NURSE FOCUS

Living, Caring, Learning – Building family relationships in haemophilia care

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With over 35 years' experience of paediatric nursing, almost half of which she has spent in haemophilia care, Robyn reflects on the importance of taking a family-focused approach and engaging with parents and caregivers. She describes her experience of providing care for a family with two boys with severe haemophilia A and inhibitors, and how listening to and working closely with the parents enabled good outcomes. Robyn points to the central role of nurse-patient relationship in haemophilia care but highlights the importance of ensuring that this close therapeutic relationship remains professional.

Keywords: *Haemophilia, Inhibitors, Nurses, Therapeutic relationship, Professional practice*

I rather fell into working in bleeding disorders care! After nursing at the Children's Hospital in Sydney for 18 years, pretty much on the same ward and in the same job, I worked in infection control for six months and realised there was life away from the wards. I went on to take a three-month secondment in haematology to set up a haemophilia treatment centre — families had lobbied government and raised money to make this possible. That was 17 years ago. Today, we care for about 300 patients with bleeding disorders, and I am one of two nurses in the haematology team. I look after people with bleeding disorders while the other nurse cares for those with haemoglobinopathies, though we cover for each other. Our team includes haematologists, registrars and a data manager, and we can access a social worker and refer patients to the psychology department. We also have a physiotherapist who sees patients with a bleed and at clinic appointments. I enjoy my role and I love seeing patients from birth and watching them growing up. I used to be really stretched because I was seeing patients all the time, but as many have changed to emicizumab in the last two years, I now see them less often. Keeping patients and families engaged is still important, though, so that they come to clinic and recognise that they still have a bleeding disorder.

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THE IMPACT OF INHIBITORS AND FINDING SOLUTIONS

I met Ricky (a pseudonym) a few days after he was born. He presented with a large haematoma after hepatitis B and vitamin K injections. There was no family history but his tests confirmed severe haemophilia A, and his genetics suggested he was at risk of developing an inhibitor [1]. After a series of factor-treated bleeds, Ricky's inhibitor was detected when he was seven months old and he had a port put in so we could start immune tolerance induction straight away. Unfortunately, he had bleeding around the port so it was changed to a central line, but skin irritation around the line meant it had to be changed back to a port.

Right from the start, Ricky's mum and dad were both very responsive to what we told them about caring for him. The mum became pregnant again when Ricky was about a year old and gave birth to a second boy with haemophilia, Jack (a pseudonym). Due to the inhibitor risk, we decided Jack should have plasma-derived product rather than recombinant [1], and only have factor when absolutely necessary. Even so, he also developed an inhibitor at the same age his brother had.

Both Ricky and Jack were treated with rituximab. This brought their inhibitor levels down but unfortunately led to hypogammaglobulinemia [2], where the immune system does not make enough antibodies. We treated that with gamma globulin, but both boys had a lot of infections. Again, the parents were fantastic about managing it all. They persevered so well that the boys had very few bleeds.

By the time emicizumab became available, the boys were doing so well that we had some big discussions about whether or not to switch their treatment. After researching it themselves as well, the parents decided they should change over. Since then, I have seen each boy once after a fall; otherwise, there have been no bleeds. The parents have gone from accessing ports every day for plasma-derived factor VIII, to giving subcutaneous injections once a fortnight and flushing the ports every six weeks or so — and we have recently been able to remove the ports.

PARENTS ARE THE BEST ADVOCATES

Knowing the family so well for so long has been incredibly helpful in keeping them engaged, giving them options and support, and getting such good results. The boys have been amazing too – always happy and laughing and ready to play, even though I have to stick needles in them.

ROBYN'S RECOMMENDATIONS FOR OTHER HEALTH CARE PROFESSIONALS

- Plan ahead with families for what is likely to happen in the future so that everyone is prepared, e.g. for bleeds when children become mobile, for inhibitors in those who are at high risk
- Inform patients and families a little at a time while you build a relationship, especially during the first six months after diagnosis. Informing them about needles or ports is important but can be frightening if you show them straight away
- Reassure families that, although there is a plan for treatment, nothing is set in stone and you can be flexible
- Get involved with haemophilia summer camps – seeing the children learn to inject themselves and enjoy all the activities is so rewarding
- Arm yourself with knowledge about research findings so you know about new treatments in the pipeline

The whole experience has taught me the importance of listening to parents. They really are the best advocates for their children. If they say there is something wrong with their child or ask to try something different, as healthcare professionals we need to be open to that and work with them. I have always tried to listen to the patients I care for, and to their families, but Ricky, Jack and their parents showed the really positive outcomes that can be achieved if you really pay attention.

Sometimes it can be hard to engage with families, and what works with one set of parents may not work for another. As experienced nurses, we may be old dogs but we have to keep learning new tricks and be prepared to change. It may just be finding a way to get through to parents — for example, explaining that by coming to the haemophilia treatment centre and seeing us during business hours, they can avoid hours spent in the emergency room when their child has a bleed. We need to understand the family's personal circumstances too — what works for a family that lives down the road from the treatment centre will be totally different from what works for a family living out in the country. Similarly, if both parents work, if they have other family commitments or if they do not have easy access to childcare, it all makes a difference.

GETTING INVOLVED BUT DRAWING A LINE

To gain this level of understanding of patients and their family needs, haemophilia nurses do need to get involved — but we also need to know where to draw the line. It may sometimes feel like we are almost part of the family but I don't want the children to think of me as their 'auntie'. It becomes tricky when families, are so grateful that they invite you to their home – which is where I personally draw the line. It is my job to look after their children and I will do that as well as I possibly can, whoever they are. It can help to explain that we have professional codes of conduct that mean we are not allowed to accept expensive gifts or to meet them socially. Most parents understand and just want to know that we are there when they have problems – which, of course, we always try to be.

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