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NURSE FOCUS

Living, Caring, Learning – Thinking outside the box to solve care challenges in a rare blood disorder

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SANDRA HIBNER is a Member of the Board of the Southwestern Ohio Hemophilia Foundation (SWOHF) and Family Annual Meeting of Hemophiliacs in Ohio (FAMOHIO)

A specialist nurse for over 20 years, Sandra reflects on her experience of finding ways to deliver effective care for a young girl with a very rare clotting disorder. Symptoms of the girl's disorder were evident from birth and through an emergency use request Sandra and the care team enabled her family to access a treatment that at the time was in clinical trial. Poor venous access meant there was a need to adapt how treatment was administered and she worked with the girl's parents to ensure that she was treated effectively. Alongside educating the parents, Sandra highlights the importance of her role in educating co-workers and other hospital staff likely to come into contact with the girl, to ensure that she always had access to timely and appropriate care. She also reflects on other instances where thinking creatively enabled patients in her care access to treatments that may not otherwise have been accessible. Now retired, Sandra continues to be involved in advocacy for people with bleeding disorders.

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In the first half of my nursing career I worked in critical care and oncology and, in surgery, as a scrub nurse. In 1997, I had a big change of role and became Coagulation Resource Nurse and Hematology Research Coordinator at Dayton Children's Hemostasis and Thrombosis Center in Ohio. It proved to be a wonderful change for me. The bleeding disorder community is so welcoming, and my colleagues were so supportive and helpful. There have been many opportunities for education, and for interactions with co-workers and other healthcare providers around the

world. I think it helped at the start because we were such a small community but, even with all the changes and advances in haemophilia care of the last 20 years, that friendliness and support has been maintained. I retired in 2019 but I have remained connected to the bleeding disorder community through the Southwestern Ohio Hemophilia Foundation (SWOHF) and the Family Annual Meeting of Hemophiliacs in Ohio (FAMOHIO) and, previously, with the National Hemophilia Foundation and the Global Hemophilia Network Support Committee.

I learned so much from my patients over the years but Mia (a pseudonym) had a big impact on how I cared for my patients and taught me to really think outside the box. Mia was born with a very rare, inherited clotting disorder. Her father was known to be heterozygous for the disorder, having inherited a gene mutation from one parent, but generally did not experience many symptoms. There was no history of a clotting disorder on the mother's side, though she was found to have a rare genetic mutation that was relevant.

Within 24 hours of birth, Mia's left foot became swollen, distorted and turned black. This is called purpura fulminans [1], and it occurs when tiny blood clots form in the skin, eventually leading to skin sloughing that looks like a third-degree burn. At her local hospital, Mia was rapidly given fresh plasma for anticoagulation and, by the time I saw her a few days later, her foot looked normal.

At that time, a purified form of the plasma concentrate that Mia needed was in development and undergoing a clinical trial. We managed to get it for Mia through an emergency use request, and we taught her parents how to give it through a central line, every other day.

Mia progressed well but, when she was about a year old, the clinical trial closed. We decided to remove the central line and move Mia on to oral warfarin and give her injections of a low molecular weight heparin if she showed signs of purpura. It wasn't long before Mia was admitted with purpura but, fortunately, the trial reopened and she was able to get back on the plasma concentrate she needed. It was subsequently approved for general use and Mia has continued using it to this day.

SOLVING ADMINISTRATION PROBLEMS

It was difficult to treat Mia intravenously. She had such poor venous access due to her abnormal clotting, so we needed to find an alternative way of administering her treatment. We started mixing it in a smaller amount of diluent than for intravenous use and giving it subcutaneously via a small pump under her skin. Mia's mum became so good at ensuring treatment was done every other day that Mia did not need any hospitalisations during that time. However, when she reached puberty she started coming in with purpura and it seemed likely she could not absorb enough of her treatment subcutaneously anymore. When we put a port under her skin to give treatment intravenously, we were concerned that she'd get clots around it but fortunately that has never happened. Today, she is doing very well, looking after her port and self-administering treatment.

Over the years, the care team and I have talked with Mia about the risks of getting pregnant [2]– both for her and for an unborn child, which would undoubtedly inherit her disorder in the heterozygous form at best. She knows the risks but it will be her decision whether or not to have a child.

WHEN IT'S RARE, EVERYONE NEEDS EDUCATION

With any patient with a rare disorder, it is important to educate not only the family but people within the hospital who come in contact with them [3]. In Mia's case, this included my co-workers and emergency room (ER) staff. The ER team needed to know that Mia could not wait for two hours to be seen and that if she came in with purpura it was not a normal bruise. We also went to Mia's school each year to talk to her new teachers and other staff members so that everyone had the information they needed.

The lessons I learned about thinking outside the box proved helpful when it came to the care I was able to provide for other patients. Being able to find assistance for my patients provided them with a sense of self-assurance and gave me the courage to try additional avenues when needed. For example, when a patient was told he would have to wait two years to access the Medicaid programme for his treatment for severe haemophilia and an inhibitor because he had not come to us through the standard channels, we opened a clinical trial for him so he could get treatment that way. As a nurse specialising in blood disorders, there were times where issues arose that posed a real challenge to providing the most effective care for my patients – but a bit of creative thinking could often help to overcome them.

Personally, I am very grateful to have met Mia and her family because they taught me so much. To start with, in the early days, I was probably more involved with them than I should have been. When the hospital updated its rules about interacting with patients, I explained that I should step back a little and could

not come to social events anymore. As I approached retirement, it was also important that the family became used to contacting other members of the team and I was pleased to see it all working out so well. After I left, though, I was no longer bound by the rules for staff and I proudly went to Mia's graduation.

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SANDRA'S RECOMMENDATIONS FOR OTHER HEALTH CARE PROFESSIONALS

- When you have a patient with a very rare condition, investigate all you can about it in the literature and reach out to clinicians with similar patients to find out what lessons they have learned
- Make sure that everyone who comes in contact with a person with a rare condition has all the information they need to help keep the patient safe
- Be prepared to think outside the box and come up with novel ways to access resources when none seem to be available

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