortally wounded on the 1<sup>st</sup> of March of 1881. His remains are buried in the Peter and Paul Cathedral, where the remains of the last Tsar and his family also lie. Among the latter can be found those of a young man of nearly fourteen years of age, to whose genetic misfortune – haemophilia, which did not allow him to lead an ordinary childhood and exposed him to an early awareness of death – was added a historical period of intolerance that finally led to the extermination of all his family.

It is not difficult to imagine that the fact of trying to find a medical solution to this problem, common to royal families, since the eldest sons were those who suffered the disease, might have been one of the reasons for our knowledge about haemophilia to evolve faster, given the pressure royalty might have exerted on the doctors who were trying to find a successful treatment for it that did not exist at the time.

The text and context of this story are still controversial, since it is impossible to establish a link between the disease, the events that took place at that time, and the revolution of 1917. While many other elements converged in the historical event, a mutation, an empress who carried the disease, and a much-longed-for but sick successor to the throne in turbulent times, are by no means lesser or marginal ingredients in such a scenario.

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