NURSE FOCUS

Living, Caring, Learning – Changing mindsets and enabling goals for people with haemophilia

Anne-Louise Cruickshank

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In a nursing career spanning almost five decades, Anne Louise has spent over 30 years caring for people with haemophilia in South Africa. She reflects on the importance of prophylaxis and how tailoring treatment to meet individual needs not only helps to prevent bleeds but can make goals and aspirations attainable. Describing her experience of how she supported a young man with severe haemophilia in achieving his ambition to become a paramedic, Anne-Louise demonstrates the importance of the nurse-patient relationship and the role of the nurse in providing individualised care. She discusses advocating for prophylaxis and the need to change mindsets, and highlights the need for further change in access to treatments for haemophilia in South Africa. The patient also describes his journey to achieving the level of fitness he needed to undertake the entrance exam to train as paramedic.

Keywords: *Haemophilia, Prophylaxis, Individualised treatment, Nurses, Professional practice*

In 1991, I began working one day a week at the haemophilia unit in Johannesburg, and I enjoyed it from the start. Having qualified as a nurse in 1975, I had worked in midwifery, intensive care and paediatric specialties, but I knew nothing about haemophilia. I had some amazing mentors, and the patients I cared for also taught me a lot.

When I first started, no patients were on prophylaxis and we held walk-in clinics that were full of people with

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bleeds. My role expanded and gave me opportunities to attend international conferences to learn more about haemophilia and meet other haemophilia nurses. I was awarded a travelling fellowship that enabled me to work with some fantastic nurses in Canada. In 2001, I moved to Cape Town and became Haemophilia Nurse Co-ordinator for three tertiary hospitals with haemophilia centres in the Western Cape, as well as doing outreach to schools and another smaller treatment centre. By this time, prophylaxis was becoming more widespread as a treatment option, but initially only for patients with insurance due to funding issues. We set up outreach clinics, brought a physiotherapist into the children's hospital, and established a haemophilia nurse training programme [1].

Today, most of our patients are on prophylaxis so they have fewer bleeds, and they get care close to where they live. It has taken many years to reach this point. Finances are limited and we needed to change minds and show the benefits of prophylaxis – but it is so rewarding to see what has been achieved.

FROM INPATIENT TO PARAMEDIC

Having worked in haemophilia nursing for over 30 years, I have cared for many inspiring patients. One who shines as a wonderful example of what people with haemophilia can achieve is David (a pseudonym).

David has severe haemophilia A and I met him as a young man in 2001 when he was admitted with a serious knee bleed. He was in hospital for three months, and during this time he told me that he wanted to be a paramedic. Although he was keen, I was not optimistic about his chances of achieving this. When David recovered, he investigated the training he would need and came back to tell me that he needed to be able to run five kilometres a day. I told him we would give it a go, and we started him on three-times-a-week prophylaxis and got the physiotherapist involved. David also went to the gym to improve his fitness.

Initially, David was turned down for the paramedic training programme because of his haemophilia. However, the South African Haemophilia Foundation (SAHF) stepped in and advocated on his behalf – and

DAVID'S STORY

My name is David (pseudonym) and I remember preparing for the Paramedic Programme exam very well. Coming out of hospital after the knee bleed, the exam was only six weeks away, so I had little time to work on my cardio fitness and strengthen my knee. Sister Anne suggested I increase my dosage slightly, prophylaxis at least 30-60 minutes prior to exercising.

I saw the public physiotherapist, who advised me on very light lower limb exercises. The exercise intensity was based on the amount of factor I had available for the month, plus enough for a spontaneous bleed in my target joint. Twelve boxes of FVIII in the initial stages were not enough – bleeds can be more frequent when your muscles are weaker. So I gradually continued building, rationing my prophylaxis until my joints were more stable.

The key exercises that worked were 10-30 minutes on the elliptical trainer five days a week, followed by light lower limb exercises (leg extensions, side raises, leg raises, assisted squats, deadlifts, plank) which gradually improved over time. The elliptical trainer was the most important – it meant I was able to strengthen the important muscles supporting my joints without any impact on my knees (unlike with regular running). I would go on it for five minutes initially, which was brutal, and eventually ended up at 30 minutes a day (approximately 6km) which made me fit enough for the entry exams.

With haemophilia, I think the key to keeping fit and well is to stay consistent with prophylaxis. It leads to better quality of life and, in the long term, costs less money than having to treat regular bleeds. I believe spending more on prophylaxis and on haemophilia clinical trials in South Africa would cut National Health Insurance costs.

Prophylaxis is not easy for patients. Try telling five-year-olds that they need to stick a needle in their arm twice a week – and it can be worse for teenagers, with all their peer pressures. From my own experience, though, I am convinced we should focus on prevention, not rehabilitation. I think maintaining good prophylactic treatment could mean we avoid spending millions on joint replacements for people with haemophilia with chronic pain and limited quality of life.

he was accepted. The Foundation also supported him financially when it looked like he could not afford to complete his studies.

Since graduating in 2006, David has responded to countless high risk and critical care emergencies as a paramedic. He is completing his Master's degree in Emergency Medical Care, and he is also involved in education in emergency care and first aid. He has travelled widely to assist with emergency care in remote areas, taking his treatment with him. It has not always been easy, and particularly in the early days, some of his colleagues found it difficult to believe that someone with haemophilia could do such a physically demanding job. Both David and I have talked about haemophilia at ambulance training schools, not only in terms of caring for patients with the condition, but also stressing that haemophilia does not necessarily prevent someone from becoming a paramedic.

CHANGING MINDSETS

To give David the best chance of training successfully to be a paramedic, it was important to make sure he was on the correct dose of Factor VIII (FVIII). At the time, limited funding meant that, patients were prescribed 7 x 500 IU of FVIII per month. It was a plasma-derived, intermediate purity factor made locally from donations at the blood bank. In truth, there was probably nearer 600-800 IU per vial – clinicians were unwittingly giving patients more than the set limit. Even so, it really was not enough, and so with the team we gradually increased the dose for David and other patients in my care to 800 IU, and the number of vials to 10 and then 12 per month, to prevent bleeds.

To ensure we could continue to do this, I had to change mindsets. I had to convince those who were rationing treatment that it was much better to spend money on factor for prophylaxis and preventing bleeds than for treating bleeds. I also needed to convince patients of the benefits of effective prophylaxis and regular attendance for clinic appointments. Some still only ask for a certain amount of factor when I know they should have – and need – more. I tell them that as factor is no longer rationed in the same way, they can have the dose they need to prevent them getting bleeds and help preserve their joints.

Prophylaxis has transformed haemophilia nursing practice in South Africa, but we are still limited in how we treat it [2]. We do not have recombinant factor or products with an extended half-life, and only a very small number of children and a few adults are on emicizumab. We are trying to get emicizumab onto the essential drug list at the hospital to ensure that it is generally available.

ANNE-LOUISE'S RECOMMENDATIONS FOR OTHER HEALTH CARE PROFESSIONALS

- Absorb all the knowledge you can and be prepared to grow into your role. We all face challenges but, as nurses, we are not alone – you can always find support from your peers
- Be sure to build good relationships with the patients in your care, managing them all equally
- Have your own ideas and make suggestions because this is how we can make haemophilia care better – we need to share our knowledge and experience
- Get involved in training the next generation of haemophilia nurses. Leaving that footprint and knowing you are making a difference is hugely rewarding
- Always be open to change because nothing remains the same. When you have been in a role for a long time, it can be tempting to continue in the same way – but we need to be open to fresh ideas from our younger colleagues

TAKING RESPONSIBILITY

From my experience of caring for David, I learned the importance of being positive and of persevering to make things happen. Over the years, I have also learned to push back when patients do not follow my advice. I can advise them to do what is in their best interests, but if they decide not to follow my advice and the plan chosen does not work or get the best outcome, they have to take responsibility. I walk their journey with them and would never give up on a patient, but there is a balance to be struck. As a haemophilia nurse, I will take patient calls after hours, and I will advocate on behalf of patients to make sure they do not end up waiting with serious bleeds in a busy emergency department. However, I believe that people with haemophilia need to live their lives, make informed decisions and be responsible for their actions.

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