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

Michelle Lavin, Rezan Kadir, Sylvia von Mackensen, Roseline d'Oiron, Debra Pollard, Anna Tollwé

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 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 menorrhagia 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 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 heavy menstrual bleeding associated with bleeding disorders.

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The effect of heavy menstrual bleeding 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 is concern that the bleeding disorders community has a double standard when it comes to heavy menstrual bleeding. 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.

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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

TALKING ABOUT HEAVY PERIODS

The highest priority when raising the prospect of heavy menstrual bleeding with a pre-pubertal girl with a bleeding disorder is to avoid a disastrous first period. 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. It is essential to involve the parents and, at an appropriate time, siblings, because they will experience the knock-on effects. Cases among girls and women in a family should be sought when a bleeding disorder is diagnosed in a male. Factor levels should be measured in girls with heavy menstrual bleeding.

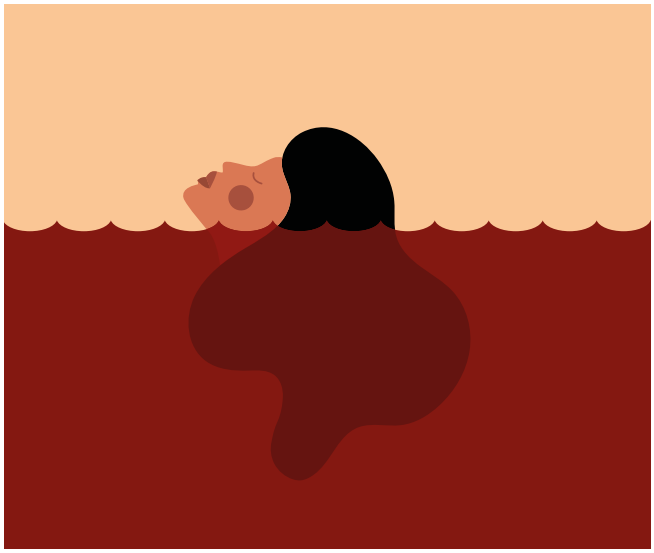
Treatment centres should be prepared to tackle this issue in advance, and establishing early contact with the family is essential. The approach should be tailored to individual circumstances, taking into account family composition and whether other family members have a bleeding disorder. Heavy menstrual bleeding may be related to causes other than a bleeding disorder; changes in the pattern of bleeding suggest the possibility of an alternative diagnosis. Conversely, it is common for a bleeding disorder to be overlooked in primary care and in menorrhagia clinics, with the result that 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. 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. This is a particular challenge when diagnosis has been delayed into adulthood –the median age for diagnosis for bleeding disorders in women in the European Haemophilia Consortium survey was 16, but the interquartile range was from age three to 29 for VWD, and up to 40 years old in other bleeding disorders, including haemophilia carrier status [2]. Peer support is invaluable in opening up conversations about menstruation, for example through participation in haemophilia camps and women's groups.

It is important that health professionals work to destigmatise menstruation and talking about periods, and seek to obtain an accurate account of bleeding severity from the women in their care. A formal instrument would be useful to document symptom severity and response to treatment. The menstruation component of the von Willebrand disease quality of life questionnaire may be helpful in this regard, but there is a need to develop a specific instrument [3,4].

TREATMENT

Diagnosis and treatment of heavy menstrual bleeding 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.



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

From the perspective of controlling heavy menstrual bleeding, the first-line choice of treatment is a combined oral contraceptive (COC). The advantages include a wide choice of preparations with different combinations of progestogens and oestrogens, convenience, effective regulation of periods that were previously unpredictable, and being able to determine the number of periods per year. It may be necessary to try several products before an acceptable balance of effectiveness and tolerability is found. Other hormonal options include use of a levonorgestrel-releasing intrauterine device. It is not medically necessary to have periods, but 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 a COC due to the systemic effects of oestrogens and progestogens. These effects may be so severe that they prefer to live with heavy bleeding. Factor replacement therapy may be considered as an alternative (except in women with rare disorders such as platelet function disorders), but it is more expensive than a COC and has not been a widely used approach historically. However, factor replacement therapy should be considered when bleeding lasts for >14 days in a woman who wants to become pregnant. Breakthrough bleeding may occur during use of a COC, which can be managed by trying an alternative product or adjunctive treatment with tranexamic acid or desmopressin. 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.

Anaemia secondary to heavy menstrual bleeding 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.

COMMUNICATION

Great care is needed over choice of language when talking about the treatment of heavy menstrual bleeding. Parents may be alarmed that hormonal treatment entails the use of a contraceptive product, fearing that this means early sexualisation of a daughter; they may also interpret the word 'coil' as meaning a copper intrauterine device, which in many people's minds is associated with increased bleeding.

Some physicians, including some haematologists, appear to believe that, unlike haemophilia, von Willebrand disease is not associated with severe bleeding. It is therefore perhaps not surprising that women experiencing heavy menstrual bleeding can have difficulty accessing care. National member organisations of the World Hemophilia Society 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 heavy menstrual bleeding associated with bleeding disorders. One approach is to develop women ambassadors who can deliver educational initiatives, although champions outside of the bleeding disorders community are also needed to promote awareness more widely.

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

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