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Focus on musculoskeletal health in women with bleeding disorders

Alison Dougall, Laurent Frenzel

Maintaining good musculoskeletal health, including good oral health, is as important for women with bleeding disorders (WBD) as it is for men. Many people with bleeding disorders ignore bleeding from their gums, believing it to be part of their condition. However, it may be a sign of periodontal disease, which left untreated can lead to accelerated tooth loss and infection, adversely affecting overall health. A good diet and access to good dental care from childhood are important to maintaining good oral health in WBD. Joint bleeding and degeneration are not limited to people with more severe forms of haemophilia; joint-related diagnoses have been shown to be twice as common among haemophilia carriers and women with mild haemophilia than in the general population. Women with type 3 von Willebrand disease experience comparable joint outcomes to younger intensively treated patients with severe haemophilia. Neither gum nor joint bleeds should ever be considered normal, as both can be treated to avoid progressive disease. Dental and joint specialists and physiotherapists should work closely with haemophilia teams to ensure optimal care for long-term preservation of musculoskeletal health.

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ORAL HEALTH IN WOMEN WITH BLEEDING DISORDERS

Maintaining good musculoskeletal health, including good oral health, is as important for women with bleeding disorders (WBD) as it is for men. Periodontal disease is the most common chronic inflammatory condition, affecting 50% of the world's adult population [1]. Left untreated, it can lead to accelerated tooth loss and infection and adversely affect overall health.

Inflamed and bleeding gums (gingivitis) are a sign of early gum disease which can progress quickly to periodontal disease in susceptible individuals. 'Pockets' form between the gums and the teeth which can trap bacteria and food particles, leading to inflammation. People with bleeding disorders are likely to bleed for longer with greater impact on oral health than those without bleeding disorders. However, many ignore bleeding gums early on because they assume it is part of their disease (Figure 1) [2]. They may avoid brushing affected areas and this makes inflammation worse. As periodontal disease progresses, teeth loosen and eventually fall out or have to be extracted.

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Figure 1. Beliefs and behaviours of people with von Willebrand disease who experienced bleeding from their gums [2]

Described themselves as having good oral health despite reporting bleeding from the gums after brushing
Thought they had little control over the bleeding of their gums
Thought bleeding from their gums was from using too hard a brush or poor oral hygiene technique
Often attributed bleeding from their gums to trauma and spontaneous bleeding
Stopped brushing in the presence of blood

Simple measures can be used to treat gum bleeding. Breaking the cycle of inflammation and bleeding is key. Brushing gently once or twice a day, especially at night, is recommended; this can be supported by the topical use of tranexamic acid. Mouthwashes are less helpful as they do not remain on inflamed areas for long enough, and anti-bacterial products may kill useful as well as harmful bacteria. Plaque disclosing tablets can be a useful way of showing a build-up of plaque and should be seen as a training – not a shaming – tool. If extractions are needed, tranexamic acid on gauze is again helpful, as is eating cold foods such as ice cream. A soft diet is recommended for three to five days.

The importance of good diet

A healthy, balanced diet, low in refined and sugary foods and drinks is important for good oral health. However, some trends towards 'healthy eating' have proved counterproductive, e.g. replacing sugar with honey or maple syrup for sweetening, and snacking on fruits that are high in sugars, such as raisins, instead of biscuits. Many seemingly healthy drinks, such as 'pure' fruit juices and smoothies also have a high sugar content and should not be consumed in large amounts.

CASE STUDY

Anna (pseudonym) has type 2 von Willebrand disease and her bleeding gums are so severe that she feels she frequently looks like a vampire. Her gums can start to bleed without any obvious trigger – she does not need to use a toothbrush to make them bleed. If she is wearing a protective mask, it can be soaked with blood in just a few hours. She assumed that the bleeding was normal for someone with a bleeding disorder and was only recently advised to seek a referral to a periodontist to treat her gum disease.

Lifelong dental care

Good dental care is essential at all stages of life. During pregnancy, even the 'best mouth' can be affected by gingivitis. New parents should be given advice about dental care around teething and every infant should be seen by a dentist before their first birthday, especially if they have a bleeding disorder. During childhood, they need good experiences with dentists to avoid developing phobias about going to the dentist later in life, and parents should be given accurate and reliable advice about teeth straightening and sealing.

Only a complete lack of gum bleeding is normal, even in type 3 von Willebrand disease, and anyone who regularly spits blood should seek dental care. It is never too late to seek help, and the earlier a person receives treatment for gingival disease, the better. With good dental care, every woman should have all her teeth in later life, irrespective of whether she has type 3 von Willebrand disease or another bleeding disorder. Guidelines are available and dental care is safe for patients with a wide range of bleeding disorders if appropriate measures are taken [3,4].

Other oral conditions

WBD may experience other oral conditions that should be rapidly diagnosed and treated. Temporomandibular pain, caused by muscle spasm, is four times more common in women than men and can be treated with soft diet and jaw rest [5]. Arcoxia (etoricoxib) for two to three weeks can be helpful while the jaw is rested.

Burning mouth syndrome is a well-documented condition and should not be ignored [6]. It is often a sign of anaemia and may arise as result of reduced oestrogen levels after the menopause. It should be treated with oestrogen rather than anti-depressants, as often happens [7,8].

Inadequate dental services

A shortage of dentists and a failure to integrate dental care with other forms of healthcare services in many countries means that many WBD do not receive the dental care they need, especially preventive measures. This situation was exacerbated during lockdown stages of the Covid pandemic, with dental practices closed for all but emergencies. Patients are also more reluctant to pay for dental care than medical care and may therefore miss out on regular checks when free consultations are not available.

CASE STUDY

Chantal (pseudonym) is 32 and has Glanzmann's thrombasthenia. She has never been to a dentist because no one told her she should. She was advised against dental treatment because of her bleeding disorder. Her oral bleeding is now is so severe and her periodontal disease so advanced that she will need to have all her teeth removed.

It is essential that haematologists are aware of the importance of referring patients to periodontists who are expert in treating gum disease and, where appropriate, performing implants. It is also imperative that patients no longer assume that gum bleeding is an unavoidable consequence of bleeding disorders.

JOINT AND BONE HEALTH IN WOMEN WITH BLEEDING DISORDERS

Joint bleeding and deterioration are not limited to people with more severe forms of haemophilia. Among 539 haemophilia carriers in Sweden followed up over 22 years, around 40% of whom were known to have mild haemophilia, joint-related diagnoses were at least twice as common as in the general population and first diagnosis occurred at a significantly earlier age [9]. Although numbers were small, joint surgery was 10 times more common in those with reduced factor activity.

Women with type 3 von Willebrand disease experience comparable joint outcomes to younger intensively treated patients with severe haemophilia. Long-term outcome data after joint bleeds in 48 patients with von Willebrand disease (VWD), 39 with moderate haemophilia and 59 with severe haemophilia, showed that, although those with VWD had repeated joint bleeding less often than the other groups (52% vs. 77% vs. 98%) joint health score was similar for VWD and moderate haemophilia [10]. Self-reported limitations in activities were comparable across VWD, moderate and severe haemophilia. Patients with Type 3 VWD had worst joint outcome, comparable to younger intensively treated patients with severe haemophilia, and limitations in activities occurred as often in VWD as in both moderate and severe haemophilia.

Goals of treatment

The primary goal of treatment for WBD is to stop bleeding rapidly in order to prevent progression to significant joint damage. This can be achieved with on-demand clotting factor (FVIII, FIX, aFVII, PCC, fresh frozen plasma) or von Willebrand factor (VWF). Prophylaxis is also an option, especially in those who have more than two or three joint bleeds per year, with clotting factors. Depending on the results of the ongoing HAVEN 6 study, the monoclonal antibody emicizumab may be a prophylactic option for people with mild to moderate haemophilia A in Europe [11,12]. A new trial is investigating its use as prophylaxis in VWD [13].

Routine ultrasound can be used to detect asymptomatic minimal bleeding so that ice can be applied and help prevent chronic synovitis. The technology is easy to use, available and cheap. MRI is less accessible and more expensive but should be carried out to detect bone lesions before orthopaedic surgery.

Treatment and prevention

Prevention of progressive joint damage is the primary focus of treatment. Chemical synovectomy using ultrasound-guided corticosteroid injections can be very effective in reducing synovitis, relieving pain and improving mobility. Treatment may need to be repeated every six to 12 months. There is little in the literature on the use of synovectomy in WBD specifically. However, the World Federation of Hemophilia (WFH) recommends that carriers of haemophilia with low factor levels are treated and managed in the same way as males with haemophilia [14]. In the case of synovitis, this includes synovectomy.

Alongside cartilage, bone and synovium, joints rely on muscle, tendon, ligament and nerve function. It is important to optimise these through regular exercise (Figure 2). People with bleeding disorders may be wary of exercise because of their increased risk of bleeds, but low impact activities such as walking, swimming, some gym work and gentle cycling can help strengthen muscles, stabilise joints, improve proprioception and avoid micro-bleeding. Exercise can also have a positive effect on mental health. Contact sports such as football, hockey or rugby should be avoided.

Regular physiotherapy is useful to develop, maintain and restore maximum movement and function and optimise quality of life. Recent guidelines recommend that physiotherapy should be available to everyone with a bleeding disorders, via self-referral [15]. As it can be difficult to find a physiotherapist who is experienced in treating people with bleeding disorders, the guidelines recommend that physiotherapists should work in partnership with other healthcare professionals to manage and provide services for people with bleeding disorders [15].

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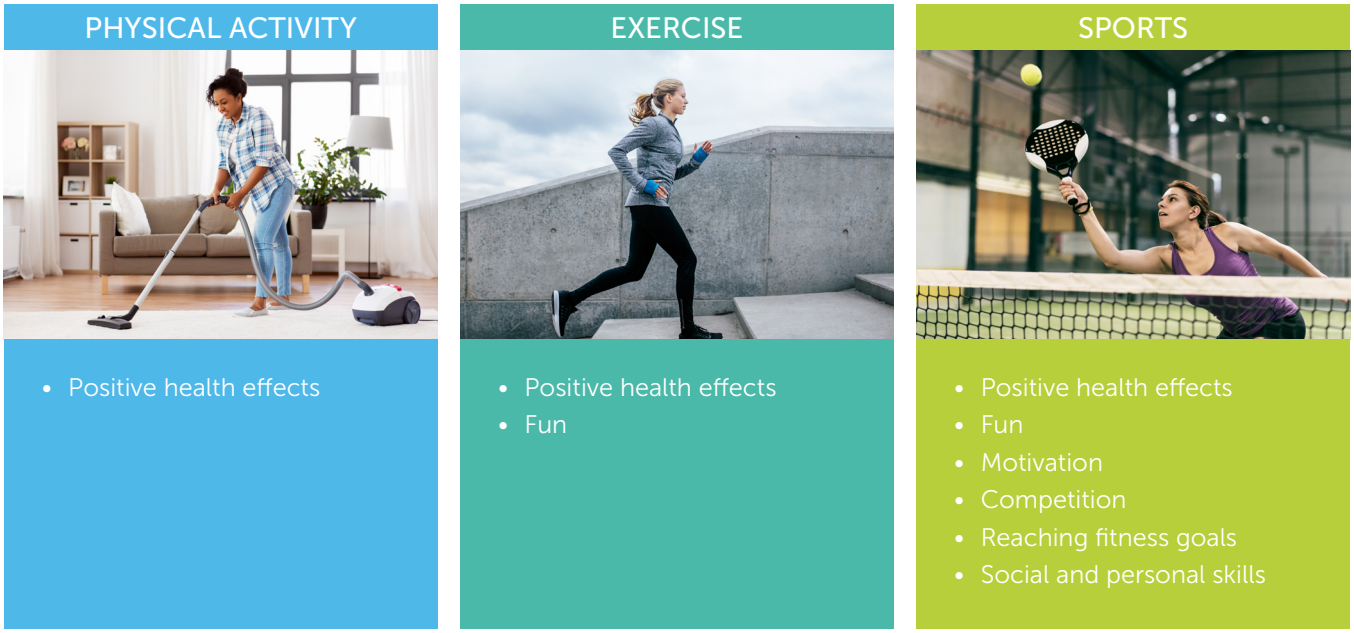
Figure 2. Physical activities for optimising muscle, tendon, ligament and nerve function, which can help to prevent progressive joint damage



Regular exercise and physiotherapy continue to be important even in patients whose joint damage has progressed to arthritis, as they will help to maintain mobility. Hyaluronic acid injections may be helpful in reducing pain and improving mobility, as will continued, regular injections of corticosteroids every six to 12 months. Stem cell injections for cartilage restoration are available at some clinics but data to support their use in people with bleeding disorders are limited. In severe arthritis, joint replacement surgery is an option when carried out with bleeding disorder protocols.

As osteoporosis is more common in women with bleeding disorders [16], it is also important to be aware of the need for bone strengthening measures such as vitamin D and calcium and weight bearing exercise.

TOP THREE TAKE-AWAYS

- People with bleeding disorders should not assume that gum bleeding is a part of their condition – it may be a sign of periodontal disease and should be treated
- WBD, including those with mild disease, may experience joint bleeding and progressive joint damage. Osteoporosis is also more common in WBD
- Regular exercise and physiotherapy are important in keeping joints healthy and can help maintain mobility even where joint damage has progressed

CONCLUSION

Neither gum nor joint bleeds should ever be considered normal or acceptable as both can be treated to avoid progressive disease. Dental and joint specialists and physiotherapists should work closely with treatment centre teams to ensure optimal care for long-term preservation of musculoskeletal health.

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