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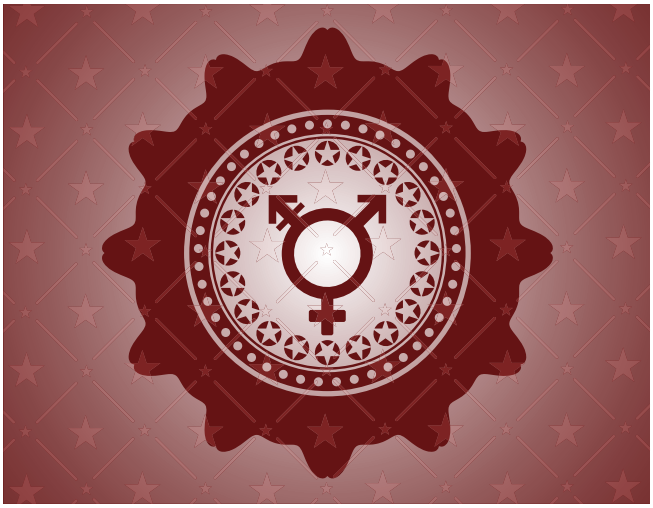
CASE STUDY

A transgender person with haemophilia

Caroline Valk

Introduction: Transgender people face many obstacles to accessing healthcare but cultural changes are likely to increase provision of sex reassignment surgery in countries with sufficient resources. Haemophilia services traditionally focus on providing factor replacement therapy for males and should therefore understand how the care they provide can be adapted to meet the needs of transgender people. Haemophilia is an X-linked congenital bleeding disorder, caused by deficiency of coagulation factor VIII (haemophilia A) or factor IX (haemophilia B). The condition is passed on through carrier females, the majority of whom have a factor level high enough to allow for normal blood clotting. However, around 10% of carrier females are symptomatic and at risk of abnormal bleeding. **Case presentation:** This case report describes a person with mild haemophilia A who, on first presentation to the haemophilia service, stated he was a transgender person in transition to becoming a male. Haemophilia was diagnosed when heavy bleeding occurred following bilateral mastectomy approximately 25 years previously. He now requested phalloplasty. **Management and outcome:** Phalloplasty was performed at a hospital geographically separate from the haemophilia centre, requiring careful coordination between the two services. A haemophilia specialist nurse provided education and training about haemophilia and its management to the surgical nurses. Twenty-four-hour support was available from the nurse and a specialist doctor. Preparation and administration of clotting factor was the responsibility of the haemophilia nurse until the surgical team was confident in its use. Clotting factor replacement was managed using standard procedures, successfully maintaining factor VIII above a target level of 100% with a twice daily dose. Surgery went well, but wound healing was delayed, in part, due to persistent bleeding. **Discussion:** Close collaboration between the haemophilia and surgical teams provided effective prophylaxis of bleeding during a complex procedure that presented new challenges. Both services now have better understanding of the needs of transgender people.



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Many female haemophilia carriers have low factor levels consistent with a mild or moderate bleeding tendency, and may experience significant bleeding episodes. For transgender people with haemophilia transitioning from female to male, this raises issues around their care both during and after transition.

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Box 1. Terminology [3]

Transgender
Adjective to describe a diverse group of individuals who cross or transcend culturally defined categories of gender. The gender identity of transgender people differs, to varying degrees, from the sex they were assigned at birth.

Transsexual
Adjective (often applied by the medical profession) to describe individuals who seek to change or who have changed their primary and/or secondary sex characteristics through feminising or masculinising medical interventions (hormones and/or surgery), typically accompanied by a permanent change in gender role

Gender dysphoria
Distress that is caused by a discrepancy between a person's gender identity and that person's sex assigned at birth (and the associated gender role and/or primary and secondary sex characteristics)

Gender identity
A person's intrinsic sense of being male (a boy or a man), female (a girl or woman), or an alternative gender (*eg* boygirl, girlboy, transgender, genderqueer, eunuch)

Gender nonconformity
The extent to which a person's gender identity, role, or expression differs from the cultural norms prescribed for people of a particular sex

Transition
Period of time when individuals change from the gender role associated with their sex assigned at birth to a different gender role. For many people, this involves learning how to live socially in the 'other' gender role; for others this means finding a gender role and expression that is most comfortable for them. Transition may or may not include feminisation or masculinisation of the body through hormones or other medical procedures. The nature and duration of transition is variable and individualised.

Keywords: *Carrier, haemophilia, surgery, transgender*

The World Health Organization has identified three challenges to the health and wellbeing of transgender people (see Box 1 for terminology): lack of evidence on the determinants and status of their health; lack of understanding about transgender-specific healthcare and preferences, and barriers to access; and social exclusion mechanisms that undermine the right to health [1]. From the health perspective, guidance on medical and surgical care has been published by the Endocrine Society and the World Professional Association for Transgender Health [2,3]; however, transgender people still face prejudice and legal obstacles that obstruct access to services in many countries [4]. Consequently, transgender people are at risk of poor physical and mental health.

It is difficult to estimate the prevalence of people who identify as transgender. Studies, mostly in economically developed Western countries, suggest that the prevalence of transgender individuals who seek care from specialist services is 1:11,900–45,000 for male-to-female individuals and 1:30,400–200,000 for female-to-male individuals [3]. However, studies published since 2012 show that the proportion of people who identify as transgender is much higher – 0.5–0.9% in adults and 1.2% in high school students [4] – and the number of people referred to specialist clinics in the UK has been increasing rapidly for several years [3,5,6]. Where services are available and individuals seek care, clinical management includes psychological assessment and support, hormone therapy and sex reassignment surgery [2,3,7]. There is no published information on the possible risks of hormone therapy in a transgender person with

J Haem Pract 2018; 5(1). doi: 10.17225/jhp00128

haemophilia. Additional planning is required for any person with haemophilia undergoing surgery in order to maintain adequate clotting factor coverage during the procedure and postoperatively. The recommended dosage and duration for haemostasis depends on the type of surgery performed, and is based on estimated blood loss during surgery and haemoglobin levels pre- and postoperatively. In major surgical procedures haemostatic support is required for five days consecutive or more.

These findings suggest that transgender people may have been reluctant to seek care in the past but, where cultural and economic factors make it possible, they are increasingly likely to come to clinical attention. Clinicians in all specialties who have previously been unaware of their patients' gender identity are now more likely to encounter individuals who openly identify as transgender. This is important in the management of haemophilia, which is a deficiency of clotting factor (factor VIII in the more common haemophilia A, factor IX in haemophilia B), for which the provision of clinical care is greatly determined by gender. Haemophilia is categorised as mild, moderate or severe according to the deficit of the clotting factor.

Males with severe haemophilia have a high risk of spontaneous and prolonged bleeding, and receive prophylactic factor replacement to reduce bleeding risk as standard of care. Those with mild haemophilia are less likely to experience bleeding unless challenged by trauma, including surgical procedures. Women with haemophilia have traditionally been categorised as asymptomatic carriers but it is now recognised that many have low factor levels consistent with a mild or moderate bleeding tendency; they may experience significant bleeding episodes, in particular severe menorrhagia [8]. Gender is also relevant to counselling about carrier status and the assessment of bleeding risk in daily life and during surgery.

This case study describes how surgical and medical services were coordinated for Eren (not the patient's real name), an individual with haemophilia who attended the haemophilia centre at the Academic Medical Center in Amsterdam, when it assumed responsibility for his care following a service reorganisation.

CASE PRESENTATION

Eren had undergone surgery at the VU University Medical Center in Amsterdam, where the Netherlands' first transgender clinic is based. Until 2013, the VU Center had also housed a haemophilia treatment centre. The haemophilia service was subsequently relocated to the Academic Medical Center (AMC), which Eren and his wife first attended in 2015. He informed the centre of his transgender status at the first appointment, saying he was born female but from a very young age had felt he did not belong in a female body. He said he was a haemophilia A carrier and his factor VIII level was 24%.

Eren was born in a small village in eastern Turkey and given the name Esila. From the beginning he felt different and preferred to play football and other rough games with his brothers rather than play with his sisters; he never wanted to wear a dress. People treated him like a little boy.

When he was eight years old, he moved to the Netherlands with his father while his mother stayed in Turkey. He went to school and learned the Dutch language. He was increasingly conscious that he felt like a boy, but did not have a close relationship with his father and did not share his feelings with him. After a few years, his mother joined them in the Netherlands and he was able to tell her how he felt. She encouraged him to seek medical help. At that time he was not aware he was a haemophilia carrier. He said he had nosebleeds, some of which had been managed by cauterisation, but he did not have excessive menstrual bleeding. His factor levels had never been checked and there was no family history of haemophilia.

MANAGEMENT AND OUTCOME

Eren underwent a bilateral mastectomy in 1982. Following the procedure he experienced a major bleeding episode, prompting the investigation which identified his low factor VIII level. He responded well to treatment with desmopressin and this was used as prophylaxis to cover subsequent operations. At the time of his consultation at the AMC he was ready to undergo phalloplasty, a genital reconstruction procedure that would begin the completion of the physical aspects of his transition. The procedure involves transferring tissue (a flap of skin, veins and nerves), usually from the forearm, to construct a penis and urethra [9]. The forearm then requires a skin graft, which is taken from the thigh. Testicular and penile implants are inserted after three and 9–12 months respectively. Phalloplasty could be carried out at the VU Center, but surgery had been delayed when its haemophilia resources were relocated to the AMC. The two sites are 12 kilometres apart, presenting a challenge to coordinating haemophilia management and surgical care. However, Eren was highly motivated and a management plan was developed.

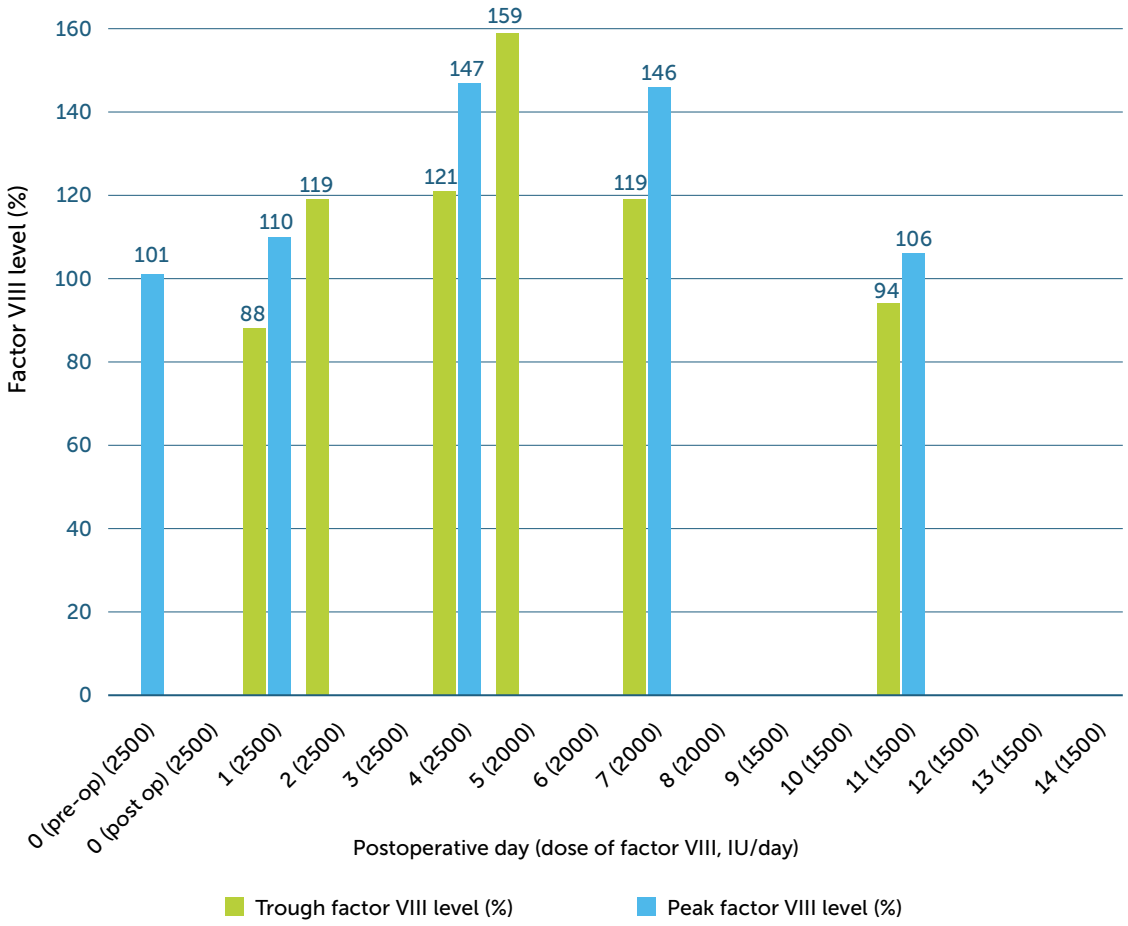

Figure 1. Total daily dose of factor VIII administered on day of surgery and postoperatively, with trough and peak levels

The haemophilia nurse delivered a presentation on the principles of haemophilia care to the nurses on the surgical ward, in which it was explained that the bleeding risk would be reduced by twice daily infusion of factor VIII, aiming to maintain a level greater than 80%. Medical and nursing staff agreed to divide responsibility for care between the surgical team, who would manage the phalloplasty, and the haemophilia team, which would manage factor replacement therapy.

Supplies of factor VIII were provided by the AMC haemophilia centre. Arrangements were made for access to laboratory measurement of factor levels at all times. The surgical ward nurses were provided with the personal contact details of the haemophilia specialist nurse so that any problems could be rapidly addressed. Support from the haemophilia medical team and specialist nurse was available 24 hours a day.

On the day of surgery, Eren was given a bolus injection of 2,500 IU factor VIII (Refacto) ten minutes before the procedure began; a blood sample taken after 10–15 minutes showed a level of 101%. Factor VIII was administered twice daily for a total of two weeks, maintaining the factor VIII level at >80% for 7 days without difficulty, and reducing to >50% from day 7 to day 14 (see Figure 1).

The haemophilia specialist nurse prepared and administered the morning dose of factor VIII and supported a surgical nurse to do so in the evening; after four days, the surgical nurses took over this role fully. Surgery and recovery proceeded well, although healing of the graft site on the thigh was delayed by persistent oozing and bleeding. This had resolved by the time Eren was discharged on day 14, and the wound was subsequently managed with daily bandaging. At outpatient follow-up after six weeks, the factor VIII level was 37%. At follow-up after one year, Eren was very happy with the result and is now awaiting surgery for implantation of testicular and penile prostheses.

DISCUSSION

Clinicians in all specialties are increasingly likely to encounter people who identify as transgender and whose medical and surgical history may present new challenges. This is important for individuals with haemophilia who opt for sex reassignment surgery, because of the need for specialist supervision to reduce bleeding risk during the many procedures necessary, and also for a full understanding of the individual's ongoing management needs. From the perspective of a service provider, the status of a female-to-male transgender person with haemophilia changes from that of carrier to having mild haemophilia, with implications for their future care.

This case shows that the support people with haemophilia need for sex reassignment surgery is similar to the approaches used for other surgical procedures, relying on maintaining trough factor VIII at a target level during the early postoperative phase, with a managed decrease over subsequent weeks. It also highlights the need for general surgical nurses to be educated about the bleeding disorder and, at least during the early postoperative period, to have hands-on support from an experienced haemophilia nurse. Additional medical and specialist nursing support was available at all times, but in this case was not required. Haemophilia nurses can also learn from their surgical counterparts about the challenges anticipated for specific procedures and individual patients, such as the risk and severity of bleeding and the likely duration of support. Each patient with haemophilia who undergoes surgery now has a personal management plan prepared by the doctor from the haemophilia service.

Transgender patients who are in transition are likely to undergo several surgical procedures, which in this case were carried out over a period of years. All disciplines involved therefore acquire experience but, with such cases occurring relatively infrequently, expertise may be lost. It is therefore important that effective working relationships between the haemophilia and surgical teams are established at the outset to ensure that information is shared, that all parties are sufficiently informed, and that lessons can be learned.

ACKNOWLEDGEMENTS

Writing services were provided by Steve Chaplin, Haemnet.

Informed consent has been obtained from the individual reported in this case study. The authors have advised no interests that might be perceived as posing a conflict or bias.

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