Haemophilia: preventing bleeding after surgery

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Congenital bleeding disorders (CBDs) are conditions in which coagulation abnormalities, such as low levels of clotting factors, disrupt haemostasis, the process that halts bleeding from blood vessels. The conditions vary in severity and symptoms: patients with mild CBDs may only require intervention when they undergo surgery, whereas patients with severe CBDs require treatment three or four times per week to prevent spontaneous bleeding. Conditions include haemophilia A and B and Von Willebrand disease.

The aim of treatment for patients with CBDs who are undergoing surgery is to correct the coagulation defect for as long as the bleeding risk persists and until wound healing is complete. Treatment regimens include intravenous infusions of specific clotting factors.

A complication of some treatments is the development of an inhibitor (antibodies that significantly reduce the efficacy of treatment). Patients with inhibitors are treated with bypassing agents such as activated prothrombin complex concentrates and activated recombinant factor VII (rFVIIa) to prevent bleeding.

Although haemostatic regimens for surgery are usually devised by specialists, surgical nurses are often responsible for administering treatment and monitoring for complications post-operatively.

Objective

This Cochrane review aimed to assess the effectiveness and safety of different haemostatic regimens administered in people with a CBD for preventing bleeding complications during and after surgical procedures.

Method

The interventions were defined as any haemostatic treatment regimen compared with no treatment or to another active regimen. Four trials (112 participants) were eligible for inclusion.

Primary outcome measures were defined in terms of mortality and blood loss.

Results

Two trials looked at patients with haemophilia A or B undergoing dental extractions with the use of antifibrinolytic agents and replacement treatment for haemostasis support. The use of antifibrinolytic agents reduced blood loss and the requirement for post-operative replacement treatment compared with placebo.

Two further trials considered patients with haemophilia A and B with inhibitors treated with different regimens of rFVIIa for haemostatic coverage of surgery. The results revealed a higher post-operative haemostatic efficacy when high-dose (90µg/kg) regimen of rFVIIa was used as compared with the low-dose regimen (35µg/kg), resulting in shorter duration of treatment, lower number of injections but similar total dose of rFVIIa.

Conclusions

The trials in this review provide some information in two specific situations in people with CBDs undergoing surgery. However, on the whole, there is not enough evidence to define the best treatments for the various types of disease and types of surgery. Consequently, treatment regimens are often based on local protocols developed by expert clinicians and on information from observational uncontrolled studies.

There are numerous reasons for this, one of which is that the rarity of the conditions makes it difficult to undertake trials with adequate sample sizes. Limitations in the availability and safety of replacement products after the transmission of HIV and hepatitis in contaminated blood products also made surgery difficult in the mid-1980s. Finally, it is argued that excellent clinical outcomes have been achieved with non-plasma-derived factor concentrates that have reduced the need for rigorous trials to consider optimal treatment regimens.

However, it is acknowledged that trials that investigate cost-effectiveness of treatment regimens in patients with CBDs undergoing surgery are now needed.

Implications for practice

Some conclusions can be drawn from this review but they apply only to specific clinical settings for patients with CBDs. As with other rare diseases, obtaining rigorous evidence is challenging. Nevertheless, it is important that nurses who are involved in the care of patients with CBDs in the surgical setting engage with specialist teams to understand the rationale behind treatment regimens they are required to deliver.

It is also worth noting that there are currently a plethora of clinical trials being undertaken to improve treatment for patients with CBDs - namely genetic therapies, subcutaneous and longer-acting products. These treatments have the potential to substantially improve both quality of life and morbidity within this patient group.

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