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Could Sleeping Beauty have been a haemophiliac?

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Abstract
In the fairy tale Sleeping Beauty pricks her finger on a spindle and due to a curse placed on her as a baby, falls to sleep for a hundred years. Here we explore the ramifications of a scenario where instead of a curse, Sleeping Beauty has haemophilia and cuts her finger at age seventeen. We first establish that to have reached her age without modern medicine, it is most likely the princess has a mild form of the disease. A model is then constructed resulting in the conclusion that if Sleeping Beauty was a haemophiliac, a relatively minor cut to her hand left untreated would be sufficient for her to 'fall into a slumber' as in the story or, put simply, faint.

Keywords: *Fairy Tale; Biology; Haematology; Haemophilia; Sleeping Beauty*

Introduction
Sleeping Beauty is a familiar fairy tale with many differing adaptations all centred around common themes. There is some argument about which story is the original, however, one of the most well-known versions is that of the Brothers Grimm [1]. It is this that will make up the source text where required in this paper.

In the fairy tale Sleeping Beauty (she is not named within the Grimm Brothers' story) is cursed, by an aggrieved fairy who had not been invited to her christening, to prick her finger on a spindle on her seventeenth birthday and die [1]. It is only by virtue of the fact that one of the other fairies was yet to bestow her own gift that the curse could be reduced from death to a hundred-year slumber [1].

The purpose of this paper is to explore the consequences of a scenario where instead of a curse from a fairy and pricking her finger on a spindle, the princess was a haemophiliac who suffers a cut on her finger.

Haemophilia
Haemophilia is an inherited blood disorder in which the individual has low or virtually absent levels of particular clotting factors [2]. These clotting factors usually bind with platelets in the blood following an injury, allowing the platelets to form clumps and stem the flow of blood [3]. There are two types of haemophilia, A and B, which are associated with clotting factors VIII and IX respectively [4]. Both of these factors are involved in the formation of the same complex and so, for our purposes, there will not be any further discussion as to their distinction as it makes no difference to their clinical presentation [3].

Haemophilia is a recessive sex-linked characteristic that is inherited due to a mutation on the X-chromosome [4]. This means that it is significantly more common in men than in women. The simplest way for Sleeping Beauty to have inherited the condition would be for her mother to be a carrier or affected and father to be an affected individual [2].

As recently as 1970 the average life expectancy for haemophiliacs was just 20 years old [5]. In the fairy tale, Sleeping Beauty is 17 years old when she pricks her finger on a spindle and so her age fits very reasonably into our haemophilia concept.

Haemophiliacs can be further classified by the severity of their affliction usually measured by the level of clotting factors they have, as summarised in Table 1 (next page).

Model
In order to model Sleeping Beauty as a haemophiliac we must make several assumptions. The first of these is the severity of her condition. The Grimm Brothers make no mention of her life from the christening to

	Clotting factor level (%)
Mild	6-40
Moderate	1-5
Severe	<1

Table 1 – A summary of the blood levels of the affected clotting factor. Data taken from [3].

the age of seventeen however, as previously mentioned, in 1970 haemophiliacs tended to die as young adults – and that is with 130 years of development in treatment, no such treatment would have been available in the context of Sleeping Beauty [1, 5]. From this we can assume that she has a mild form of haemophilia i.e., she does not suffer from frequent spontaneous bleeding into her joints and muscles, but she does bleed for a long time even following minor injuries [2]. Individuals with haemophilia are currently advised to contact health services if they can't stop the bleeding at home after 30 minutes and so we will take that as the duration of our model [6].

We also need to establish what volume of blood our threshold for 'success' is. It is extremely difficult and comes with many ethical issues to try and measure blood loss following a physical trauma, but generally once you lose ~40% you are considered to have a 'preterminal condition' and it is vital that you receive a transfusion [7]. Since there is no possibility of a transfusion for Sleeping Beauty, and in the fairy tale she does not actually die we will take a slightly reduced volume of 30% as our threshold. If we use Nadler's formula (for females) [8]:

$$V = (0.3561 \times H^3) + (0.03308 \times W) + 0.1833$$

Where V = blood volume (L), H = height (m), W = mass (kg). Taking values of H – 1.625m, W – 56kg gives us a total blood volume of 3.564L which we will approximate here as 3600ml. This means if we deem it reasonable that Sleeping Beauty could lose 1080ml of blood in half an hour from a cut to her finger her '100-year sleep' could feasibly be due to her being a haemophiliac.

The last and most difficult assumption we have to make is the flow of blood from the wound. As mentioned previously, there is considerable difficulty involved in measuring losses from injuries and quantifying exactly how this changes with severity. It has also been noted that within the literature the same terminology is used in relation to perfusion (millilitres per minute per millilitre/gram) while actually referring to different measurements without clarifying this [9]. To circumvent these numerous difficulties primary measurements were taken using a dripping tap over the course of a minute. These gave a rate of 34ml lost per minute from a moderately bleeding cut, see figure 1 below. Note – the water was *not* a constant stream but a steady drip (20-25 drips every 10 seconds).

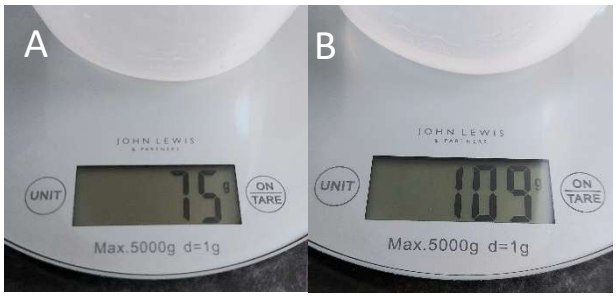

Figure 1 – [A] Mass of empty jug (g). [B] Mass of jug after being filled by a steadily dripping tap for 1 minute (g). Photos by Sarah Hume.

A simple multiplication (34×30) tells us that if Sleeping Beauty lost blood at this rate for half an hour, she would lose 1020ml. As this is 60ml less than our proposed threshold value it does not quite meet our criteria, however, it is remarkably close (0.0555…% less). While not incontrovertible proof of haemophilia in the way that a more extreme value may have been, this boundary result actually supports the idea of a relatively mild form of haemophilia such as would've allowed the princess to have reached the age of seventeen without access to the prophylactic measures available today.

Conclusion

Haemophilia is a blood disorder characterised by excessive bleeding following injuries due to reduced or absent numbers of clotting factors. If left untreated this blood loss can have significant negative physiological impacts or even lead to death. Were the story of Sleeping Beauty to include no curse, our model shows that it's possible for her to have fainted following an injury to her finger (although it would have to be more severe than a prick) if she were a haemophiliac. Mild haemophilia is most likely, due to her age and the marginal result of our calculations.

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