

Preparing for menarche: treatment and management of heavy periods in women with bleeding disorders

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Prolonged menstrual bleeding interferes with daily life and causes marked blood loss, resulting in anaemia and fatigue. Treatment centres should address the issue of heavy menstrual bleeding (HMB) with pre-pubertal girls in advance of their first period, in order to best prepare them. It is common for a bleeding disorder to be overlooked in primary care and in gynaecology clinics, and women sometimes struggle to get a correct diagnosis. There are cultural taboos that inhibit open discussion of menstruation, and women tend to minimise the severity of their symptoms. Health professionals should work to destigmatise the issue and seek an accurate account of bleeding severity, with diagnosis and treatment provided in a joint clinic combining gynaecology and haematology expertise. Treatment

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should be individualised, taking into account personal, social and medical factors, with the aim of improving quality of life. Great care is needed with regard to choice of language when talking about treatment, and treatment centres should consider offering open access to women who need support in dealing with adverse effects. National member organisations have an important role to play in educating people with bleeding disorders, health professionals and the wider public about the burden of HMB associated with bleeding disorders.

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The effect of heavy menstrual bleeding (HMB) on quality of life in women with a bleeding disorder is not fully recognised, despite being a prevalent issue (Table 1). Prolonged bleeding interferes with daily life and causes marked blood loss, resulting in anaemia and fatigue. Girls and women have to take time off school or work to cope with the impact, marking them out as different from their peers, which in turn risks causing isolation. There

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Table 1. Prevalence of menorrhagia in women with bleeding disorders^[1]

DISORDER	PREVALENCE (%)
Von Willebrand disease	32-100
Platelet dysfunction	
Glanzmann's thrombasthenia	51
Bernard-Soulier syndrome	13-98
Haemophilia	10-57
Factor XI deficiency	59
Factor XIII deficiency	35-64
Rare factor deficiencies	35-70

is concern that the bleeding disorders community has a double standard when it comes to HMB. If a man with a bleeding disorder were to report an annual bleed rate of 12, clinical alarm bells would ring; however, women tend not to receive the same degree of attention when experiencing heavy periods.

TALKING ABOUT HEAVY PERIODS

There are cultural taboos that inhibit open discussion of menstruation, and women tend to minimise the severity of their symptoms. Heavy bleeding may become normalised in some families; for example, when a mother has lived with heavy periods and is not aware that her experience is unusual. Even when women present for assessment of HMB, the possibility of an underlying bleeding disorder is commonly overlooked in primary care and in gynaecology clinics. As a result, women often struggle to get a correct diagnosis: the median age at diagnosis for women in the European Haemophilia Consortium survey was 16, but there was a broad interquartile range (3–29 years) for von Willebrand disease (VWD), and some women were not diagnosed until 40 years of age in other bleeding disorders^[2].

It is important that health professionals and women work together to destigmatise menstruation and talk more openly about periods. For doctors, consideration of a bleeding disorder and obtaining an accurate account of bleeding severity from the women seeking their care is important. A formal instrument would be useful to document symptom severity and response to treatment. The menstruation component of the VWD quality of life questionnaire may be helpful in this regard, but there is a need to develop a specific instrument^[3,4].

In pre-pubertal girls with a known bleeding disorder, the highest priority is to avoid a disastrous first period. Treatment centres should be prepared to tackle this issue in advance, and establishing early contact with

the family is essential. As a general recommendation, it is important to develop the topic gently from around the age of seven or eight years old, with the aim of preparing the individual for the fact that her periods are likely to be heavy and that she will need different care as a result. This approach applies to all types of bleeding disorder in women, and it may be useful to determine factor levels and activity in advance. The approach should be tailored to individual circumstances, but it is essential to involve the girl and parents while taking into account family composition and whether other family members have a bleeding disorder. Cases among girls and women in a family should be sought when a bleeding disorder is diagnosed in a family member.

In all women with a bleeding disorder, HMB may be related to other causes; changes in the pattern of bleeding should prompt consideration of an alternative diagnosis.

TREATMENT

Diagnosis and treatment of HMB in women with a bleeding disorder should be provided in a joint clinic combining gynaecology and haematology expertise. Haemostasis in the endometrium is not fully understood; bleeding severity does not correlate well with factor level and some women with levels within the normal range experience heavy bleeding. Treatment should be individualised, taking into account personal, social and medical factors, with the aim of improving quality of life.

From the perspective of controlling HMB, the choice of treatment depends on whether or not the woman



Improving diagnosis, care and treatment for women and girls who experience heavy menstrual bleeding is a multi-faceted challenge, involving the education of people with bleeding disorders and health professionals, early contact with the families of pre-pubertal girls, the breaking down of taboos around discussing menstruation, individualised treatment plans, and general awareness-raising

is planning a pregnancy. Where this is not the case, the choice of treatment is hormonal contraceptives. For younger females, parents may be alarmed that hormonal treatment entails the use of a contraceptive product, fearing that this means early sexualisation of a daughter. Great care is needed with regard to the choice of language when discussing the treatment of HMB.

Combined oral contraceptives (COCs) include a wide choice of preparations with different combinations of progestogens and oestrogens. They are effective in reducing menstrual loss, with regular menstrual periods, and prevent ovulation – and, thus, the risk of ovulation bleeding. COCs can also be used as an extended regime, enabling women to determine the number of periods they have per year. It may be necessary to try several products before an acceptable balance of effectiveness and tolerability is found. Breakthrough bleeding may occur during use of a COC, which can be managed through trying an alternative product or adjunctive treatment with tranexamic acid or desmopressin.

Another effective hormonal option is the levonorgestrel-releasing intrauterine device (IUD). The term ‘coil’ should also be avoided as many people use this term for the older copper IUDs, which may be associated with increased bleeding. The use of levonorgestrel-releasing IUDs is often linked with irregular bleeding and spotting up to six months after insertion, which is the main cause for discontinuation of this treatment. It may also cause amenorrhea. While it is not medically necessary to have periods, individuals may consider this to be unnatural, preferring to have some periods in the course of a year if they are predictable.

Some women cannot tolerate hormonal therapy due to the systemic effects of oestrogens and progestogens. In addition, hormonal therapy is not appropriate when pregnancy is planned. In these circumstances, haemostatic agents such as tranexamic acid and/or desmopressin are used to treat HMB. Factor replacement therapy may also be considered when appropriate, but is expensive, may not be available, and carries risks (e.g.

inhibitor formation, viral infection and risk of thrombosis). Tolerability may also be a problem with tranexamic acid, which is sometimes associated with nausea and diarrhoea. This may improve with dose adjustment; less frequent dosing should be considered on the grounds that taking some doses is better than none at all. Desmopressin is an antidiuretic hormone and can be associated with water retention and hyponatremia. This treatment should be limited to a maximum of two to three days, and fluid restriction advised.

In women who have completed their family and are not responding to medical treatments, surgical treatment with endometrial ablation is an option to control HMB.

Anaemia secondary to HMB should be treated with iron replacement. This is usually taken orally, but gastrointestinal tolerability may be a problem with the conventional twice daily dose regimen. If this occurs, the dose should be reduced to once daily. When rapid restoration of iron is required, intravenous administration is preferred. This is very effective but more expensive and should be administered in a clinical setting due to the risk of an anaphylactoid reaction.

The menopause can be a challenging time for women with a bleeding disorder, just as it can be for other women. Systemic symptoms are the same in both groups and equally likely to be overlooked by clinicians. There is no specific approach for women with a bleeding disorder. Endometrial ablation is an option to control heavy bleeding.

Treatment centres should consider offering open access to women who need support dealing with the adverse effects of their treatment. It would be useful to measure and record treatment outcomes; however, there is currently no guidance on how best to do this.

ADVOCACY

Some physicians, including some haematologists, appear to believe that, unlike haemophilia, VWD is not associated with severe bleeding. It is therefore

KEY MESSAGES

- The highest priority when raising the prospect of heavy menstrual bleeding with a girl with a bleeding disorder is to avoid a disastrous first period
- Health professionals should work to destigmatise menstruation and talking about periods
- The words chosen to describe heavy periods and their management should be chosen with great care
- Education and awareness raising among people with bleeding disorders, health professionals and the wider public are essential in improving access to care for women experiencing heavy menstrual bleeding

perhaps not surprising that women experiencing HMB can have difficulty accessing care. National member organisations of the World Federation of Hemophilia have an important role to play in educating and raising awareness among people with bleeding disorders, health professionals and the wider public about the burden of HMB associated with bleeding disorders. Peer support is invaluable in opening up conversations about menstruation, and can be facilitated through haemophilia camps and women's groups. NMOs can also assist in developing women ambassadors who can deliver educational initiatives, although champions outside of the bleeding disorders community are also needed to promote awareness more widely.

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