HAEMOPHILIA AND INHIBITORS 2: SURGICAL MANAGEMENT

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Part 1 of this two-part unit on haemophilia and inhibitors examined the condition of haemophilia, the problem of inhibitors and practical challenges nurses may face in surgical management. This second part examines a case study of a 58-year-old man with severe haemophilia and an inhibitor. Communication and involvement with the patient, caregivers, allied departments and the multidisciplinary team were integral to the procedure’s success. This, in addition to detailed planning and continuity of care, is crucial for a successful outcome.

ORTHOPAEDIC SURGERY
In the past, due to the potential for life-threatening bleeds, surgery in patients with haemophilia and an inhibitor was rarely performed. The advent of bypassing therapies – such as factor VIII inhibitor bypassing fraction and recombinant factor VIIa (rFVIIa) – has made surgery possible for such patients. However, orthopaedic surgery in this group is still a clinical challenge.

Although many authors (Goddard, 2005; Ludlam, 2005) have discussed the difficulties of managing haemophilia patients with inhibitors during surgery and post-operatively, few have discussed the nursing role in overcoming these challenges.

CASE STUDY
History
George Greenwood*, aged 58, had been diagnosed with severe haemophilia shortly after birth as there was a strong family history. Unfortunately, at this time there was no effective treatment for bleeds, and tissue damage caused by recurrent bleeding into the joints progressed to painful, advanced, degenerative haemophilic arthropathy.

George had relatively infrequent treatment for bleeding episodes and developed an inhibitor in his 30s. His musculoskeletal problems were evident from the age of eight and deteriorated over the years. Where someone on effective prophylaxis would expect to have relatively few joint bleeds, George was having more than 120 bleeds into his lower limb joints a year. He was unable to work and became increasingly housebound and isolated, which had a huge impact on his quality of life and psychological well-being.

Although George received his haemophilia care locally, he was referred to St Thomas’ Hospital in 2001. It was evident that without surgical intervention his prognosis was poor, so planning and negotiation with his PCT was undertaken to enable surgery to take place. The plan was to replace both knee joints and perform arthrodeses on both ankles within the same year.

Surgery
In January 2006 a surgical team performed a total knee replacement (TKR) of the left knee. The nursing team was asked to monitor haemostasis throughout the procedure. Haemostatic control was provided by the bypassing agent factor VIII inhibitor bypassing fraction.

At the pre-surgery visit to the centre, a baseline assessment was performed. This consisted of pre-blood sampling and administration of the bypassing agent followed by post-blood sampling.

As well as a thrombin generation assay, tests on the blood sample included a full blood count, liver function tests and coagulation screening.

Post-operatively, the bypassing agent was given as prescribed every 4–6 hours for at least three more doses with pre- and post-bloods taken as directed by the surgical protocol.

One nurse remained with George throughout the procedure, with a second to assist with having the samples labelled, recorded and dispatched to the pathology laboratory. A haemophilia nurse was rostered to be with him overnight for two nights, while daytime treatment and sampling was to be covered by a designated nurse from the centre. This meant there was no need for an ICU bed and that George’s haemophilia care was provided by experienced nurses working in liaison with ward staff.

Outcomes
This surgical intervention was the first of four procedures. Using similar protocols and nursing input, George had a left ankle arthrodesis 10 days later (on the same admission) and returned to hospital in August 2006 to have the same surgery carried out on his right knee and ankle.

The procedures have made a substantial difference to George’s life. Despite immense post-operative pain, he reported within the first 24 hours that the underlying chronic pain was already reduced. At follow-up, three months after his final procedure, he...
was free from analgesia, able to sleep and less irritable. To date, his mobility has improved greatly, he has abandoned his crutches, can stand for long periods and is able to drive a car using the pedals.

As with most patients with haemophilia who undergo joint replacement, bleed frequency into the new joint also decreased significantly, from up to 120 times per year to just 50 per year post-operatively. As a result of decreased pain and reduced bleeds, George began to think and talk about his future, something which, in his pre-operative situation, he did not do.

Below we outline some important challenges that we overcame in this case study. The box (below left) outlines some important issues to consider in planning similar processes.

CHALLENGES AND PRACTICALITIES Communication and involvement
Communication and involvement in specific areas were key to success. Communication was vital with the patient and his caregivers, and within the multidisciplinary team itself, as was early involvement of allied departments such as the pain team, pathology and physiotherapy.

A first step in open communication with and gaining George’s committed involvement was to help him cope with the impending challenge. Both George and his caregivers expressed daily anxiety about the procedures, especially the possibility of physical disability and the fear of post-operative pain. It was vital that they all received information about the procedure and regular one-to-one contact to discuss any concerns or questions.

From the start, communication within the multidisciplinary team was excellent. To aid communication further, this team discussed and planned the corrective surgery of the affected joints over regular meetings.

Planning
Before surgery a protocol was prepared detailing instructions for pre-operative clinic assessments and tests required before, during and after the surgery. It also outlined any important considerations including contraindications for the patient such as the use of antifibrinolytics. During the operation our role was to monitor haemostasis closely.

Working alongside our colleagues in pathology, we performed a trial run of the monitoring, covering practicalities around performing the thrombin generation assay, getting results in the expected time frame and even climbing flights of stairs between theatre and the pathology laboratories. As no ‘runner’ was available, it was agreed that we would deliver samples in half-hour intervals.

Due to the large volume of ancillaries needed for blood sampling, labelling and the FEIBA administration, we decided to prepare dated and timed packs containing everything needed for each occasion. We liaised with theatre staff and were given space within the anaesthetic room to store and prepare what we needed. Similarly, packs were made to be kept in George’s side room for the post-operative period.

Other planning considerations included: organising a pain team assessment; communicating with anaesthetists on the placement of a triple lumen central line for blood sampling; ordering blood resources; liaising with orthopaedic nursing staff; and arranging specialist nursing staff cover for the night recovery period post-operatively.

Continuity of care
Post-operatively, our role was to provide comprehensive care, maintaining the desired level of haemostasis, monitoring and assisting with rehabilitation therapy, and detecting the occurrence of early bleeding.

In recovery, George experienced immense pain. Having the pain team already involved and prior assessment of the patient greatly helped in managing his pain quickly. One-to-one nursing was provided on the orthopaedic ward for 48 hours. This ensured continuity of care through this early stage and also took pressure off the regular orthopaedic ward staff. Nursing staff also had direct access to a consultant’s mobile number in case of emergency.

After the initial post-operative period, the dosing interval (around 12 hours) of bypassing agent was especially beneficial as it was less labour intensive. From George’s perspective, bolus administration meant that he did not need to be attached to a drip or pump. The central line was removed by day seven to reduce the risk of infection.

CONCLUSION
In summary, the availability of bypassing therapy has changed the management of haemophilia patients with inhibitors undergoing surgery.

This case study demonstrates that with planning, multidisciplinary communication and continuity of care, orthopaedic surgery should no longer be avoided due to concerns over maintaining haemostasis, and it can greatly improve quality of life.

For nurses working in other specialties there are many transferable skills and lessons learnt that could be directly applicable to the nursing of other patients with complex needs.

*The patient's name has been changed