NURSE FOCUS

Living, Caring, Learning – Collaborating across borders to support a haemophilia patient with a rare complication

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Cyrus, a haemophilia nurse in Kenya, reflects on his experience of treating a patient with a rare complication of haemophilia in a setting with limited access to treatment. The patient developed a debilitating abdominal pseudotumor following an inadequately treated traumatic bleed. After consulting with experts in other countries, surgery was deemed too great a risk. Initiating FVIII prophylaxis with the aim of shrinking the pseudotumour was identified as the most appropriate approach to treatment, however supply issues made it difficult to maintain a prophylactic regimen. When the pseudotumour continued to grow, posing a risk to adjacent organs, an international partnership enabled the initiation of emicizumab prophylaxis through a clinical trial. It was hoped that emicizumab would prevent further bleeding into the pseudotumour and allow it to shrink over time. The patient continued to experience breakthrough bleeding and was withdrawn from the trial after 2.5 years. By this time the supply of FVIII to Kenya through the through the WFH Humanitarian Aid Program had increased and the patient was able to resume FVIII prophylaxis. The pseudotumour is now stable and he continues to be cared for by the same nursing team. Reflecting on this case, Cyrus highlights the value of sharing expertise across borders and working in partnership to try to ensure the best treatment outcome for a patient with a rare complication in a resource-constrained setting. He also reflects on being compassionate in his practice and the importance of a holistic approach to patient support.

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In 2012, I graduated as a Registered Nurse from Moi University in Eldoret, Kenya. A year after completing my internship, I joined the AMPATH [1] (Academic Model Providing Access to Healthcare) programme at Moi Teaching and Referral Hospital (MTRH). AMPATH is a collaborative partnership between MTRH, Moi University College of Health Sciences (MUCHS), and a consortium of North American Universities led by Indiana University. MTRH and MUCHS share a campus in Eldoret in western Kenya, about 300km northwest of the capital, Nairobi. It's the second largest tertiary public health hospital in Kenya and one of two comprehensive haemophilia treatment centres (HTCs) in the country. AMPATH covers MTRH's catchment area, and over the last 30 years or so has expanded, growing significantly in the early 2000s as a response to Kenya's HIV pandemic, and now including care for a range of chronic conditions.

I have benefitted from involvement in AMPATH's health research programmes and its focus on developing health care leaders. Importantly, our patients benefit from this too. Over the last seven years, I have been involved in the sickle cell and haemophilia research and care programmes, and I now have a senior role in the wider non-malignant haematology programme, mainly focusing on sickle cell disease, haemophilia and other rare bleeding disorders. It's been a transformative journey for me, with so many changes and opportunities. I am still very much involved with patient care, outreach and support, and it is a privilege to work across so many aspects of haemophilia care for the 270 patients who attend our HTC.

I'm going to share the story of one of the inspiring patients who has taught me so much about putting all the pieces together to optimise care. This experience also taught me that, even when we don't succeed in the way we had hoped, our patients benefit from knowing that everything possible has been done to help them.

A RARE HAEMOPHILIA COMPLICATION

Moyo (a pseudonym) has moderate haemophilia A. As a young man, he had a serious accident at work and subsequently developed an iliopsoas muscle bleed. This was inadequately treated due to challenges in accessing coagulation factor concentrates (CFCs). At the time, the supply of CFCs we had access to in Kenya was solely through donations from the World Federation of Hemophilia (WFH) and Indiana Hemophilia and Thrombosis Centre (IHTC) [2,3].

Moyo went on to develop a large swelling in his lower abdomen and his health deteriorated. On further assessment we established he had a pseudotumour, a rare complication of haemophilia where an encapsulated haematoma forms following recurrent bleeding. It was the first time the team had seen this complication and thanks to our AMPATH partnership we were able to get expert advice from the IHTC team. We also had discussions with specialists in other HTCs including in Italy, Spain, Argentina and Israel. Everyone agreed that surgery as an option was too risky, and we started to treat Moyo with factor VIII (FVIII) prophylactically to try to shrink the pseudotumour [4].

As a country, we did not have enough CFCs to meet our on-demand treatment needs, let alone support a prophylaxis programme. The limited and intermittent supply meant we just couldn't access enough FVIII to sustain prophylaxis for Moyo. This meant the outcomes were not as we had hoped and the pseudotumour continued to grow. Moyo's physical activity was extremely limited due to the pain and swelling it caused. We were very concerned as it was also displacing and compressing adjacent internal organs, threatening to compromise their function. Home visits were an important part of the care we could give, and we saw that the situation put a lot of pressure on both Moyo and his family. In addition to his treatment needs and the debilitating physical impact of the pseudotumour, it was evident that he needed support for his psychological wellbeing and the social and financial impacts too.

THE VALUE OF INTERNATIONAL PARTNERSHIP

At this time, the IHTC had initiated a clinical trial for treating pseudotumours with emicizumab. They very generously flew Moyo to the USA for further assessment and treatment, and he was enrolled in the trial. Emicizumab mimics FVIII but behaves slightly differently – it has a longer half-life and provides constant coagulant activity equivalent to FVIII levels of 15-20% [5]. It was hoped that prophylaxis with emicizumab would prevent further bleeding into the pseudotumour, allowing it to shrink over time, improving the pain and swelling Moyo was suffering, and relieving the compression on his other organs.

When Moyo returned home, he was followed up by our research team at MTRH, in line with the trial protocol. Sadly, again, things didn't go as hoped. Moyo had frequent bleeds despite prophylaxis with emicizumab and the pseudotumour increased in size. On top of this, CFCs to treat breakthrough bleeding remained in very short supply. After 2.5 years, he was withdrawn from the trial.

Since this time, the supply of CFCs through the WFH Humanitarian Aid programme has increased and Moyo has been able to resume prophylaxis with FVIII infusions. The pseudotumour is stable and he has reported feeling better. He continues to be cared for by the team at MTRH. Progress is also now being made to ensure the Kenyan government is able to offer expensive coagulation factors to the people who need them.

EXPANDING HORIZONS

My involvement in Moyo's care taught me a lot about the importance of a holistic approach – not just considering his physical needs but also the wider impact on his wellbeing and that of his family. Home visits as part of outreach activities are an integral part of healthcare delivery. I learned a lot about being compassionate and gaining greater understanding of what our patients go through.

This situation also challenged me to look further afield for help for patients and not to be limited by the space in which we work. If we don't know what to do for a patient there is always somebody somewhere who has the experience to advise the best course of action. If we hadn't explored all the opportunities for Moyo, I don't think he would have received the care he needed. Even though emicizumab didn't work for him, he knew that we were all behind him, doing everything we could to help – not just us, but his family, friends and community too. I think all of this contributed to the fact that he feels better now and is getting on with his life. He needs crutches to get around but he now runs his own small business and he can support his family. He is building a freedom for himself, even though we couldn't achieve the perfect outcome for him.

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CYRUS'S RECOMMENDATIONS FOR OTHER HEALTH CARE PROFESSIONALS

- Listen to patients and their families and really try to understand their situation before you make decisions about optimal care – then combine this with your professional knowledge to give care based on what is in their best interests
- Showing love and compassion brings responsibility but also bring rewards for your patient and for you
- Always be open to new ideas and be ready to contact experts in your own country or abroad who may be able to help or give advice – make good use of options for online discussions and consultations if it is difficult to meet in person
- As individuals, always aim to be better, do better and do good.

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