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NURSE FOCUS

Living, Caring, Learning – The treatment centre as family for a woman with severe haemophilia

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A nurse at Israel's National Hemophilia Center for over 35 years, Dalia reflects on lifelong relationships with patients and the 'family approach' to care in the context of her experience of caring for a woman with severe haemophilia A. Having cared for the patient since she was a young child, Dalia describes the difficulties she has faced and the importance of ongoing conversations about her life in shaping her care, including risks around pregnancy and having a child. The patient went on to have a son by caesarean section, covered by factor VIII and tranexamic acid; her son was diagnosed with severe haemophilia A at birth and Dalia now sees them both in clinic. The patient also contributes her view on the importance of the close therapeutic relationships she has with Dalia. While acknowledging the importance of setting boundaries, Dalia reflects on the role of the haemophilia centre as an extension of the patient's family and the nurse as a 'mother' figure. She feels the centre's family approach is reflected not only in the care team's knowledge of their patients but also through close and effective working relationships between staff, built on experience sharing, discussion and mutual support. Dalia believes it is this combination that has made her centre so successful in its involvement in clinical trials.

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I have been a nurse for 43 years and Head Nurse at the Israeli National Hemophilia Center since it was established in 1987. I had previously worked full time on a medical ward and was looking for a part-time role to give me time to gain academic qualifications at the university. I had treated a patient with haemophilia and HIV on the medical ward who sadly died – I found it very emotional, but it was through my contact with this patient that I became interested in haemophilia care. When I first started working at the haemophilia centre, there were two doctors, a part-time social worker and me.

In Israel, early diagnosis of haemophilia is common. Male patients often present with bleeding after the first surgical challenge, circumcision, performed in Jewish boys at the age of eight days, so we get to know them and their families very early. Our relationship with them continues throughout their lives, supporting early prophylaxis and providing physiotherapy, medical consultations and psychosocial support groups for various ages. Today, the Israeli National Hemophilia Center provides comprehensive care to over 700 patients with haemophilia (adults and children) and another 1,500 patients with a range of bleeding disorders. The first babies I treated are now 35-40 years old!

LEARNING FROM A GIRL WITH HAEMOPHILIA A

I have more than 700 haemophilia patients in my care, but only four women with severe haemophilia. I've known Noa (pseudonym) for over 30 years and she has taught me so much about what it's like to be a girl, and then a woman, with haemophilia – their medical care, ethical dilemmas and reproductive choices [1,2].

Noa was diagnosed with haemophilia A at the hospital near where she was living, but as she had repeated knee haemarthrosis from an early age, she was referred to the centre at Ramat Gan when she was about two years old. There was no haemophilia in Noa's family and she was diagnosed with an inversion 22 mutation carrier, with X inactivation (lyonization). She always wanted to do everything that other children did – especially the boys – and she had a lot of injuries. In the early days, we didn't have prophylaxis so we gave her factor treatment on demand when she had bleeds.

When Noa was nine, I started teaching her the principles of self-infusion, but she never really learned how to do it properly. She went to the summer camps organised by the Israeli Haemophilia Association with the boys with haemophilia and played football with them. By then, she was on prophylactic treatment but was still bleeding significantly, especially during her menses, and she had a major bleed into her abdominal cavity following ovulation. She would come to the unit twice a week for her prophylaxis, and more often when she was menstruating. These visits were obviously essential for her treatment, but it also gave us the opportunity to talk about different aspects of her life, including family, educational and financial issues. We also talked a lot with Noa about the risks of becoming pregnant, the likelihood of her having a child with haemophilia and what this could mean for her.

CHALLENGES OF PREGNANCY

When she was in her early-mid-twenties and recently married, Noa decided she wanted to become a mother. The medical team were concerned about this as her extreme lyonization might 'cover up' another X-linked disorder. I and several other members of the team, including the doctor, psychologist and social worker,

NOA'S STORY

My name is Noa (pseudonym) and I am a woman with severe haemophilia A. I met Dalia when I was three years old, when I was referred to the Israeli National Haemophilia Center from another hospital due to a major bleed. Since then, the center has become my second family. As my haemophilia was a sporadic case and there was no haemophilia in my family, everything was new to me. Dalia treated me like a daughter and has taken care of me for 35 years.

Dalia has always had so much patience with me. She not only treated me and trained me but also encouraged me in difficult times and gradually became like a second mother to me. She tried to teach me self-infusion for many years but I constantly refused to do it because of fear.

During my pregnancy she supported me in various ways. She spoke to me every day. It was a journey we went through together until I gave birth to a boy with haemophilia. Only when I became a mother did I understand that I couldn't avoid self-infusion anymore. Dalia was so supportive, and by the time my son was three years old I succeeded in overcoming my fear and started self-infusing.

My son is eight now and we visit the center at least twice a month. To be honest, I sometimes miss Dalia and it's good to have the opportunity to update her on various things that have happened in our lives. I am very grateful for the treatment I have received from the Haemophilia Center through the years. As a way of giving something back, I volunteer to collect donations to buy special gifts for children who are hospitalised.

discussed the risks of becoming pregnant with her and we recommended that Noa should wait for a fuller genetic analysis to be carried out so we had a better idea of the potential risks. However, she quickly became pregnant. Thankfully, she had no complications during her pregnancy, and her baby boy was born at term by caesarian section aided by Factor VIII (FVIII) and tranexamic acid cover [3]. He was diagnosed with severe haemophilia A at birth and his circumcision was performed under FVIII.

Noa's son is now eight years old and lives as normal a life as possible, including attending karate class! He has received physiotherapy and hydrotherapy by our centre's physiotherapist. Although Noa knows how to self-infuse now, she often comes to the centre for FVIII – I think it's an excuse to come and visit us – so both mother and child come in for prophylaxis with recombinant FVIII. When her son is nine he will be able to go to summer camp – he is eager to learn how to self-infuse so we are teaching him. Noa still calls me occasionally about incidental bleeding episodes, trauma events and other issues. For a while, the team at the haemophilia centre became an 'alternative family' for Noa and her son, though we are doing our best to enable greater independence.

CARING AS A FAMILY

As I've known Noa since she was so young, it's easy to think of her almost as another daughter. I invited her to my house as a child during the school holidays and she came to me for the Sukkot celebrations for harvest. My staff tell me that I am like a mother to my patients and to them, and I like it that way! However, I do recognise the importance of setting boundaries.

Dalia's recommendations for other health care professionals

- Really get to know your patients and their families – building trusted, supportive relationships helps to optimise care
- Take the time to understand the specific issues facing girls and women with bleeding disorders, such as pregnancy and childbirth – it is important that they have the same level of medical attention as boys and men
- Offer support to patients but also set boundaries so that patients and their families understand what you and other team members are able to do

The centre's 'family approach' is the legacy of its founder, Professor Uri Martinovitch. He led from the top with a very warm and supportive manner which our current head, Professor Gili Kenet, continues today. The staff – now 25 people including four nurses – know each other very well and often socialise together. This closeness is something special and it enables us to share our experiences and support each other in providing the best possible care for our patients.

I believe this has also been important in enabling us to work so successfully in haemophilia research. We currently have 13 clinical trials running at the centre, and about 10% of our patients with severe haemophilia are in clinical trials. We are consistently among the high recruiters in the studies we're involved in. When we are approached about a new trial, we get together to discuss it and consider who will be good candidates for inclusion. We know all our patients so well that we can predict who will adhere to the protocol and who will benefit. Five of our patients have been in gene therapy trials and we are now recruiting for another gene therapy study. The fact that we work so well together helps with the smooth running of our studies. We have built trusted and supportive relationships with our patients so they are happy to participate.

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