Sexuality and Bleeding Disorders

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‘I am not living with my haemophilia’

‘If she doesn’t accept my haemophilia she doesn’t accept me’

‘I always told the girl that I have got haemophilia’

Sexuality and Bleeding Disorders

Series: Haemophilia care and treatment

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Content

Introduction

In 2014, it will be fifty years since Professor Simon van Creveld (1894-1971) established a clinic for patients with haemophilia. Van Creveld became a professor of paediatrics at the University of Amsterdam in 1938, but had been working on haemophilia from 1935 onwards. His hypothesis was that there is a factor in the blood promoting coagulation and that this factor is absent in people with haemophilia. In vain he tried to isolate this factor from blood plasma and sera, it was Edwin J. Cohn (1892-1953) in the U.S.A. who developed a method to obtain fractions from blood plasma containing coagulation factors. Nevertheless, in the Netherlands Van Creveld was the leading figure in haemophilia research whose work was widely recognised and honoured. He raised funds for a clinic to be built in Huizen (the Netherlands) where he could treat boys with haemophilia. On June 26th 1964 this clinic was opened by Her Majesty Queen Juliana.

Thanks to the scientific progress made in the sixties of the twentieth century boys with haemophilia had a life ahead of them. In the seventies the boys, who had spent long months in plaster casts in the Haemophilia Clinic, could get factor VIII (FVIII) infusions at home. There was less and less need for hospital beds. This meant an important change in the aims of the clinic that had been named after its founder, the Van Creveldkliniek (VCK). In the nineties the VCK moved to the University Medical Centre (UMC) in Utrecht, where today it is still accommodated as the Van Creveldkliniek, and since January 2014 as

Centre for Benign Haematology, Thrombosis and Haemostasis Van Creveldkliniek. The VCK has been designated as European Haemophilia Comprehensive Care Centre (EHCCC) by EUHANET.

The Van Creveldkliniek is a national centre for treating patients – both young and old – with coagulation disorders such as haemophilia and von Willebrand Disease. The VCK is an international training centre for the World Federation of Haemophilia (WFH) and participates in the WFH Twinning Programme.

To commemorate the fiftieth anniversary of the Van Creveldkliniek, this publication in the series *Haemophilia care and treatment* has been compiled. It addresses an issue that is often not discussed: sexuality. For patients and doctors, sexuality is not an easy topic to talk about and therefore it is frequently neglected. This is the case in general practices and in the interaction between patients and their specialist, so little is known about what might be a problem for a person with haemophilia, let alone what might be a solution to the problem.

Our question for this book was: ‘Is there something special about sexuality and people with haemophilia that needs attention and are there ways to improve guidance and intervention?’. Research on sexuality and bleeding disorders is lacking. Bleeds caused by sexual activity are rare, but psoas bleeds may occur in young adults. Such bleeds are treated as any other bleed in haemophiliacs but for complete rehabilitation and prevention of reoccurrence of a bleed, practical guidelines given by a physiotherapist are essential. How haemophilia patients (i.e. males) adapt during sexual intercourse has not yet been studied.

Probably more of a problem is the absence of lust and difficulties with erection or ejaculation, problems that increase in older age. Pain and invalidity in older patients may also have a negative influence on sexuality. Last but not least, there are the men with haemophilia who have been infected with hepatitis c (hcv) and/or hiv. They can have physical and emotional problems with sex, especially because of fear for virus transmission and side effects of the medication.

Finding a way to deal with a problem starts with a diagnosis and no diagnosis is possible when people don't talk about sex. This book aims to improve openness and discussion between people with haemophilia and the caregivers.

In this book personal experiences are described to illustrate the solutions patients found to enjoy sexuality despite limitations. In a series of interviews, people with a bleeding disorder – alone or together with their partner – tell how they cope with sexual relations and the limitations in their sexual behaviour caused by their physical condition, the risk of bleeding, arthropathy, hcv/hiv-infection and the side effects of their medication. In general it can be stated that the more partners are able to discuss difficulties, the better they will be able to adapt their sexual activities or temporarily avoid some of them. All persons were asked more or less the same questions. The interviews can be found throughout the book.

We are very pleased that we found the specialists in this field to write a chapter for this book. Firstly Doctor Woet L. Gianotten, who is a consultant in physical rehabilitation sexology and one of the few experts who has published on haemophilia and sexuality before. Secondly, there are colleagues from the vck team: Ruud Bos, Piet de Kleijn, Lily Heijnen and Eveline Mauser-Bunschoten.

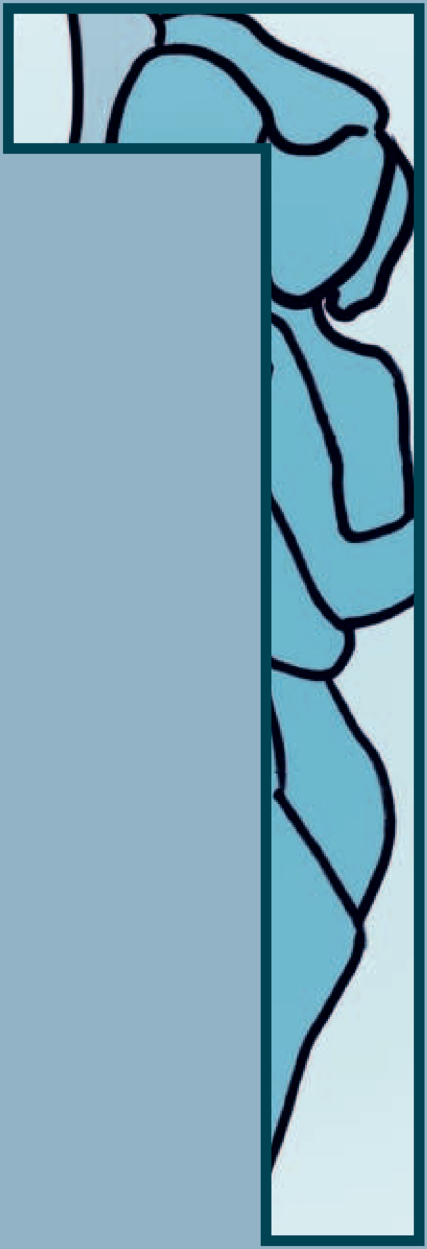
Bos has been employed as a medical social worker at the vck since 1999; De Kleijn has been involved in haemophilia care since 1980, he is responsible for physiotherapy care in the vck; Dr. Heijnen is specialist in physical medicine and rehabilitation, before her specialisation (1973-1979) she worked as a physician at the VCK and since 1983 as consultant for rehabilitation. Dr. Mauser-Bunschoten has been a member of the VCK team since 1979; she is widely involved in the organisations for and treatment of people with haemophilia.

Other colleagues from our hospital UMC Utrecht who participated in writing this book are Danielle van Elst-Laurijsen and Andy Hoepelman. As a Master Advanced Nursing Practice D.H.M. van Elst-Laurijsen, MA MANP, works at the Department of Infectiology of the UMC Utrecht. Prof. Dr. Andy I.M. Hoepelman is specialist for Internal Medicine and Infectiology. He is the medical manager of the Department for Internal Medicine, Infectiology and Geriatrics UMC Utrecht. His special area is HIV and viral hepatitis.

Dr. Annemarie de Knecht-van Eekelen has been teaching medical history at the Free University of Amsterdam. She has participated in the production of several publications of the Van Creveldkliniek on bleeding disorders.

The editors express their gratefulness to April Jones, Haemophilia Nurse Specialist at the Haemophilia Centre, Royal Victoria Infirmary, Newcastle upon Tyne (UK), who went through a draft of this text and greatly improved our English.

Utrecht, July 2014
The Editors



WOET GIANOTTEN

Aspects of sexuality

This chapter is a general introduction on some aspects of sexuality that are relevant for professionals dealing with bleeding disorders. It starts with the questions 'What is normal' and 'Why do people have sex'. The answers to these questions give an indication as to why it is important to pay attention to this subject, and elucidate the great diversity in people and their views on sexuality. In spite of all the current openness on sexuality, many people are not sure about 'What is normal?'.

WHAT IS NORMAL?

There is neither a statistic 'normal' for our patients, nor for us professionals. So, we have to deal with this variety. To simplify this statement: Some people are completely happy with sex once every two months, whereas others need sex every day. Some are happy and satisfied with short intercourse encounters, whereas others prefer long-lasting sensuality. Some have only conjoint sex, whereas others tend to do it on their own. And in between there are all kinds of combinations.

Another important aspect of sexuality that needs to be addressed is the difference between male and female, so we will deal with that difference.

After this overview of sexuality, we will discuss sexual function and dysfunction. Sexual function deals with the sexual 'machinery' and as such it provides a relevant start for better understanding of disturbed sexuality. Since chronic diseases predominantly interfere with the sexual function, the larger part of our considerations will be addressed to sexual function.

The last part of this chapter looks into the health benefits of sexual expression. Paying attention to sexuality and intimacy can be an aspect of good care, it can benefit the patient-professional relationship and it even can include elements of cure.

WHY DO PEOPLE HAVE SEX?

The usual answers are: for relation (love, commitment), for procreation (child wish) and for recreation (fun, lust, adventure). Even so, probably the most common reason is habit or custom (for instance ‘because we always do it on Sunday’). Maybe that doesn’t sound very romantic, but it is a very reliable argument, especially since it keeps sexual expression going, which appears to have health benefits. On the other side long standing habits can easily cause disappointment when they are disturbed. That regularly happens when sexuality becomes disturbed by a chronic disease.

Research in this area indicated more than 230 different motivations to have sex, including very physical reasons like muscle relaxation, sleep induction and pain relief (Meston & Buss, 2007). There are many less physical arguments like emotional relaxation, comforting each other, heightening the sense of male or female self, dealing with emotional disturbance and re-establishing contact after gradually drifting away or after a mutual quarrel. It may be tempting to value one argument more than another, but that disregards the human complexity.

MALE-FEMALE DIFFERENCES

Parents, peers, media, and culture together influence the way we behave as a girl or a boy and later as a woman or a man. However, behind those nurture elements, nature also has a strong influence. During the first months of intrauterine life androgen hormones are responsible both for the development of the male genitals, and for the typical wiring in the male brain. Although there are no gonadal hormones acting till the beginning of puberty, there are plenty of differences between boys as a group and girls as a group. Within each group there are also internal differences, but when comparing the two groups, one can say that boys / men are more function oriented and that girls / women are more people oriented. The one is not better or worse than the other, it simply is the way it is.

With the onset of puberty, the hormones start changing the body (from boy to man and from girl to woman) and they strongly influence behaviour. The androgens have a major role in sexual behaviour with testosterone (T) as the number one for sexual thoughts, for sexual desire and for arousability, but also for some less sexual aspects of behaviour like mood and assertiveness. In the adult male 95% of testosterone is produced by the testes and 5% by the adrenal glands, whereas in the adult female 50% of the testosterone comes from the ovaries and 50% from the adrenal glands. In men the T-level stays more or less the same until 40 years of age, after which it goes down with approximately 1% per year. In women the gonadal hormones fluctuate in a monthly cycle until the menopause, when the ovaries gradually stop producing testosterone. After some years of menopause the woman gets her testosterone only via the adrenal glands. As long as men and women are healthy enough, their T-levels will maintain sexual desire till they grow really old.

Males have T-levels between 10-15 times higher than females and that probably is an important part of the explanation for the differences in sexual behaviour. Men tend to be more focused on penetration, on genitals, on orgasm (and on sexual performance), whereas women tend to be more focused on relationship and on sensuality. The high T-level is also responsible for the male's higher assertiveness and lower emotional sensitivity. Another important difference is the rather even-tempered mood in most males versus the rather fluctuating mood in most women because of hormonal changes throughout their monthly cycle and pregnancies.

Although there is much variety among people and cultures, some other male-female differences deserve to be mentioned: For the majority of men visual stimulation is very effective. That's why many men want to make love with the lights on. Another strong turn on for most men is when the woman clearly shows her sexual arousal. For women, undiluted attention is a strong aphrodisiac and many need time to build up sensuality (*'good foreplay starts with breakfast!'*).

SEXUAL FUNCTION

In a simplified explanation of sexual function we tend to say that there are three phases: sexual desire, sexual arousal and orgasm. Originally these were described as successive in this sequence. Indeed, that is what happens in many men. If not geared down by the partner, many men tend to continue more or less in a linear process from desire to arousal (horny and erection) and then orgasm / ejaculation as the rounding off. For many women it is rather different, as is seen especially in longer standing heterosexual relationships. A common scenario is as follows: the man (with his higher T-level and accordingly more sexual desire) initiates erotic contact. The woman is not (yet) in the mood. When the man plays it well and pleases her enough, she will develop sexual excitement before getting sexual desire. The 'last' step of orgasm is also different for many women. Depending on mood and context of the moment she may have an orgasm, but many women can have full sexual satisfaction without having had an orgasm.

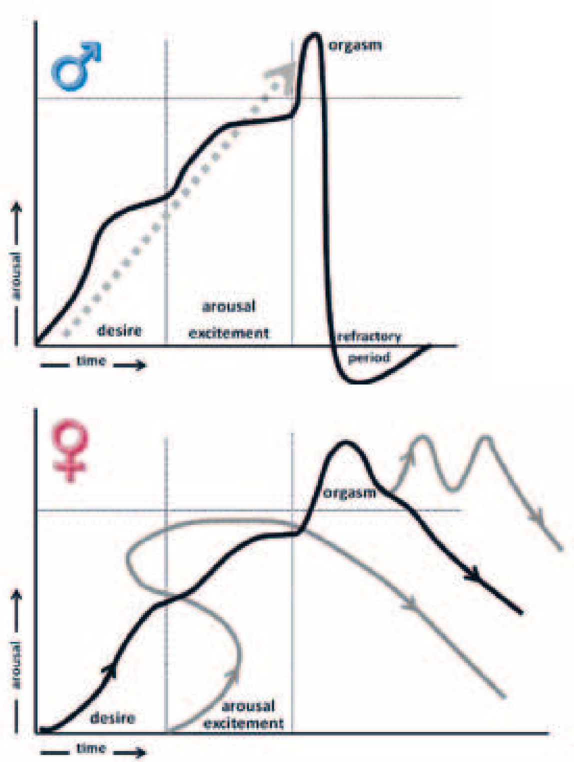


Figure 1. Sexual response cycle in male and female.

SEXUAL DESIRE

Although we tend to call it ‘butterflies in the belly’, sexual desire is situated in the head. Especially this part of sexuality is guided by hormones (particularly testosterone). It is also dependent on the neurotransmitter balance (with dopamine as desire-increasing factor and serotonin as desire-diminisher). Another relevant physical factor for desire is energy. Besides, one also needs sufficient stimuli to get into the sexual mood.

SEXUAL EXCITEMENT / SEXUAL AROUSAL

This phase is mainly guided by the circulation. The genital expression of arousal (erection and lubrication) is the result of hyper congestion of the penile and the perivaginal circulation. In the man, the smooth muscles in the cavernous body relax and the intracavernous space is filled with blood. Then the veins are compressed so that the pressure can increases to a ‘full penis’. Next, because the space is surrounded by the very tight *tunica albuginea*, the pressure increases more and the penis becomes hard and erect.

In the woman there is no *capsula*. Her hyper-congestion takes place in the vessels surrounding the vagina and consequently fluid permeates through the vaginal wall. This lubrication has two functions: One is for fertility (good lubrication favours the sperm cells). The other function is a mechanical one (‘oiling the vaginal cylinder’) to prevent damaging the vaginal mucosa and to prevent pain during intercourse. As a result proper circulation is a major condition for good arousal.

Two centres in the spinal cord regulate sexual arousal. Part of the erection and lubrication are the direct (reflex) result of genital stimulation. Testosterone has a minor role in this phase by creating arousability in the brain (‘becoming horny’).

ORGASM PHASE

Here again, there are large differences between males and females. For many males, orgasm and ejaculation are the same, and in ± 96% of men they coincide. The other 4% of men have learned to have an orgasm without ejaculation (and they can have that several times consecutively).

After having had an ejaculation, the man enters a refractory period (‘falling in a black hole’). His system has to be ‘reset’, which can take 15 minutes in a young man and up to a full day in an old man. Most women don't have such a refractory period. Besides, women can have an orgasm in more different ways. Roughly 90% of the women can get an orgasm via the clitoris, 25-35% via penetration (tapping the cervix and anterior vaginal wall), some only via fantasy or via breast stimulation. At least half of the women can have more than one orgasm in a row.

The major physical prerequisites for orgasm are intact nerves and proper neurotransmitter balances and of course proper stimulation.

SEXUAL DYSFUNCTIONS

We will successively discuss disturbances of the above-mentioned functions (desire, arousal, erectile functions and orgasm) and two additional disorders: sexual pain (or 'dyspareunia') and disturbed muscle function. We will focus on those aspects that are relevant for professional work with patients with bleeding disorders.

SEXUAL DESIRE PROBLEMS

When talking about desire, one has to realise the importance of context. During the larger part of the day people have no sexual desire. When one partner of a couple gets 'in the mood' and invites the other for action, the other partner is not always ready for that or may not yet be ready. That does not mean that there is a desire problem. We should differentiate between sexual aversion ('I really don't want to be involved') and absence of desire (or no desire yet). The last situation is rather common in normal daily life and very common in all kind of chronic disease related situations.

Absence of sexual desire is normal when testosterone levels are low. In haemophilia this can be the case in young / middle-aged HIV-infected men on combination antiretroviral therapy (cART). One in six men on cART was found to have premature decline of serum testosterone (Rochira et al., 2011). Lowered bio-available testosterone levels were also found in chronic HCV-infection and IFN-alpha therapy (Nguyen et al., 2006).

Other reasons for low desire are fatigue, a very common symptom in all patients with a chronic disease; and pain, common in many aged men with joint disturbances due to their haemophilia.

A relevant medical reason for not having pro-active sexual desire is the side effect of medication. Especially the SSRI (selective serotonin re-uptake inhibitor) and SNRI (serotonin noradrenaline re-uptake inhibitor) antidepressants are known to negatively influence sexual desire (and as a matter of fact also the other two phases of the sexual response). The group of paroxetin, citalopram and venlafaxine (responsible for >50% of outpatient antidepressant prescriptions in many western countries) is known to influence sexual desire negatively in 61-79% of patients (Serretti & Chiesa, 2009).

Besides physical joy-killers, there are also the more emotional reasons with depression and low mood in front. The middle-aged and older men with haemophilia have had a very tough life with many insecurities, considerable joint pain and social restrictions. That could be a reason to have less sexual desire. However, people don't always conform to expectations. Many of the aged men who survived the risks and hassles of haemophilia are real survivors and tend to be more creative than average people in finding benefits of life.

AROUSAL PROBLEMS

Problems with arousal are mainly a male issue. When women lack lubrication they tend to solve this with some saliva or lubricant or they have a little bit pain, but usually this is not a big problem. Many men, on the contrary, confuse erection with masculinity and male identity. For them no erection means not being a man. This causes an estimated 40% of men with erectile insecurity, who may completely stop making love to the partner. Their argument: 'I cannot finish the job properly!' (Meaning: I cannot give her sexual excitement or orgasm just by penetration).

ERECTILE PROBLEMS

Why do men with haemophilia have erectile problems? There are many possible reasons. We'll first look at the non-organic reasons, then the non-haemophilia related organic reasons and finally the potential problems caused by haemophilia.

Non-organic reasons

Many men grow up with a set of ideas on how sex should be. Some examples:

- Men are always ready and willing to have sex.
- The man is responsible for the woman's sexual pleasure. He should be the initiator and the actor in bed.
- Sex requires an erection.

These views provide neither an attractive background for sexual fun, nor are these very reassuring for a sexual encounter. Such ideas can cause a lot of nervousness in the process of dating or later in a relationship. Almost every man will once in a while experience moments of not having an erection during a sexual encounter because of low desire, fatigue, too much alcohol, or a weak condition due to a disease. In a man who believes in the traditional sexual myths, such an experience can cause fear of failure and become part of a vicious circle in which he is more occupied with fear than with sexual pleasure and sexual stimuli.

Organic reasons

Non-haemophilia related organic reasons are abundantly present in populations with a chronic disease. Especially all conditions that impair the circulation (like diabetes mellitus, atherosclerosis, hypertension and antihypertensive medication). Vascular emergencies tend to occur when the lumen of an end artery becomes too narrow. Since the penile end arteries have a small diameter (Ø 1-2 mm), atherosclerotic narrowing will cause problems, before problems arise in those end arteries with a bigger lumen, like the coronary (Ø 3-4 mm) and the internal carotid (Ø 5-7 mm). A decade ago erectile dysfunction was a luxury problem, but nowadays it has become an indicator to search for circulation disturbances.

Haemophilia related causes

Erectile dysfunction in aged males with haemophilia can be the result of hypertension which is not uncommon in this patient group. Subsequently, the prescribed antihypertensive drugs can deteriorate the erectile function. Besides viral infections, older men with haemophilia usually have disturbed joints with reduced flexibility causing pain. That can be a reason for diminished stimulation or diminished arousal (fearing pain), adding to fewer erections.

ORGASM PROBLEMS

Like many other men, males with haemophilia can have problems with ejaculatory control and may have premature ejaculation. With painful joints, some men can deliberately be fast, to prevent the development of pain.

No orgasm – the other problem in this phase – happens much more frequently. That too can be the result of (anticipating) pain and insufficient stimulation, which can also be caused by fatigue. However, most orgasm problems are the result of antidepressant-induced disturbance of the neurotransmitter balance.

DEALING WITH SEXUAL DISTURBANCES

THE BALANCE MODEL

The ‘balance model’ is a more or less holistic approach to ‘manipulate’ the various sexual functions. With the ‘balance model’ one can explain how a sexual function apparently has changed into a dysfunction, or how a dysfunction can be returned to proper functioning. It is not about ‘being in balance’, but about a pair of scales with on one side the inhibitors and on the other side the stimuli. Whereas many conditions can function as an inhibitor and deteriorate sexual function, many other conditions can be sexual stimuli that could improve sexual function. That is not only a fact of life, but, when understood well, one can proactively work on diminishing the inhibiting conditions and increasing the sexual stimulating conditions.

Especially in chronic disease there are many ‘somatic conditions’ that interfere with sexuality. When those conditions are irreversible, one maybe has to learn to accept that reality. Examples of somatic and non-somatic irreversible conditions are: anatomical changes, disfigurement, a very rigid coping style or an uncooperative partner. However, most conditions can be influenced by adapting time, medication, expectation, treatment etcetera. Examples are fatigue, pain, fear of failure, ‘poor sexual logistics’.

Next to diminishing the negative conditions, one can also add sexual stimuli. Nowadays, spicing up a sexual encounter with an X-rated movie, a little bit of alcohol, sexy underwear, or a vibrator is common practice in many relationships. So why not use these possibilities with our patients?

DISTURBED SEXUAL DESIRE

Diagnostic perspective

Some questions to ask patients are:

- Is sexual desire only absent or is there a real sexual aversion? (That last situation will ask for a very different approach).
- Does the absence of desire happen in most situations (general)? Or is desire only absent with the partner, but not towards masturba-

tion (or not with another partner)? In that case the physical origins become far less relevant.

- Could the low desire be a result of lowered androgen levels in the blood? The relevant laboratory tests (T, total T and Sex hormone-binding globulin (SHBG)) should be done early in the morning (between 07.00 and 11.00 hrs. AM).

Total T	> 12 nmol/l	no substitution required
Total T	between 8 and 12 nmol/l	a trial with substitution can be considered
Total T	< 8 nmol/l	substitution required!

Therapeutic strategies

1 Desire is lowered by fatigue factors. In that case several strategies can be used.

- Changing the timing. People tend to be less tired in the morning or after a nap. It seems strange that the professionals sometimes have to remind people about such a logical solution. But it really helps for the couples who get stuck in routine, and it can be a relief for the one who needs to change, but is too scared to mention that. Besides, in the morning, men and women have the highest testosterone levels!
- Sometimes the fatigue is 'local'. When stimulating the clitoris for a long time is too strenuous, a good vibrator could take over the action of weak or tired muscles.

2 Desire is lowered by too low androgen levels. Testosterone replacement should be considered (or definitively prescribed). An additional advantage of androgens is that the mood will improve and the fatigue will be less.

3 Desire is lowered by disturbed neurotransmitter balance.

There are many interventions for antidepressant-induced sexual dysfunction (Taylor et al., 2013). Some of the frequently used strategies are:

- Alteration of antidepressant. It seems useless to change from one SSRI to another SSRI.
- 'Drug holiday'. Whereas the antidepressant action builds up slowly (over weeks) and also disappears slowly, the sexual side effects develop within some hours and they also disappear quickly. That can be clinically used in antidepressants with a short half-life (for

instance in paroxetin, but not in fluoxetin). After 2-2 1/2 days not taking the medication, the sexual side effects have sufficiently disappeared to allow good sexual function, but after that, the next dose should be taken immediately. This strategy should of course not be used very frequently. However, it is important that the couple learns how to deal with this possibility, to keep it at hand when needed.

- Adding another agent ('antidote'). Many agents have been used: dopaminergic (f.i. amantadine); antiserotonergic (f.i. cyproheptadine); PDE5i's (sildenafil, vardenafil or tadalafil); and bupropion.

Whatever enhances sexual desire in the realm of context and conditions can be part of the strategy. Everything that in the past has been erotic could serve in the foreplay. Dancing, bathing together, sauna, watching a movie (romantic or X-rated), erotica, tools and toys, perfume, incense, etcetera. Professionals can give hints, but should be careful not to virtually enter the patient's bedroom. There is much difference between saying 'Use a vibrator!' and 'Some patients in the same condition used a vibrator. It helped some of them. Maybe you could consider trying that!'.

DISTURBED AROUSAL / ERECTION

Diagnostic perspective

In the past urology / andrology had several diagnostic tests available to investigate the erectile capacity, but these tests have all disappeared after the introduction of the PDE5-inhibitors.

The following additional information adds some insight and direction.

- An erection on waking up is nearly always a sign that 'the sexual machinery' functions well. Problems with getting an erection most probably derive from the emotional context. Usually, because the man fears to fail and as such distracts himself from the chances to get aroused.
- An erection disappearing during the sexual play is usually also a sign of emotional distraction. Such a situation tends to happen during removing his underwear, when entering the bedroom or when the woman indicates that it's time for penetration.
- An erection whilst watching 'X-rated' material is also an indication that the sexual machinery functions properly. One should be aware of how some men deal with porn when having erectile problems. It is as if they look via their penis. If no erection occurs, they conclude

‘that the movie is not good’, whereas the same movie could make him very horny before the erectile disturbance.

Therapeutic strategies

To enhance erection, both the man and his partner could pay attention to the conditions for sexual arousal. On the positive scale: a sexy atmosphere, sexy talk, sexy clothes and seductive behaviour of the partner (most men are very visual in their arousal). On the negative scale: fatigue, expectation that the sex should be perfect and that the female orgasm is often dependent on male erection. The items on the negative scale should be reduced. Usually less focus on erection and more focus on pleasure will enhance the sexual play. It will also improve the erection itself, as long as there still is erectile capacity left.

For the erection itself various strategies are mentioned in ascending order of being ‘medical’.

- Proper penile stimulation (by self or partner, by hand or mouth) will go directly via the reflex arc.
- A good lubricant. Lubrication usually will improve erection. One should choose the right one. Intravaginally, one can use a water based lubricant. However, that very quickly becomes sticky when used outside the vagina (for instance with masturbation) for which an oil-based or a silicone-based lubricant is recommended. For an oil-based lubricant one can look in the kitchen (pure olive oil or the like). Silicone-based lubricants are relatively expensive (and can afterwards make the floor of the shower dangerously slippery).
- A (strong) vibrator. Be aware: vibrator means vibration and not an artificial penis. Vibrators not only increase stimuli, but they can be used as a substitute for muscular activity as well.
- A constriction ring (or cockring) for when the erection gradually disappears. When an erection has developed (for instance after much oral or vibrating stimulation) the ring is tightened around the base of the penis, so that the blood stays inside the penis. This erection is a bit wobbly and also cold (because of no circulation). So, the ring shouldn’t stay on too long.
- When the erection cannot be provoked in other ways, it can be forced by a vacuum device. The device is (in an airtight way) put over the penis. After applying vacuum, the under-pressure will fill the penis with blood. Before removing the device (and the vacuum) a constricting ring is put round the base of the penis. For the rest,

see above. This vacuum method is not recommended for men with haemophilia, because it can cause extensive subcutaneous penile bleeding.

- Intra-urethral application of a small pellet with alprostadil. The prostaglandin permeates into the cavernous bodies and causes via vasodilatation an erection as long as it is combined with sexual stimulation. Applying the pellet can cause pain and in a small percentage also a minor urethral bleeding.
- Intracavernous application of alprostadil. The patient learns to inject himself. For men with haemophilia this is contraindicated because of the bleeding risk.
- Oral use of an 'erectile pill' with a phosphodiesterase-5 inhibitor (PDE5i). The three types are sildenafil, tadalafil, and vardenafil, with tadalafil the long-acting one (24-36 hrs.) and the others acting 4-8 hrs. The erectile action starts from ±1 hour after taking the pill, but erection happens only when there is sexual stimulation. PDE5i's don't cause an erection, but prevent the disappearance of blood from the cavernous bodies!

  A very important contraindication is the use of nitrates, because that combination can cause fatal hypotension.

  Nearly all side-effects of these drugs are the result of local hypercongestion: headache (in over 12%), flushing, nasal congestion, rhinitis, dyspepsia and dizziness. In haemophilia patients the nasal hypercongestion is a reason for caution, since there are some reports in the literature of serious epistaxis (nose bleeds) after a PDE5i. A small amount of people develop a rhinitis immediately after orgasm. An additional reason for such an epistaxis could be sildenafil induced platelet aggregation inhibition as suggested in in-vitro research (Berkels et al., 2001).

  Knowing these effects might create a dilemma in the care of men who crave for a proper erection. With only few reports of PDE5i induced epistaxis one could consider giving detailed information, prescribe a short acting erection pill (sildenafil or vardenafil) and start with a low dose.

DISTURBED ORGASM / EJACULATION

At age 55 most men have gone >4.000 times through the process of erection followed by ejaculation. When his erection disappears the man cannot imagine having an orgasm without an erection first. So, he doesn't try which implies that two people might be left out in the cold. In this situation, it can really be a gift when the professional provides information that the man can use in regaining the pleasure and benefits of an orgasm.

Diagnostic perspective

Relevant questions are:

- Is the orgasm disturbance due to context (time, place, relationship)?
- Is the orgasm disturbance induced by medication (especially antidepressants)?
- Is the orgasm disturbance a sign of avoiding pain?
- Has there been enough stimulation to reach an orgasm?

Therapeutic strategies

For orgasm one needs sufficient and strong stimulation. That is where the above-mentioned balance model can help a lot. Some of the stronger stimuli are:

- Visual stimulation. The required stimuli become stronger from desire via erection to orgasm. Whereas a romantic seductive face can suffice to enhance desire, and naked breasts will suffice to reach erection, for orgasm the naked crotch or watching people having sex can be needed. In most Western societies sufficient material is available on internet or in sex shops. But when apparently necessary, it is useful to indicate to patients that these materials exist and that in view of their disease it is sensible to use those possibilities.
- Although maybe less beautiful than the porn stars, their own partner can provide a lot of visual stimulation and add intimacy. Many couples experience the sexual power of the combination of the intimacy and excitement when watching their own partner masturbate.
- Imagination. Most people do have hidden sexual fantasies. Sharing those fantasies can be both very intimate and very exciting.
- Vibration. Except in cases of disturbed neurotransmitter balance or neurological damage, it is very difficult not to get an orgasm when properly using a strong vibrator.

Dealing with antidepressant-induced dysfunction has been discussed in the paragraph on desire strategies.

For dealing with orgasm problems caused by pain (and the fear for pain) the following should be noticed: People should learn how to use painkillers effectively, with a higher dose for special occasions and adapting the peak of maximum effect to the moment of making love. Make use of many pillows and use positions with the least pain. Maybe the couple should also accept that orgasm should not be obtained through intercourse. One can use the scenario of penetration without moving, talk lovingly to each other and afterwards give or receive an orgasm with a vibrator.

THE HEALTH BENEFITS OF SEXUAL EXPRESSION

Over several decades the medical profession was heavily confronted with the negative consequences of sexual contact. As a result of AIDS, infertility by chlamydia, and some cases of myocardial infarction or stroke during sex, the idea took root that sex is dangerous. It is rather new that the wide range of health benefits of having sex has been rediscovered (Whipple et al., 2007; Gianotten et al., 2006).

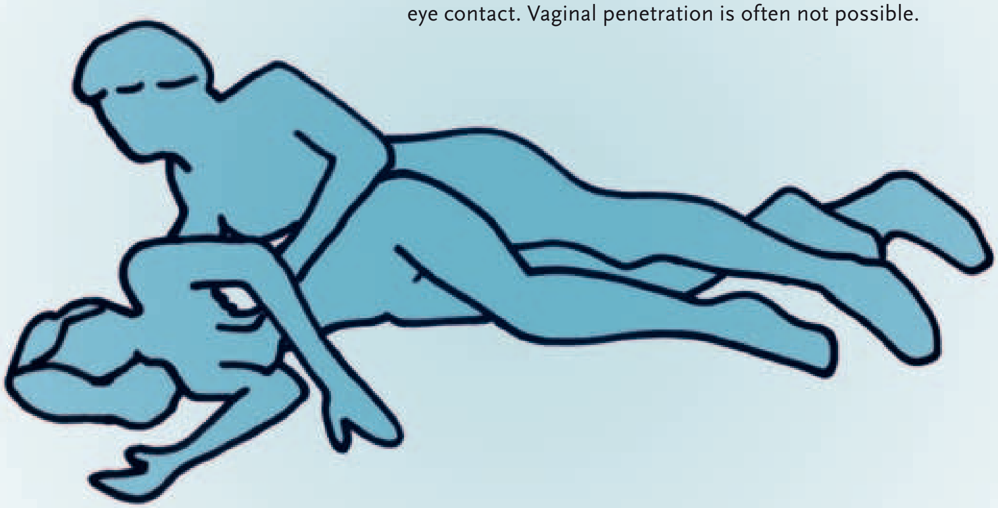


Figure 2. Side position with the man behind. Advantage: This position is relaxing for both partners. The woman may use a pillow between her knees. Disadvantage: There is no eye contact. Vaginal penetration is often not possible.

In this paragraph only the more impressive facts are addressed, starting with long-term benefits and then the direct (short-term) benefits. That difference is meaningful for the kind of relationship one has with the patient (or the couple). Long-term health benefits are relevant for disease risk reduction, but not so much when a patient is admitted or has an acute problem. However, when treating men with haemophilia the long-term health benefits could be relevant when the professional is also the one who is the general practitioner / carer and as such has to advise about lifestyle.

The real long-term effect worth mentioning is the effect on longevity. Men live longer when they have had more frequent intercourse, whereas women live longer when their sex life has been more satisfying, which for women does not mean having an orgasm (Palmore, 1982).

The strongest arguments for the correlation between sex and cardiovascular condition were found in a British longitudinal study among a group of men aged 45-59 years. After 10 years, the routine of having ≥2 orgasms per week statistically gave a 50% reduction in fatal coronary events when compared to the men who had ≤1 orgasm per month (Davey Smith et al., 1997).

More frequent ejaculation over the years also showed to reduce the prostate cancer risk. In that case, the causality is expected to run via more frequent secretion of oncogenic material with each ejaculation (Leitzmann et al., 2004).

The short-term benefits are relevant for other reasons. Sex can improve various aspects of body, mind and relationship. Part of these effects goes via an increase in testosterone and oxytocin. Whereas a higher testosterone level will facilitate sexual desire, sex can also increase the testosterone level. Testosterone decreases fatigue and it increases sexual arousal, mood and sexual desire.

The neurohormone oxytocin, well-known in giving birth and for lactation, is also called the social hormone. It has important functions in contact and sex. The oxytocin level goes up by massage, touching and being touched and then strongly increases with orgasm. Oxytocin has many different functions. It relaxes striated muscles (that is why some men fall asleep after orgasm), but it also increases mutual trust and openness (that's why some men start talking after orgasm). Oxytocin increases the perception of less explicit information, diminishing the autism resembling aspects of behaviour. Besides, oxytocin has anxiolytic and sedative properties. Some of the more striking short-term health benefits are:

- Pain reduction. Like other enjoyable things in life, good sex will distract from pain. It seems that part of the pain reduction is because of an increase in oxytocin level. For women there is an additional pay-off. Genital (especially vagino-cervical) stimulation and orgasm pro-actively elevate the pain threshold (Komisaruk & Whipple, 2005).
- Muscle relaxation. This was investigated in patients with spinal cord injury. Sexual stimulation (especially genital vibration) and orgasm decreased muscular tension for several hours (Biering-Sorensen et al., 2005).
- Sleep enhancing. Good sex (with or without orgasm) is for many people an introduction to healthy sleep. Probably caused by a mixture of being tired (in fact 'satisfied'), relaxed muscles, relaxed mind and maybe also neuroendocrine changes.
- Strengthening pair-bonding. This is a direct effect of the increase in oxytocin levels.
- Mood-enhancing and antidepressant effects. Oxytocin is also negatively correlated with anxiety and depressive symptoms (Scantamburlo et al., 2007).
- Alleviation of allergic symptoms and increasing immunity. Kissing decreases allergen-specific IgE production (Kimata, 2006). Young people with a moderate frequency of sexual intercourse had 30% higher IgA levels than the sexually abstinent youngsters (Charnetski & Brennan, 2004).

Beside these there are many emotional consequences of good sex (or good enough sex):

- Reducing the emotional distance between partners.
- Increasing the sense of being male / female (and boosting one's self esteem).
- Rewarding because of the accompanying pleasure and excitement.

In brief: having a sexual encounter together or having sex by oneself can in many ways promote good quality of life.

THE BENEFITS OF ADDRESSING SEXUALITY AND INTIMACY IN CONTACT WITH OUR PATIENTS

Patients and their partners don't feel safe to bring up the topic of sexuality and intimacy to their healthcare professionals. Even when there are real sexual disturbances for which they dearly want to get advice. According to the patient's opinion the professional should start asking. Few professionals on the other hand feel sufficiently safe to pro-actively bring up the topic.

These were also the findings after an investigation in our own rehabilitation centres (Gianotten et al., 2006). Based on that information, we developed for the various rehabilitation disciplines a training programme, with elements of knowledge and training skills. Part of the programme was that in the interval between training sessions, the professionals were obliged to address sexuality with their patients. The feedback and the evaluation afterwards gave two very relevant pieces of information:

1. None of the patients were shocked when the professional included the sexual topic, while the majority were very happy that those questions were raised.
2. Many professionals discovered that bringing up sexuality created in some way a better professional relationship (Post et al., 2008). Although this was not our aim, we presume that this will also improve adherence.

Besides all benefits in the 'Quality of Life' realm, sexual expression can carry direct positive elements in the field of care. Better sleep, relaxed muscles, more emotional relaxation, less stress regarding their partner and a better sense of self can enhance the conditions to recover from an acute physical problem or facilitate dealing with the hassles of physical suffering.

It is important to be mindful of this when dealing with patients admitted for longer periods of time. Allowing them once in a while to be undisturbed with their partner (or alone) can be a very robust piece of holistic care.

Maybe there are other benefits in the range of cure as well. Gradually some indications pop up in the scientific literature that sexual expression can have neuroprotective effects (Spence & Voskuhl, 2012). One area where the benefits of sexual expression become visible is in the prevention of cognitive decline (Hartmans et al., 2014).

EPILOGUE

An important effect of properly addressing the topic of sexuality and intimacy is the permission given. Like everybody else in Western cultures, patients, partners, and health professionals have been brought up with the idea that sex is for the young, the healthy and the beautiful. That is not a convenient position for good care. Many haemophilia patients and partners are suffering from the damage that the disease, the accompanying viral infections and the treatment have inflicted to their sexual and intimate life. We know that the incidence of sexual damage is high, especially in aged haemophilia patients. We also know that not all patients (and partners) need attention. The only way to discover their needs is very simple: *Just ask!* Ask every patient, the young, middle aged, old, married, single or gay. To a lesser extent the same goes for women with von Willebrand Disease.

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Annemarie de Knecht-van Eekelen and Lily Heijnen interviewed an older man with haemophilia. He is married and has a son and a daughter. The daughter is an obligatory carrier. He worked full-time and retired at the age of 65. Nowadays he likes to travel together with his wife.

‘I am not living with my haemophilia’

Did the fact that you are a person with haemophilia cause problems with starting a relationship?

“Not really. I myself never made a problem of having haemophilia. I tried to live a normal life. However, that was not that easy in those days when no clotting factor was available. When I was young I did have a lot of bleeds and my mother was very anxious that I should not hurt myself. If I played soccer I made sure she would not see me. Often I had a bleed afterwards and could not walk for weeks, but having played soccer was more important to me.

I was quite young when I got a relationship; in fact I have been married for over forty years now. Others warned the girl and her parents for the boy with haemophilia. They thought many problems would arise, bleeds, and of course there was the issue of heredity. But my girl and me, being young, and in love, we went our own way”.

When did you tell the girl that you have haemophilia?

“I have always been open about my haemophilia. When people ask ‘what is wrong with you?’, I say ‘it is haemophilia’ and I give a short explanation. That is all there is to it. Same with a girl. The parents of my future wife did try to talk her out of it, in vain, but it did take a couple of years before we could get married, we were engaged for quite some time”.

When you make love, are you afraid of having a bleed?
“No, never. I am not living with my haemophilia. When I look back at what I did when I was young, it was sometimes really reckless. I just don’t want to feel haemophilia interfering with my life”.

Does pain in your joints or muscles limit your sexual activities?
“You have to be creative. Pressure on my elbows is painful, same for my knees. So the classic position with the man lying on top of the woman is not an option. You have to find another position that is comfortable for both partners. My wife has nice ideas about how together we can be at ease and enjoy sex. If your relationship is ok, it is not a problem; your partner doesn’t want you to be in pain. Sex is something you have together”.

Did you consider not having children because of your haemophilia?
“We always wanted to have children. And haemophilia was not that much of a problem anymore when the factor products became available. When our son was born professor Van Creveld congratulated us and wrote ‘leave it at that’. But we did get a daughter too and of course she is carrier. In her perception having haemophilia is acceptable, she looks at me and sees how I deal with it. I am her reference. For my children I am not a patient, they say ‘you could do everything’, it is important how you deal with your own situation”.

Do you have hepatitis C and did the medication for hepatitis C treatment have an effect on your relationship?
“I have had a hepatitis C infection but I have been treated and I am no longer positive. The treatment was awful. For half a year I could not work. The day I was taking the injection I just laid on my bed, I felt so tired; I didn’t have the power to get up. I wasn’t depressive, just extremely exhausted. During that time I wasn’t capable of anything, let alone sex. My wife had to accept this, she did, but even though we knew that this was the side effect of the cure, it was very confronting. I had thought that I could cope, but you can’t tell beforehand what it does to you. The only thing is that you know why you do this to yourself. And fortunately it did help… no hepatitis C for me now”.

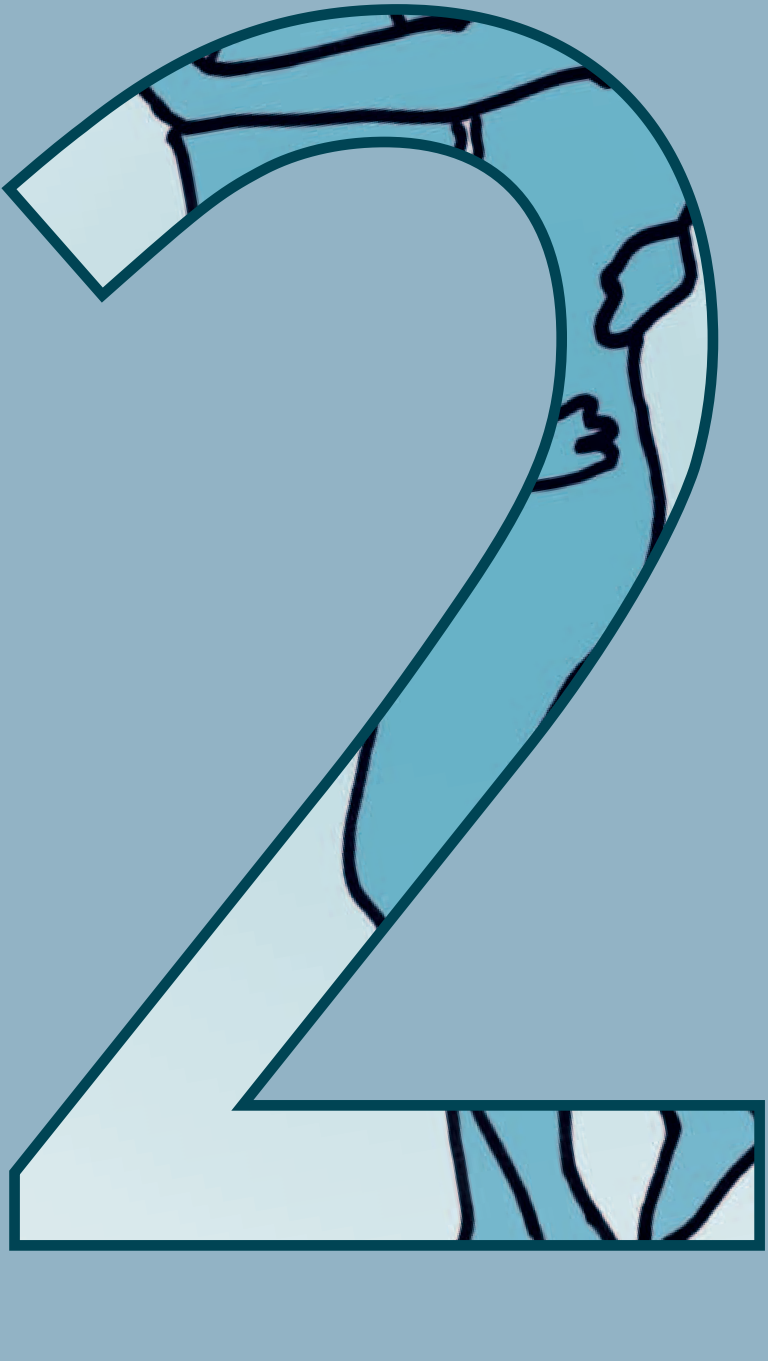
Are you HIV-positive?
“No. But in the eighties we did follow the information on HIV very closely. As soon as possible I took a test to find out if I was infected.

There were people who didn't want to take a test, which is so stupid. We had been using condoms from the start of our relationship on, and continued using them in that period, but fortunately I was free of HIV. After that my wife took the contraceptive pill".

You are getting older. Do you experience problems related to old age ailments?
"I have to take medication for high blood pressure and these pills have side effects. I have read the instructions for use before taking the medication and it does mention effects on erections. I thought it might be temporary, but I just can't have an erection. I am unhappy about it, and so is my wife. But on the other hand, we are no longer thirty; we have to live with it. It does influence your relationship. We did discuss it together, but not taking these pills is no option. Anyway, you can make love without intercourse".

Did you try a drug (sildenafil) for erectile dysfunction?
"In the past I did, but it is no fun; taking a pill a few hours before having sex lays so much emphasis on your performance, my wife needs to want to have sex at that certain time, it doesn't work that way. When we were young we used a condom, but that is different. For us it did hardly interfere with our love-making".

What is your advice to other people with haemophilia?
"Don't let your life be ruled by your haemophilia. In my youth my peers didn't take my haemophilia into account, they had no pity with me. I learned to accept and how to use my strength. I am not a macho, because with my haemophilia I will never fight: 'adapt and be resourceful, that is my advice'".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WOET GIANOTTEN

Young people and sexual development

In this chapter attention will be paid to several aspects of sexuality during the phases young people with a bleeding disorder pass when reaching puberty and adolescence.

INTRODUCTION

Like all others, youngsters with a bleeding disorder have to make transitions in the process of maturation: From a child under the wings of his parents to a person living alone with his own responsibilities. From visiting the paediatric professional together with his parents to visiting the adult care haematologist on his own. From schooling and financial dependency to a job and social independency. And from a kid playing with Lego and toys to a sexual person, eventually with a sexual partner.

DIFFERENCES BETWEEN BOYS AND GIRLS

Professionals dealing with bleeding disorders are very aware of male-female differences in disease patterns of their patients. There are many other areas where boys differ from girls. A considerable part of these differences develop in the first months of intrauterine life. In that period not only the genitals are shaped, but the male brain is bathed in testosterone hormone causing dimorphism in the wiring in the brain. That different wiring is for an important part responsible for the typical boys' behaviour. From shortly after birth the behaviour of children is no longer influenced by gonadal hormones until they enter puberty. Their behaviour is, on the other hand, also shaped by nurture elements (for example, education of parents, peers, society, school). Not all boys are the same, but for the majority this combination of nature and nurture means a life of 'rough and tumble'. Boys and girls differ in those early years not only in competition and play, but also in many other aspects like socialising, fine motor skills, verbal fluency, etcetera (Brizendine, 2006; Brizendine, 2010).

CHANGES DURING PUBERTY

During puberty and adolescence body and mind undergo many changes, becoming independent from parents and developing towards sexual partnership and reproduction. The adult secondary sexual characteristics and reproductive capacity develop and the growth spurt takes place. These physical changes are accompanied by alterations in sexual thoughts and behaviour in the relation to the opposite sex (at least for heterosexual kids) and in the relation to the surrounding society. All these processes are on the one hand orchestrated by the hypothalamic-pituitary-gonadal axis, but they are on the other hand also guided by genetic differences and influenced by environmental signals.

For (Caucasian) boys, the mean age of the onset of puberty is 11 years (between 9 to 13.5 years of age). For girls the range of pubertal onset lies between age 8 and 13 years, influenced by many factors including race, birth weight and maternal age.

SEXUAL INTERACTION CAREER

When discussing relevant stages of sexual development in this chapter, Dutch data is used (de Graaf et al., 2012). This is not because the behaviour of Dutch youngsters is so remarkable, but because there is ample reliable information on their behaviour. The Netherlands have a robust tradition in investigating the sexual behaviour of young people in representative national samples.

In the common sexual trajectory the first step for many boys is masturbation. When they subsequently get into erotic contact with other youngsters they tend to follow a gradual pattern of increasing more intimacy: from kissing to French kissing (tongue in mouth), to extensive petting to finally oral sex and sexual intercourse. This process from the beginning to intercourse is called the 'sexual interaction career'. For most youngsters this process takes a period of approximately four years.

Starting with boys and continuing with girls, each of these steps is addressed and the possible consequences for youngsters with a bleeding disorder are discussed. This information is relevant in the context of sexuality education, so that is the first area to be considered.

SEXUALITY EDUCATION

Some countries are far advanced in sexuality and relationship education of young people, whereas in other countries this specific element of education is rather absent. There are traditional religious leaders and politicians who believe that sexuality education induces young people to have an early sexual start. The reality is different. Proper sexuality education not only reduces sexual risks and disturbances, but it also slightly delays the sexual debut (WHO Regional Office, 2010). Proper sexuality education should include prevention of undesired pregnancy, prevention of sexual transmitted diseases (STD), and prevention of sexual abuse. It further should include the topics of diversity (prevention of homophobia) and pleasure. That pleasure element is a strong motivator to attract young people's attention.

A very relevant aspect of sexuality education is the near absence of '*Don't*!'. We prefer not to say '*No!*', but to focus on explaining the potential risks of specific behaviours and how to prevent those risks as much as possible.

BOYS

MASTURBATION

Approximately 70% of boys start masturbating in the window connecting one year before, to one year after the first nocturnal semen emission. In the Netherlands, 50% of the boys have masturbated at age 13 yrs. and 8 months. For the boy with a bleeding disorder there are three areas of worry or consequence.

1. HAEMOSPERMIA (BLOOD IN THE SPERM)

This happens sometimes, and in some boys it happens more frequently. It can create unrest, so it is relevant to have this phenomenon explained to the boy when entering puberty. However, there is no real danger in haemospermia and in general it does not require treatment with clotting factor concentrate.

2. SUBCUTANEOUS BLEEDING OF THE PENILE SKIN

Especially at the beginning of masturbation many boys still have to learn how to masturbate ‘smoothly’. When much force is used during this manipulation it can cause subcutaneous bleeding. The solution is to develop masturbation techniques with less force on the penis.

One way to prevent subcutaneous bleeding is the use of a good lubricant. For external use (as in male masturbation) one should not use a water-based lubricant since that quickly becomes dry and sticky. Good alternatives are oily or silicone-based lubricants.

Another way to enhance sexual excitement is by using a vibrator at the various sensitive areas of the penis.

In general clotting factor replacement for the subcutaneous bleeding is not indicated, only in case of a massive bleed or much discomfort, a single dose of clotting factor can be given aiming at a clotting factor level between 40-50%.

3. PSOAS BLEEDING

Bleeding in the ilio-psoas muscle is a painful and serious situation with dangers of involvement of the femoral nerve. The psoas is the muscle used for the typical male thrusting movement with rhythmic lumbar lordosis during sexual intercourse. Besides, in some men the contractions of the psoas muscle contribute to the development of sexual tension during masturbation. Psoas bleeding happens in 11% of young men with haemophilia (Gianotten & Heijnen, 2009).

Next to adequate clotting factor replacement to stop bleeding and apart from rehabilitation to prevent long-lasting functional limitation, attention has to be paid to sexuality with advice on a scenario with minimum muscular tension during masturbation. A way to reach the peak of ejaculation / orgasm more easily can for instance be the use of visual stimulation (especially moving pictures). The treatment of a psoas bleeding is discussed in Chapter 4.

KISSING AND FRENCH KISSING

At the age of 14 yrs. and 1 month 50% of Dutch boys have experienced French kissing. When youngsters have started with French kissing, they usually are also ready for hickeys (suction kiss) and love bites. It will be clear that this should be done with caution when a boy has haemophilia. So, information on this behaviour should be part of the sexuality education. Although in sexuality education it is preferable not to say 'don't', this is one of the few situations of a real 'don't or be careful' advice.

For the boy this situation could be a moment of his 'coming out' regarding his haemophilia. He should tell his date about his haemophilia. That could be a difficult decision: do you or don't you tell your date?

EXTENSIVE PETTING, MANUAL AND ORAL SEX

At the age of 14 yrs. and 8 months 50% of the boys have reached the level of extensive petting. At the age of 16 yrs. and 6 months 50% of

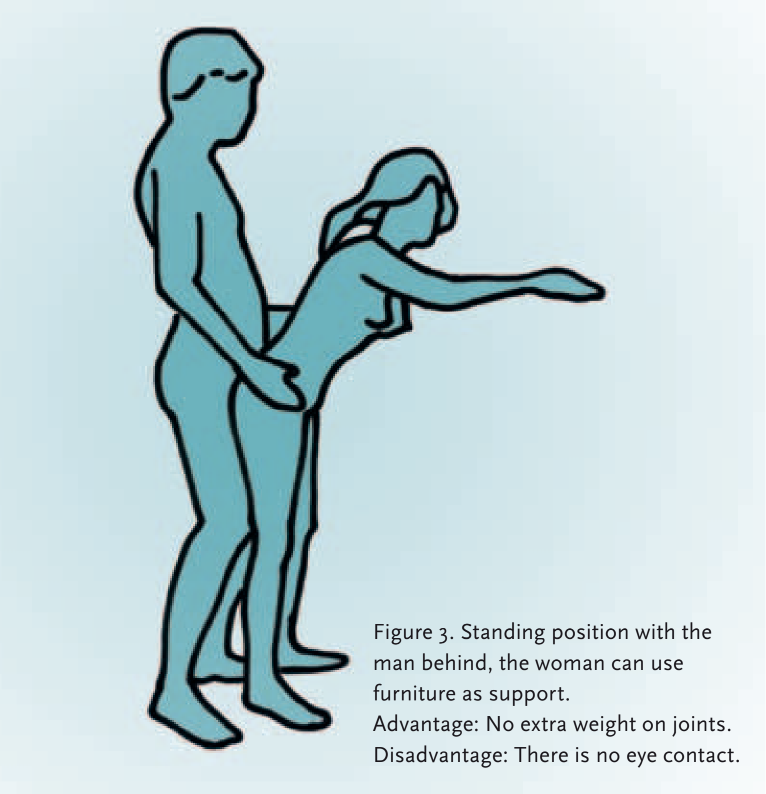

Figure 3. Standing position with the man behind, the woman can use furniture as support.
Advantage: No extra weight on joints.
Disadvantage: There is no eye contact.

the boys have had manual sex (with mutual masturbation). At the age of 17 yrs. and 1 month 50% of the boys have experienced (receiving) oral sex.

Especially in the beginning, those new experiences are so exciting, that one easily forgets to take proper care for muscles and joints. So it could be wise to discuss with the boy how he can be kind to his joints and muscles and how he could make use of a soft underground, of pillows, etcetera.

SEXUAL INTERCOURSE

At the age of 17 yrs. and 5 months 50% of Dutch boys have experience with intercourse. Intercourse asks for some additional preventive strategies. Depending on the sexual position different joints and muscles can be strained. So it is for instance important to make use of a soft underground.

During intercourse the ilio-psoas muscle is used for the coital movements. That should not be done too forcefully. Especially when the pumping movements of the erect penis are used to bring about the female orgasm, there is an increased risk of damage for the ilio-psoas or other muscles (and for the joints). In that situation, it is wise to consider fingers or a vibrator for that purpose. When intercourse is due, don't forget to think about additional preventive measures to avoid sexually transmitted diseases and undesired pregnancy.

HOMOSEXUAL CONTACT

Most of the recommendations given above apply in the same way for male-male sexual contact. With the same major advice: be gentle for joints and muscles. When anal intercourse is part of the sexual repertoire, additional attention is needed. The inserter in anal intercourse can (more easily than in vaginal intercourse) get a penile subcutaneous bleeding. The receiving man can more easily get a bleed from the anus, since the rectal mucosa is very sensitive for microtraumata. So in both situations much attention should be paid to copious artificial lubrication (oily or silicone), proper muscular relaxation and gentle penetration. An unexpected anal bleed should be treated properly without delay.

GIRLS

SEXUAL BEHAVIOUR

The age period at which girls start masturbating is extended over many more years than in boys. Having the first orgasm before puberty happens in 12% of the girls even at an earlier age than in boys. This is probably the result of the androgen increase from the *adrenarche*, the development of a new zone of the adrenal cortex (at 7-9 years of age).

Whereas in the Netherlands 50% of the boys had masturbated at age 13 years and 8 months, it took until 17 years and 3 months before 50% of girls had masturbated. That is later than other sexual activities.

At the age of 14 years and 9 months 50% of the girls have experienced French kissing.

At the age of 15 years and 5 months 50% of the girls have experience with extensive petting.

At the age of 16 years and 1 months 50% of the girls have experience with mutual masturbation.

At the age of 16 years and 9 months 50% of the girls have experience with sexual intercourse.

GIRLS WITH VON WILLEBRAND DISEASE

We are not aware that in girls with a bleeding disorder sexuality can be impaired via muscle or joint problems as happens in men. But when there is joint bleeding or arthropathy, as is not uncommon in type 3 von Willebrand Disease (VWD), this may have the same impact as in haemophilic men. For girls with VWD most sexual disturbances can be expected to be the result of long and irregular menstrual blood loss with two major consequences:

1. Anaemia, with fatigue (and decreased sexual desire).
2. Diminished days available for genital sexuality.

Cunnilingus will be strongly diminished, which in some relationships could mean that more fellatio will take place.

In addition there is the increased risk of ‘contact bleeding’ after sexual intercourse. Every woman should know not to start (the penetration part of) intercourse before there is sufficient arousal, shown in sufficient vaginal lubrication. Without proper lubrication penetration can more easily cause fissures. That advice is extra relevant for the woman with VWD since those (micro)traumata can cause much blood loss.

In case the couple intends to include anal penetration in their sexual play they should know that the anus is even more vulnerable to minor traumata than the vagina. More than average care should be given to copious use of artificial lubrication (with silicone lubricant), maximum pelvic floor relaxation and a very gentle approach.

For girls with VWD in puberty extra attention has to be given to proper cycle regulation (eventually in the form of hormonal contraception).

Combined with education on sexual transmitted diseases and pregnancy, sexuality could be an area for discussion although we have far less concrete advice as for boys in puberty.

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Ruud Bos visited an adolescent with severe haemophilia and spoke with him about his previous girlfriend and other relations.

‘If she doesn’t accept my haemophilia she doesn’t accept me’

I told you that we are writing a book about sexuality and bleeding disorders. You told me that you had a girl?
“Yes, when I was seventeen I got in a relationship with a girl, I knew her already for quite some time. We went to the same school since I was twelve. She was my first love, but unfortunately our relationship lasted only for two years. That is one year ago now. I am not going steady at the moment”.

Did your girlfriend know that you have haemophilia?
“Yes, she did. I told her myself. She knew it before we got in a relationship because we went to school together. And at school I once gave a presentation about haemophilia, so everyone knew about it”.

Was it a problem for her to have a relationship with you because of your haemophilia?
“No not at all. I give myself an injection with factor concentrate two or three times a week and because of this prophylaxis I never have a bleed. I can do everything I want, there are no restrictions. I never experienced a spontaneous joint or muscle bleed, so I can move and walk without limitations. People don’t notice that I have got haemophilia.

For my girlfriend my haemophilia was not an issue. She saw me injecting myself and that was it”.

Did you have a sexual relationship with your girlfriend?
"Yes, we had sex together. When we did it, it was the first time for both of us. The first time was exciting and thrilling, because it was new. We never discussed possible problems related to haemophilia".

What do you think might be risky for someone with haemophilia having sex?
"Well, well, gosh ..., I don't know, perhaps if a couple plays it rough, or when they fall out of bed?".

Some men with haemophilia have pain in joints or muscles in certain positions or they have blood in the sperm. Did you ever experience a thing like that during sex?
"Oh, well, I didn't even know that such things could happen. That is completely new for me. We never took special care. We had quite a lot of sex during these two years and we did it everywhere, outside, in the car ... Well, gosh, I have never been afraid that something could happen, I just didn't know. I am glad that I didn't know, because sex has to be nice and fun. If you start worrying it is less fun".

It is not my intention to frighten you, but you can imagine that older people with haemophilia have such problems.
"Sure, but that is because of their past, the bleeds they had when they were young. I never had a bleed, so for me that is no issue. I would never have thought about blood with my sperm, now you told me so, well, well, have you ever ..., I would hate it when I had to deal with this".

Do you ever have pain during sex?
"No, I never have pain in my muscles or joints. That is because I never had a bleed. As I said, my life is just like that of any other person".

Are you HIV- or HCV-positive?
"No, fortunately I have no infections. I have been tested and I am fine".

What would you do when you get another girlfriend? Will you tell her that you have haemophilia?
"I will tell her as soon as possible. Its better she knows. She has to

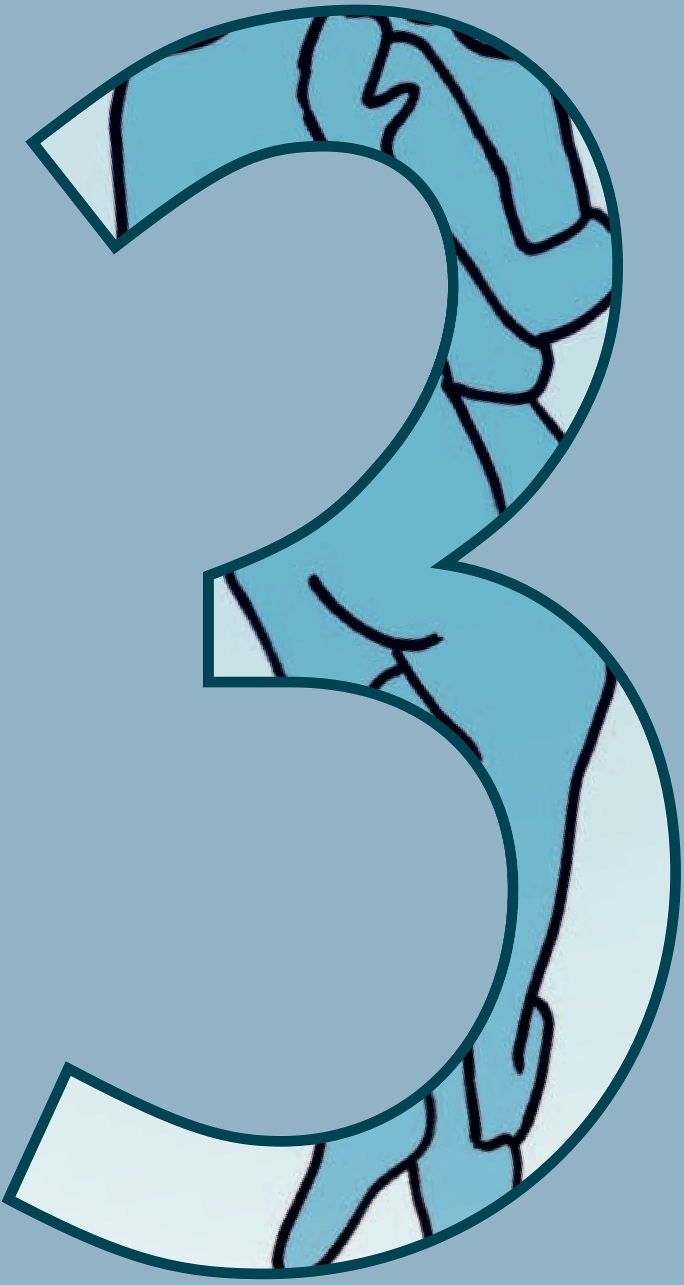
take me for what I am. And if she doesn't accept my haemophilia she doesn't accept me. Having a relationship for the second time will be much easier; I have experience now and that is more relaxed".

What is your mother's opinion about your relationships?

"My mother is always worried that something might happen to me. No wonder, as she has two sons with severe haemophilia. I sometimes feel controlled. I often go out at the weekend; I might stay with a girl and come home at six in the morning. My mother can't be awake till six, so I send her a text message about my whereabouts".

What do you expect from the future?

"I expect to get another girlfriend. I had one for a short time some months ago, but it didn't work. Not because of my haemophilia. I'm sure I will get another, I don't worry about that. My life is OK, I have nothing to complain about".


3

EVELINE MAUSER-BUNSCHOTEN
AND WOET GIANOTTEN

Sexuality in the ageing male

With ageing, haemophilia patients will not only suffer from haemophilia related diseases like arthropathy, HIV- or HCV-infection, which may have an impact on sexual life, but also from co-morbidity with various age-related chronic diseases.

INTRODUCTION

Older people enjoy sex too. Just as in younger people, a healthy sexual life has a positive impact on quality of life. In the process of ageing there is a gradual decrease in testosterone levels. Together with medical and psychosocial problems this can lead to a decline in sexual desire, to erectile disturbance and to delayed ejaculation, all factors that can diminish sexual activity.

EFFECTS ON SEXUALITY

The most frequent diseases affecting sexuality are diabetes mellitus, cardiovascular diseases, like coronary heart disease, myocardial infarction, and hypertension (Corona et al., 2010). Over the last decade, erectile dysfunction has been ‘upgraded’ from a luxury problem to a relevant and very robust indicator of cardiovascular pathology for the 30+ age group.

Furthermore the use of medication, for example combination antiretroviral therapy (cART) for HIV, medication against high blood pressure, SSRI (selective serotonin re-uptake inhibitor) and SNRI (serotonin noradrenaline re-uptake inhibitor) antidepressants may affect several sexual functions.

SEXUAL ACTIVITY

In general older people are not as sexually active as younger people. In the study of Helgason et al. (1996) among 50-80 year old Swedish men, the frequencies of desire, erections, orgasm and intercourse all gradually diminished by age. Good physical health, a (healthy) partner and a regular and stable sexual activity throughout life predict sexual activity when people get older, together with the ability to renegotiate intimacy in a way that the sexual focus changes from performance and intercourse to a more sensorial sexuality. As especially in older patients with severe haemophilia physical health is impaired by arthropathy this may have a great impact on psychosexual health. An unsatisfactory sex life may cause distress and result in depression and further deterioration of quality of life. It is therefore important to address sexual health adequately and where possible educate and inform patients about the why and how of healthy sexuality.

SEXUAL DESIRE

Sexual desire indicates the presence of desire for, and fantasy about sexual activity. A very relevant condition for sexual desire is sufficient levels of bio-available testosterone. From age 40 onwards there is an annual 1% reduction in testosterone levels. Chronic disease, a high body mass index (BMI) and a large waist circumference significantly decrease the testosterone levels (Wu et al., 2008). In an Australian study 60% of men over 70 years of age reported a decrease in sexual interest (Holden et al., 2005). This may be caused by impaired physical or psychosocial health or depression. Although sexual desire may be lower in the elderly, this will not always be perceived as problematic, especially not when both partners have an equal decrease in sexual desire. In such cases low sexual desire will often have no effect on quality of life. On the other hand when there is an imbalance between partners with regard to sexual desire this may cause stress, affect quality of life and lead to depression.

ERECTILE PROBLEMS

When becoming older it will take more time to develop an erection and more direct physical / genital stimulation is needed in addition to the visual, psychological or non-genital stimulation (Meston, 1997). Erectile disturbances can have many different causes, as there are: impaired physical or psychological health, problems within the relationship or death of a partner. Inappropriate erection may lead to impaired sexual intercourse, which can cause problems within a relation leading to stress and loss of self-confidence. However an erection is not absolutely necessary to remain sexually active, especially not in ageing men who may find alternative ways to live a satisfactory sexual life (Blanker et al., 2001). Older men are often not concerned about reduced erections. They can cope with this and do not experience distress. This may be one of the reasons why medical help often is not asked by this age group. However, we should realise that it may affect a relationship and quality of life while persons involved do not dare to seek help.

EJACULATION AND ORGASM

Impaired ejaculation may be premature or delayed. Premature ejaculation is mostly not perceived as problematic and is less often seen in elderly men compared to the younger age group (Corona et al., 2013). Delayed ejaculation may come together with a delay or absence of an orgasm despite sufficient sexual stimulation (McMahon et al., 2004). It increases with age and can cause personal distress (Corona et al., 2006).

The majority of absence of ejaculation / orgasm is the result of disturbed neurotransmission with SSRI and SNRI antidepressants as the major culprit. This group of drugs can influence all three stages of sexual response, with orgasm dysfunction as the most disturbing sign (Serretti & Chiesa, 2009).

Various treatment strategies can be used to deal with this antidepressant induced orgasm dysfunction, including anti-serotonergic or dopaminergic antidotes, change to non-SSRI antidepressants, and temporary (‘weekend’) pause when using SSRI with a short half-life.

HOW TO ADDRESS SEXUAL DYSFUNCTION

Therapy for sexual dysfunction in ageing men is initially the same as for the younger age group. It does not make much difference whether a man suffers from haemophilia. However, we should realise that decline in sexuality is a physiological part of ageing and often not pathologic. It is useful to explain the physiology of sexuality and at the same time provide the patient with information on a more sensory sexuality, less focusing on performance and coitus. For patients such experiences might lead to less distress, a more satisfactory sexual life, and better well-being. Improvements in general health and physical condition have a positive effect on sexuality too.

When there are signs of hypogonadism, additional testosterone could improve sexual life, although it is contraindicated in case of prostate carcinoma and haematocrit >52%. Advices with regard to sexuality and arthropathy are given in Chapter 6. Adequate painkillers and adaptation of positions may be of help to improve quality of sexuality.

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Safe sex

Remarks on safe sex of an older male with haemophilia who is HIV-positive

“In the eighties when the AIDS problem became prevalent I had no relationship, so safe sex wasn’t an issue. This changed in 1991 when I met my present wife. Right from the start of our relationship we discussed how to deal with the fact that I am HIV-positive. For me and my partner it always has been obvious that we should practice safe sex. You know how to handle a condom, at first it causes an unpleasant interruption, but you get used to it.

Things get different when your medication causes erection problems and your erection disappears when putting on the condom. The interruption is no longer unpleasant, it is annoying and the urge of unsafe sex increases. It is possible to use other ways of having sex when you have erection problems, but for us this wasn’t too good. I tried a drug for erectile dysfunction (sildenafil), but that was no success, I suffered from heart complaints. This is one of the side effects of sildenafil and I wonder if this side effect is stronger in combination with HIV-medication.

Eventually a condom is still the best assurance for safe sex. Don’t let the surprised look of the young sales woman bother you when buying a package regularly”.

Adapted from: C. Smit et al. (eds). Oud worden met HIV. Gezondheid en ziekte van oudere HIV-patiënten: een inventarisatie. [Ageing with HIV. Health and disease of older HIV-patients: an inventory]. Amsterdam: Aids Fonds, 2009.

4

PIET DE KLEIJN AND EVELINE MAUSER-BUNSCHOTEN

Acute bleeds and sexuality

Sexuality and bleeds affect each other in different ways. On the one hand sexuality may cause bleeds, on the other hand sexual activity may be impaired during the acute phase of a bleed due to physical and psychological distress.

INTRODUCTION

In rheumatic disease both males and females have problems with their joints during sexual activity, although they do not differ from controls regarding their sexual satisfaction (van Berlo et al., 2007). The classical intercourse position ('man on top', see Figure 4) has consequences for almost every joint of a man's body and will be used in this chapter as an example to show the effects of intercourse.

Most literature we found on this topic concerns people with rheumatic diseases and as far as we know nothing has been published specifically focussing on male sexuality in haemophilia. In order to discuss the consequences of sexual activities for each separate joint we will distinguish between bleeds in lower and upper extremities. Different from regular activities (e.g. walking) sexual activity relies on upper as well as on lower extremities. Different from rheumatic disease, in haemophilia both intra-articular and intra-muscular bleeds may counteract sexual activities. As a result of pain, sexual desire will be limited during the acute phase of a bleed in many patients. After a bleed has stopped, sexual activity may be impaired due to functional limitations or pain, but with some adaptations it may be restored satisfactorily for both partners.

SEXUAL ACTIVITY AND INTRA-ARTICULAR BLEEDS

UPPER EXTREMITY

Wrist

Sexual activities are comparable to the use of crutches during walking, as leaning on one's hand during the 'man op top' position forces the wrist in maximal dorsiflexion. This position is mostly limited after intra-articular bleeds, especially when these occur in the distal part of this composite joint. But even when the bleed is located in the more proximal part of the wrist, the dorsiflexion will always be limited and painful. Stabilising the wrist could be an option if a bleed has stopped, but certainly in the acute phase this is too painful, and therefore not possible.

Elbow

Especially older patients with haemophilia are used to crutches to decrease weight bearing of the lower extremities. During this activity they stabilise their elbow in a 10-20 degree flexed position. In this position the optimal moment of force is obtained in the triceps mus-

Figure 4. The classical intercourse position ('man on top').

cle, and resembles the position of the upper extremity during the 'man on top' position. Adaptations of the 'man on top' position after an acute elbow bleed are difficult; the best option might be to choose totally different positions or alternative ways of making love.

Shoulder

Stability of the shoulder is dependent on the rotator cuff (capsula, ligaments and muscles). The position of the shoulder during sexual activity is different compared to the position when crutches are used. During the 'man on top' position the upper arm cannot be supported by the trunk ('supplying upper arm'). As a consequence bleeds in the shoulder may affect sexual activity comparable to an elbow and wrist bleed.

LOWER EXTREMITY

Hip

It is important to distinguish between a hip and a psoas bleed. Differentiation can be assessed, after a thorough anamnesis, by inspecting active inner and outer rotation of the hip. In most acute hip bleeds rotation is not possible, but in case of a psoas bleed it is. However, when there is too much pain, testing will not be possible at all. In this case ultrasound is indicated to distinguish between a hip and a psoas

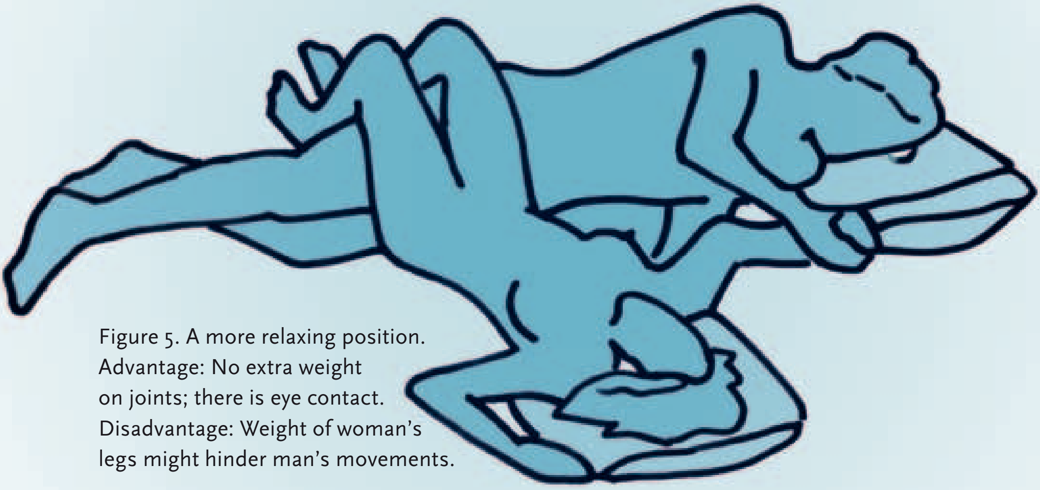

Figure 5. A more relaxing position.
Advantage: No extra weight on joints; there is eye contact.
Disadvantage: Weight of woman's legs might hinder man's movements.

muscle bleed. Unless the 'man on top' position can be adapted towards slightly more bended hips, which is the 'position of most comfort', the total lack of rotation will disable these activities to a large extent.

Knee

Like the elbow, the knee is the central joint of a relative 'long' extremity, and maximal power is needed to stabilise the leg during sexual activity. Adapting the 'man on top' position into a less weight baring position with a slight flexion of the knees seems a fair solution, at least from a functional point of view. Also the frequency of the rhythm can be adapted without losing quality of satisfaction.

Ankle

Dorsiflexion is the most limiting and painful position after an acute ankle bleed and even though the rest position is in slight plantar flexion, the maximum position, as during sexual activities might be asked, will also be painful. The alternative, stabilising ankle joints in an acute situation, means that maximum force is needed when the knees are more or less in extension. In a case of chronic ankle arthropathy supportive shoes may be a solution, in acute situations it is not. Adaptation of the knees towards a more bended position makes it easier to rest the ankles over the lower legs of the partner. As mentioned before: adaptation towards other positions can be more satisfying.

SEXUAL ACTIVITY AND INTRAMUSCULAR BLEEDS

The risk of sexual activity mostly described is the 'psoas bleed'. There have been presentations on psoas bleeds during World and Musculoskeletal Congresses and they are adequately described in a booklet written by Greg Blamey (2012). A bleed in the ilio-psoas muscle may result from various causal factors.

The ilio-psoas muscle

The ilio-psoas muscle (psoas or groin muscle) is a typical postural muscle consisting of the psoas major and the psoas minor muscle (de Kleijn, 1995). The muscle closely cooperates with the iliacus muscle to support the standing position, stabilising the lumbar spine and preventing the person from falling backwards. From a functional point of view, this is the adequate activity for this typical postural muscle. The bending of the hip joint is also assisted by this muscle.

Between 10-30% of intramuscular bleeds occur after trauma and/or stretching of muscles (Beeton et al., 2001). However, in early adulthood after a period of growth, the postural muscles have to adapt to this 'new' postural situation, and can be relatively 'short' for often a longer period of time. Between the age of 10 and 20 the risk of a psoas bleed is increased. For this effect the term 'surmenage' is often used, which means 'mental or physical overwork or overtraining'.

Sexual activity is a sudden, strong and very energetic activity, in an unusual position of the body with extended hips. Unlike regular sports activities, sexual activities are not trained for, often the opportunity suddenly arises. This implies that sexually active boys are more at risk for bleeds in their postural muscles. Bleeds may be underestimated as the first signs are neglected by sexual arousal. This risk may even be greater when the first signs of an ilio-psoas bleed are not recognised because a patient is not familiar with it, and therefore no clotting factor therapy is given in the early phase of a bleed. This further increases the risk of the development of a severe bleed.

Every health care worker has to be aware that in this specific age group 'pain in the groin' could be a bleed of the ilio-psoas muscle, due to surmenage caused by partying, playing soccer (de Kleijn, 1995) or by sexual activity. Generally the bleed can be detected by a thorough anamnesis. When a groin bleed is not recognised, as is sometimes the case in patients with mild haemophilia, they might develop flexion contractures of the hip (up to 80 degrees), requiring admission into the hospital (de Kleijn, 1995). Shortening of the psoas muscle, results in a bended (flexed) hip, but also in a lordosis of the low back spine (Blamey, 2012; Kendall et al., 1971). This is most clearly visualised by the typical 'psoas walk', characterised by a stance phase with bended hip and hyperlordosis.

REHABILITATION

Acute major joint and muscle bleeds are very painful and therefore have a negative effect on sexual desire and sexual excitement. Fear that the bleed worsens when having sex, results in even more pain and restricted joint function may also be a factor in refraining from

Figure 6. a) Patient walking with a flexed hip after a psoas bleed; b) Functional exercise: practice hip extension during stance phase to prevent contracture and improve gait (photographs: VCK Utrecht)

sexual activity. This is an important issue as we normally advise patients to maintain a period of relative rest in order to support recovery and to prevent further deterioration of the bleed. Analgesics may temper the pain caused by a bleed but have the disadvantage that when pain is not experienced any more, patients will become too active, with a possible negative effect. Depending on the site and severity of the bleed, sexual activity should be temporarily discouraged or performed in alternative ways in case of an acute bleed (Blamey, 2012).

The very first step of the rehabilitation process is to decide when a bleed has stopped. The decision is not based on the opinion of doctors and physiotherapists only, but merely on the patients' opinions. They know their body well and it forces them to learn from every bleed. In general, when pain vanishes, and movement of joints or muscles recover, patients become more active in daily life. But whether this also implies sexual activities is hardly known. This process has to be properly advised, because recurrence rates up to 41.5 % are described in literature (Domula et al., 1985). And every recurrence of a bleed will require a more extended rehabilitation process with an increased risk of complications, the major one being pressure on the femoral nerve (Nobel et al., 1980).

Once a bleed has stopped physiotherapy should be started. This is always aimed at functional recovery, at first avoiding weight bearing of an affected joint. For some reason the process of rehabilitation in haemophilia is hardly described in literature, but rehabilitation as advised after tendinitis resembles our clinical experiences with haemophilia This rehabilitation process is based on psoas inhibition exercises (Figure 6). Nothing has been published about how to translate these exercises towards sexuality and sexual activities. The use of the psoas position (Edelstein, 2009), in which the muscle is totally inhibited, is a reasonable advice for sexual activities in an alternative position. Such a 'functional milestone' (Guccione et al., 1996) is the first step aiming at restoration of activities of daily life, and is guided by disappearance of swelling and warmth, and recovery of muscle strength.

CASE HISTORY

A 20 year old patient is known with moderate haemophilia (FVIII 3%). During childhood he had repeated bleeds in his right ankle and elbow for which prophylaxis was given with three times a week 15 IU/kg FVIII. As he reported hardly any bleeds, prophylaxis was decreased to twice a week at age 15 and the patient stopped prophylaxis at age 18.

His bleeding frequency is low (0-2 per year) and bleeds are mostly caused by a trauma. He called our centre from his holiday address indicating pain in his right hip region. Despite treatment with 20 IU/kg FVIII for 3 days there was hardly any improvement. Assessment showed a limitation of both flexion and extension of the right hip; both movements were painful as well, whereas rotations were normal.

We diagnosed a bleed of the ilio-psoas muscle and asked him whether he had been sexually active during the past few days. He confirmed he was enjoying his holiday having fun with girls. We explained to him that the bleed might be caused by his abrupt sexual activity and advised to intensify treatment with factor FVIII to stop the bleed (twice a day 25 IU/kg BW), avoid weight bearing, and to refrain from sexual activity temporarily. After returning home he was seen in the haemophilia centre by a physiotherapist who advised to start rehabilitation and gave suggestions on sexual positions. He completely recovered within 4 weeks after starting the rehabilitation programme. No relapse of the bleed occurred. No limitations with regards to sexual behaviour were reported afterwards.

This case history illustrates that haemophilic men who develop a quite abrupt sexual activity, as is seen shortly after marriage or after falling in love and sometimes during holidays, are at risk of an intramuscular bleed of the ilio-psoas muscle. It is important to treat bleeds adequately with clotting factor in combination with (relative) rest. Once the bleed has stopped, physiotherapy aimed at functional recovery and participation should be installed. This implies that attention should be paid to sexual behaviour, which may be even more important because patients may, due to negative experiences, develop distress and fear for sexual activity.

Alternative sexuality or sexual positions may be of help to enjoy sex and to prevent recurrence of a bleed. In the case of elbow, knee or ankle bleeds, the missionary position is discouraged. Just like in the case of low back surgery, woman on top will be less stressful for the affected joints or muscles of the affected man as the partner does the 'heavy' work. Because not many patient records contain a step by step description of sexual activities as these are hardly mentioned during a consultancy, baseline programmes are difficult to provide. Common sense, however, is often enough to reach full functional recovery.

HAEMOSPERMIA

Although little has been published on haemospermia in patients with congenital bleeding disorders, the phenomenon is probably not very rare as has recently been shown by Sato et al. (2014). In this study 47 (32%) out of 148 patients experienced haemospermia, the average number of incidents was 4.6 per person. The age at the initial incident ranged from 15 years to over 50 years with a peak during the thirties. Patients perceive haemospermia as threatening. Some of them will contact their haemophilia specialist for advice, but many will be reluctant to do so and therefore the phenomenon might be underdiagnosed. Haemospermia may have a negative psychological impact on quality of life and sexuality. Because of fear for haemospermia a patient might refrain from sex. Girolami et al. (2009) describe three patients with haemospermia: one with haemophilia, one with von Willebrand Disease and a patient with a factor II deficiency. In all three cases the clinical course was benign. Substitution therapy with clotting factor concentrates was given in two of the patients. Additional measures may comprise rest and temporarily abstinence of sexual activity. In general haemospermia is an innocent phenomenon and does not require clotting factor replacement. In some cases a single dose factor VIII or IX is given aiming at levels between 40-50%. When repeated haemospermia occurs referral to a urologist for further examination is indicated to exclude a urological cause.

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Annemarie de Knecht-van Eekelen and Lily Heijnen interviewed a younger man (early thirties) with haemophilia. He is married, without children.

'I always told the girl that I have got haemophilia'

Did the fact that you are a person with haemophilia cause problems with getting a relationship?
"When I was a child I got some serious bleeds in my ankle. That influences my physical condition. Some of my joints are really bad. The bleeds had an important impact on my career, I couldn't choose the school I wanted and I missed so many lessons because of my bleeds. I like sports, but I can't participate in the way I want.

A relationship? I had no problems finding a girl and I always told the girl that I have got haemophilia. That is the best: just tell. They see you with crutches so they wonder, what is wrong with him? On holidays with a group of friends they see me injecting myself, so then they know. That is what I can add, that there are wonderful medicines allowing me to be almost the same as them.

I had a few girlfriends before I met my present wife, but I didn't feel the urge to jump into bed with each girl I met".

When did you tell the girl that you have haemophilia?
"As I said, I didn't make a problem of telling. My wife wanted to know about heredity, she asked a lot about that, and about the injections. Her family also knew it right away and had no problems with me; they accepted me the way I am".

When you make love, are you afraid of having a bleed?
"We are married now and we have sex regularly. There are some is-

sues. When we want to have sex, she asks if I injected clotting factor concentrate before. That is a reassurance that I won't get a bleed. So we preferably have sex when my clotting is high. But on the other hand that might make sex less spontaneous, having to think about your clotting. Anyway, clotting factor concentrate always lies nearby.

For me it is important that she doesn't touch my nose; when I was young I had a lot of nose bleeds, and I am more or less afraid of getting one".

Does pain in your joints or muscles limit your sexual activities?
"I never experienced a psoas bleed because of the coitus, and I do know what a psoas bleed is, I had problems with my psoas at high school.

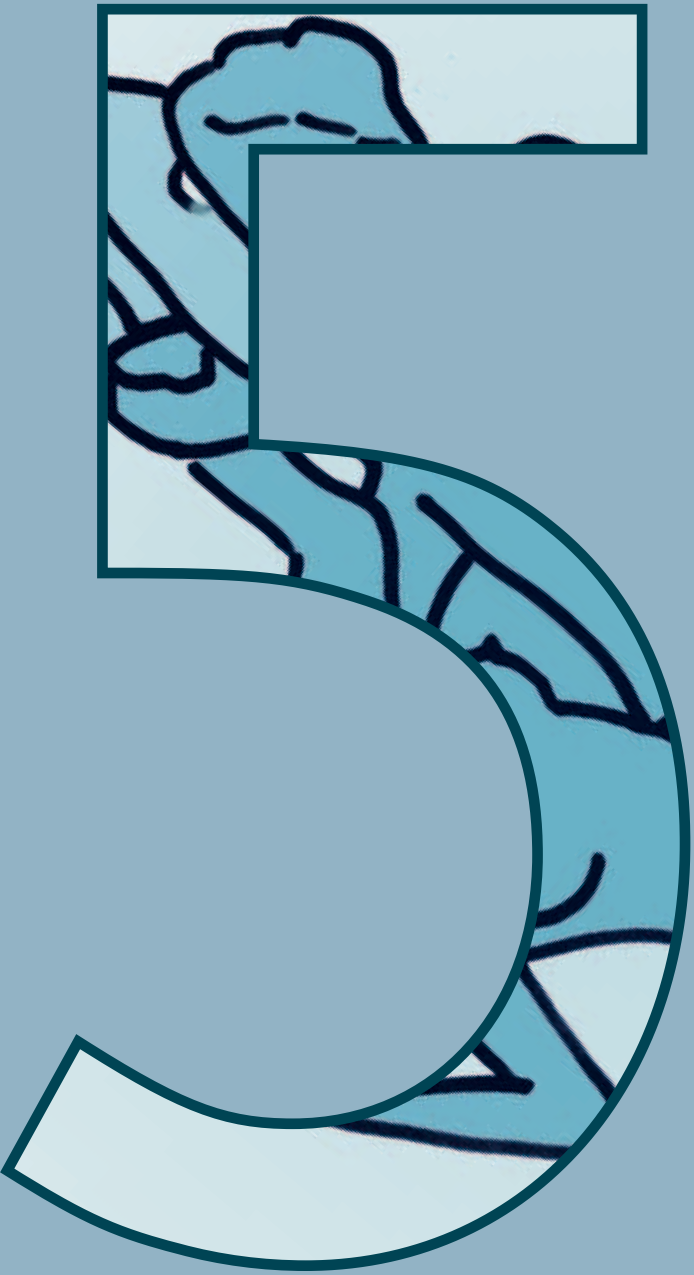
When she sits on top of me I don't like too much pressure on my legs. That is the same when we sit together on the couch and she puts her legs in my lap; when I feel pressure I am afraid of getting a bleed.

Also I don't like to rest on my elbows too much. As the plantar flexion of my ankles is limited, I put my feet out of bed most of the time, to relieve the pressure on my ankles".

Do you consider not having children because of your haemophilia?
"We would like to have children, we know all about heredity, but notwithstanding my haemophilia, we take the chance. We also discussed probable risks with our parents. My wife has stopped using the contraceptive pill so we will wait and see. When it is a boy he isn't affected with haemophilia. When it is a girl she is a carrier and she might have a low FVIII with bleeding problems. We haven't discussed prenatal screening yet. In one of the meetings of the Dutch Haemophilia Patient Society there was a presentation about having children. That was useful".

Do you have hepatitis C and/or are you HIV-positive?
"No. Fortunately I am from a younger generation for which HCV and HIV are not a problem. I don't use any medication".

What is your advice to other people with haemophilia?
"Work together. Know how to help each other. Provide information, show where information can be found. It is so important that people know that they are not the only ones dealing with a problem".



5

EVELINE MAUSER-BUNSCHOTEN

Sexuality in females with von Willebrand Disease

There are three types of hereditary von Willebrand Disease (VWD) with various subtypes. Patients show varying degrees of bleeding tendency. This chapter focuses on women with VWD who often experience heavy menstrual periods that might have a negative influence on their quality of life and sexuality.

INTRODUCTION

Von Willebrand Disease (VWD) is the most common inherited bleeding disorder affecting 1% of the population. It is a heterogeneous disorder and both men and women are affected. In type 1 VWD, which is most common, there is a quantitative deficiency of Von Willebrand factor (VWF). Type 2 VWD is the result of a functional VWF defect. The most severe form is type 3 VWD with a (near) complete lack of VWF synthesis. Clinical signs vary from person to person and in time. Bleeding tendency is not always related to the severity of the disease. In women, VWD is often diagnosed after the menarche because of menorrhagia. Blood loss during menstruation may be profound and prolonged, in some cases over two weeks, and is sometimes irregular. This may lead to physical problems like fatigue and dyspnoea due to anaemia for which substitution with iron may be indicated and in extreme situations blood transfusions are required. In the past women with VWD even died from blood loss during menstruation as has been described by the Finnish doctor Eric A. von Willebrand (1870-

1949) for the first time in 1926 (Nyman et al., 1981). Due to the severity of menstruation and its irregularity, sanitary protection has to be constantly available and this may have a psychological impact too.

QUALITY OF LIFE

Psychological and physical complaints caused by menorrhagia in females with VWD affect quality of life as was reported by De Wee et al. (2010, 2011). In a Dutch nationwide study, over 1000 patients with severe and moderate VWD were interviewed using a self-administered questionnaire. 423 were females >16 years, 81% of them suffered from menorrhagia, and 78% of the women >16 years received any kind of medical intervention for menorrhagia. In the same study, females with VWD compared to men, scored statistically significantly lower on general health, vitality, and physical health. Besides a physical and psychological impact there is a social effect of the disease, as due to the amount of blood loss women are not able to participate in sport, go to school or work.

PREGNANCY AND DELIVERY

Although during pregnancy the levels of FVIII and VWF increase, a substantial proportion of VWD patients will still have levels below normal at the time of delivery. This may lead to increased and prolonged and even extreme blood loss during and after delivery.

Shahbazi et al. (2012) assessed in a retrospective cohort study the occurrence of obstetric bleeding in the three trimesters of pregnancy, along with primary and secondary postpartum haemorrhage among 100 women with inherited bleeding disorders. In comparison with normal women, VWD patients showed a higher frequency of bleeding in the first trimester of pregnancy but the rate of miscarriage was lower. Vaginal bleeding in the postpartum period was often seen, not only in women with VWD but even more often in haemophilia carriers.

So these women require replacement therapy. Programmed delivery is recommended. Spinal analgesia is contraindicated because of the increased bleeding risk. Vaginal delivery is generally safe, however the danger of postpartum bleeding complications cannot completely be predicted, neither by past history of bleeding episodes or haematological laboratory tests of VWF activity levels. Clotting factor concentrate replacement should be initiated already in the first stage of labour. In patients with type I disease, desmopressin can be given during the postpartum period. Delayed postpartum bleeding may occur when replacement therapy is not continued for some days. In women with moderate and severe VWD laboratory monitoring measures should be continued for 8-10 days after delivery and – depending on laboratory results and bleeding – doses of clotting factor concentrate should be adjusted. Addition of tranexamic acid is indicated and must be continued post-partum until bleeding has stopped.

SEXUALITY

One may expect that profound and prolonged menstrual bleeding also has a great impact on sexuality. Although there are some publications on sexuality during menstruation and spotting there are no specific publications on sexuality and women with VWD. In their study Davis et al. (2002) report that sexual behaviour changes during vaginal bleeding or spotting. Kissing did not change, breast touching and non-genital touching slightly decreased. However, genital activity significantly decreased. Oral sex (cunnilingus) decreased to zero and vaginal intercourse declined from 67% to 7% during vaginal bleeding. Extrapolating these data to women with VWD in whom duration and severity of menstruation are more pronounced, the menorrhagia will have a major impact on sexual functioning and relationships.

CULTURAL RULES

Sexuality is influenced by religion and rules set by different cultures. According to the Bible, a woman with a vaginal bleeding is unclean. Sexual relations are forbidden within seven days from onset of menstruation (Ezekiel XVIII) and during pathological vaginal bleeding (Leviticus XV). Seven days after the cessation of vaginal bleeding, a woman is considered as clean and sexual contacts are permitted. In the Islam comparable guidelines are given. These rules will affect sexuality as the number of days when genital and non-genital sexuality is permitted is restricted.

TREATMENT

Women with VWD often need medical intervention for heavy vaginal bleeding. In our centre oral contraceptives are given to reduce and regulate menstruation and blood loss. In general this has a positive effect on the amount of blood loss, but sometimes antifibrinolytic drugs like tranexamic acid are needed in combination with clotting factor correction.

Hormone containing intrauterine devices have a positive effect to stop bleeding completely, although some women report an increase in blood loss.

HYSTERECTOMY OR ABLATION

In the past hysterectomy was performed in women who had completed their family. In the above mentioned Dutch study 20% of the interviewed women with moderate or severe VWD underwent a hysterectomy, mainly because of menorrhagia (de Wee et al., 2010). This percentage is twice as high as in women without VWD. Today, ablation is performed more often to stop bleeding. Hysterectomy and ablation will stop vaginal bleeding, which improves physical and psychosexual health. However, this intervention in itself may affect sexuality. One disadvantage of hysterectomy is that the menopausal hormone changes will start earlier (which could add to sexual disturbances).

McPherson et al. (2005) investigated sexual health five years after hysterectomy or ablation. Especially when hysterectomy was combined with removal of ovaries, sexual functioning and well-being was often impaired (mainly loss of desire, difficulties with sexual arousal and vaginal dryness). After ablation fewer psychosexual problems occurred. Compared to hysterectomy ablation is a less invasive procedure requiring a lower amount of clotting factor correction, and it has a lower risk of post-surgical bleeding. Therefore in VWD ablation is often preferred in case there is an indication for surgical intervention for heavy vaginal bleeding. However, it has the disadvantage that heavy bleeding may reoccur and hysterectomy has to be done anyway.

POST-COITAL BLEEDING

There is no literature on post-coital bleeding (PCB) in women with von Willebrand Disease. In women without von Willebrand Disease, post-coital bleeding is an indication for reference to a gynaecologist. Khattab et al. (2005) investigated a group of 116 women with post-coital bleeding without abnormal cervical cytology. 72 women were over 35 years of age and had PCB for more than 4 weeks; 94 women were younger than 35 years and they suffered from repeated unexplained PCB. At examination 23% of the women had an *ectropion*, 20.5% contact bleeding, 8% a polyp, 4% cervical ulceration, 3.7% suffered from cervical cancer, and in the others no pathology was found.

Infection with chlamydia is a risk for PCB. In women with von Willebrand Disease PCB may be more common due to their increased bleeding tendency, however, pathology should be excluded when repeated PCB occurs and patients should therefore be referred to a gynaecologist.

CASE HISTORY

A 25 year old woman with severe von Willebrand Disease (FVIII 0%, vW ristocetin cofactor <10%) is seen at the first aid department because of increasing abdominal pain, nausea and a swelling. Pain refers to the right shoulder.

The evening before admission she had been sexually active using a vibrator deeply intravaginally.

At admission her blood pressure was 90/40 mm Hg, pulse 100/minute. At physical examination an abdominal distension with peritoneal irritation was seen.

Ultrasound revealed free abdominal fluid with blood clots lateral from the uterus. There was no vaginal bleeding. A CT scan with contrast showed an active bleed median from the right ovary, probably originating from the ovarian artery. Haemoglobin level was 6.5 mmol/l and dropped to 5.4 mmol/l. Conclusion was a traumatic bleeding caused by deep penetration of a vibrator.

She was treated with 50 IU/kg (total 3000 IU) Haemate P and blood transfusion, followed by daily 2000 IU Haemate P for five days. Recovery was uncomplicated and complete. She has no limitations with regard to sexual functioning.

This case shows that in rare cases vaginal sexuality may lead to intravaginal bleeding. When treated adequately with clotting factor concentrate bleeding will stop and recovery will be uncomplicated.

PREVENTION OF BLEEDS

Depending on the amount of lubrication, the health of the vaginal mucosa, the size of the penetrating penis (or vibrator) and the energetic force of the movements, there will be more or less microtraumata in the vaginal wall. The woman with VWD (and her partner) should be aware of the importance of proper lubrication, and eventually make copious use of a good lubricant. Since the rectal mucosa is even more sensitive for microtraumata, it is absolutely necessary to pay attention to proper artificial lubrication when anal penetration is part of her sexual repertoire.

Soft cups or 'instead cups' are an alternative to pads and tampons. These cups hold rather than absorb menstrual fluid, they can be worn up to 12 hours. In case of menstruation or post-coital bleeding soft cups can be worn during intercourse to prevent disturbing blood loss.

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Lily Heijnen interviewed a woman with von Willebrand Disease. She is in her fifties, married for almost forty years, without children.

'I have never been afraid of a bleed'

Did the fact that you are a person with von Willebrand Disease cause problems with starting a relationship?

"No, I was sixteen years old when I got to know my present husband. I did have problems with my monthly periods but that is not a topic to discuss with your boyfriend. When I was seventeen I had a major bleed in my ovaries, this caused quite a panic. Then my boyfriend heard about my condition. It was a severe bleed and I was taken to the haemophilia clinic where first professor Van Creveld treated me. He told my grandma, who was with me, to ask if I was pregnant. But I wasn't, we didn't have sex at that age. My condition was really bad; I had to have surgery in one of the regional hospitals.

As a child I suffered from nose bleeds, there was no treatment at that time. They gave me full blood, and later plasma or cryo. At the age of twelve I was knocked down by a car, which resulted in a knee bleed, after that I often had a knee bleed twice a year".

When did you tell your future husband that you have von Willebrand Disease?

"I can't remember exactly, but when I had this major bleed in the ovaries he knew already. Later we discussed having children, we deliberately choose not to have any. Especially my husband was afraid that children would have problems with the disease".

Did you use preventive measures?
"As I told you, I was twelve when I was admitted to the haemophilia clinic for my knee bleed and I had to stay in the clinic for a year for rehabilitation. At that time I got my first menstruation, which was extremely severe and went on for three weeks. They gave me tranexamic acid (this is used to control unwanted bleeding; it reduces the amount of blood loss) and a pill. That pill I had to take for three weeks and then stop for one week. During that week I still had strong menstrual bleeding. When I was eighteen I got to know that this pill was THE pill, the contraceptive pill. I used it from age twelve to fifty, till my menopause".

When you make love, are you afraid of having a bleed?
"I have never been afraid of a bleed. But the severe bleeding during my period has its influence on our sex life. During my period, which continues for 7-10 days, we don't have sex. I lose too much blood, the blood flows in waves with clots and constantly I am busy with sanitary pads".

Is there blood loss during coitus?
"Yes, postmenopausal, not before. There is a little blood loss, but that is over after a visit to the toilet. I think it is caused by the dry, thin vagina wall, even though we do use lubricant and condoms".

Did you know that hepatitis C and HIV could be transmitted by blood products and did that influence your sex life?
"Yes, when we heard that HIV can be transmitted by blood we started to use condoms and we still do, that was our own initiative. Our physician thought it wasn't necessary because we had no multiple other sexual partners".

Do you still use condoms even though blood products are safe now?
"Yes, we continued using condoms and it doesn't bother us. We also find a condom more hygienic for my husband, because sometimes I lose some blood from the vagina during coitus".

Did the knee bleeds limit your sexual activities?
"No, in the past my knee never obstructed sex and I wasn't afraid of getting a bleed. But seven years ago I fell and broke my upper leg. They put a pin in and they also gave me an artificial knee, because my

knee was ‘worn out’. Unfortunately this went wrong, I have a stiff knee since, but it doesn’t hurt. With a stiff knee the positions during sex are limited and I don’t like to lie on my back or sideways with the stiff knee. Lying on my belly during coitus feels best”.

Does the position you take during sex influence your orgasm?
“Yes, getting an orgasm is less easy, but during the prelude there are many possibilities. And, getting older, my sexual needs diminish. We have sex about once a week, which is enough for me.”

What is your husband’s opinion, would he prefer to have sex more often?
“Yes, I suppose so, but it is once a week”.

Do you notice limitations or effects of ageing?
“I take medication for high blood pressure, but I don’t think that affects my sex life. I often have cystitis, it is a family disorder, and when I have cystitis we don’t have sex”.

Are you happy with your life?
“Yes, together with my husband, who is retired now, we enjoy ourselves. I am an optimistic type. Compared to men with haemophilia my limitations are minor. Yes, the stiff knee is a pity, I had to stop cycling, but we often take trips in the weekend, we have nice outings. Yes, I am happy with my life”.


6

LILY HEIJNEN, PIET DE KLEIJN AND
EVELINE MAUSER-BUNSCHOTEN

Sexual activities and haemophilic arthropathy

Haemophilic arthropathy is characterised by a limited range of motion, pain and stiffness. In this chapter possible treatment and advice to improve sexuality are given to men with haemophilic arthropathy.

INTRODUCTION

In general in developed countries, patients with severe haemophilia born well before replacement therapy was available, suffer from haemophilic arthropathy of five synovial joints (Ahlberg, 1956). Today, haemophilic arthropathy is still a major complication for people with haemophilia in developing countries as well as for patients worldwide with an untreated inhibitor.

In a recent Dutch study among patients with severe haemophilia between 18 and 70 years of age (mean age 40.5 years) two-thirds of the patients indicated having one or more painful joints, 52% had two or three painful joints, and 14% reported five painful joints. Arthropathic pain is mostly seen in ankles, knees and elbows; haemophilic arthropathy in wrists, hips and shoulders are no exception (van Genderen et al., 2006a).

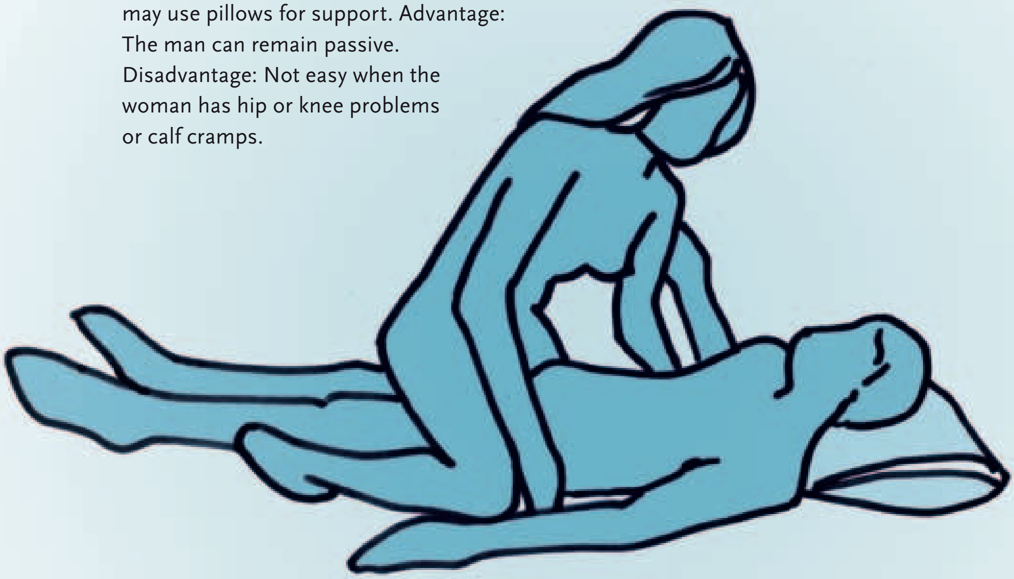
Haemophilic arthropathy is characterised by a limited range of motion, pain and stiffness. Depending on pain intensity, in combination with limited muscle strength and bad condition, these patients experience functional limitations in their regular activities of daily life (ADL), of which sexual functioning is a part (van Genderen et al., 2006b).

LIMITATIONS OF SEXUAL ACTIVITIES

Unfortunately, in haemophilia care, no large survey exists on the specific topic of haemophilic arthropathy in relation to sexual functioning. Literature on relevant other diseases, like rheumatic disease, are often more female oriented (Yilmaz, 2012). Usable questionnaires were developed for patients suffering from rheumatoid arthritis (Gossec et al., 2012).

Either pain or functional limitations may lead to limitations of sexual activities. Pain not only causes physical limitations but also mental problems like depression (Kraaimaat et al., 1996). Mental problems by definition have an impact on sexuality. In a study by Laffosse et al. (2008) 19% of patients with arthropathic pain in the hip complained of severe to extreme pain during sexual activity. In another study (Kraaimaat et al., 1996) 48% of patients suffering from rheumatoid arthritis experienced pain in this situation, indicating that this may cause problems in haemophilia too.

Figure 7. The man lies on his back and may use pillows for support. Advantage: The man can remain passive. Disadvantage: Not easy when the woman has hip or knee problems or calf cramps.



DISCUSSING SEXUALITY

Dealing with sexual problems due to disease or disability is part of medical rehabilitation, but both patients and professionals experience barriers to discuss sexual issues (Post et al., 2008). Treatment of sexual problems caused by haemophilic arthropathy, if given, is based merely on advice, i.e. coaching.

In order to stimulate haemophilia healthcare workers to include questions regarding sexual activities in their regular anamnesis, a standard questionnaire can be added to the medical patient file (paper version or electronic), which has to be completed by the doctor or other team member. Training relevant team members in discussing sexuality is a fair option to develop skills and abilities to advise patients and spouses how to improve sexuality (Gianotten et al., 2006; Post et al., 2008).

From then onwards regular evaluation is of main importance. Have couples been able to find sufficient adaptations in their sexual relationship? It is of utmost importance to make the following inventory of their adaptations:

- Were you able to find alternatives?
- Do you have the feeling that your partner is satisfied with these adaptations?
- Are you able to discuss this with your partner?
- If there is a problem we could ask:
- What limits the patient most?
- What does he want to achieve most?
- Is pain causing the problem?
- Is he feeling depressed?
- Or is it the limited range of motions that hampers the activity?
- Or is it the weakness of muscles or a coordination problem?

GENERAL TREATMENT MODALITIES

There is no literature on physiotherapy focussing on sexuality in haemophilia and we can only hope that functional milestones are used by couples to improve sexual activity according to their desires and needs. In a guideline written by Blamey (2012) a series of pictures is given showing alternative positions. Alternatives for positions during sexual activities are more or less the same as described in Chapter 4. The musculoskeletal system acts in functional chains so a complaint in one arm may originate from a functional problem in one leg or from the other arm and vice versa.

In general, the treatment of sexual dysfunction will depend on the individual patient's symptoms and desires. However, there are some general recommendations including: exploring different positions, using analgesic drugs, heat application and muscle relaxants before sexual activity, and exploring alternative methods of sexual expression.

Always look for the most feasible and reasonable options. In the case of a sore knee joint, why not apply heat (warmth) before starting sexual activity. Or combine heat with rest and keep the total body warm under the blankets. Larger heat packs, for example for the lower back, can be applied to both partners and bring them into the same mood.

Some patients complain of disturbing noises during sexual activity, caused by arthropathic crepitation. In this situation a solution may be to play some favourite music.

PAIN

When pain is the most limiting factor of chronic joint problems we refer to the general pain schedule of the *Haemophilia Guidelines* (Srivastava et al., 2013). We have to distinguish pain in a single joint, and pain in multiple joints. In the last situation pain more often causes other problems like depression (Kraaimaat et al., 1996). As pain in a joint may vary during the day, not all joints are painful at the same time. This may result in 24 hours pain and has a great impact on ADL and quality of life. In this situation sexual dysfunction may be present and painkillers and adaptations are essential, because despite pain, satisfaction of being together with a partner will help to retain quality of life. Moreover an orgasm releases endorphins that diminish pain.

ORTHOPAEDIC INTERVENTIONS

We assume that the quality of sexuality also benefits from successful orthopaedic interventions resulting in pain reduction and improvement of the range of motion. Haemophilia arthropathic pain is in almost all cases the indication for surgery of ankle, knee, hip and elbow. Only during the first weeks after surgery, especially after hip and knee replacement, couples temporarily have to stop their regular sexual activities and find alternatives. When flexion of knees has improved after knee replacement and knees are better stabilised in a ‘doggy position’, this will have a positive effect on sexuality. Unfortunately in haemophilia patients joint function often does not improve after joint replacement.

EPILOGUE

Patients with haemophilic arthropathy may experience limitations in sexual functioning as well as in other activities of daily living (ADL). Although haematologists, physicians and physiotherapists have a tendency to focus on the physical aspects of haemophilia and arthropathy, sexual counselling should be part of the regular activities of the haemophilia care team. As both professionals and patients experience barriers to discuss sexual issues, training of professionals is a necessity. Adequate pain killing is important to improve sexuality and general quality of life. When interviewing patients regarding sexual aspects (see interviews in this book) we learned that couples are very well able to find ways to cope with a limited range of motion of different joints. Referral to a social worker or psychologist may be of help to improve sexuality, but in some cases a sexologist is the better option.

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Lily Heijnen interviewed a homosexual man with severe haemophilia in his early forties.

'My partner is very understanding and actively looks for practical solutions'

Did the fact that you are a person with haemophilia cause problems with starting a relationship?

"It is and was not a real problem, but it plays its part at the back of my mind. It is not obvious that I have haemophilia, but as soon as I have to do certain activities, for instance when on holiday, one can notice my limitations".

When did you tell boyfriends that you had haemophilia?

"Soon after I meet someone and the relation gets steady I say: 'You have to know that I have haemophilia A and that I have certain limitations'. I also tell colleagues and casual friends because when going on holiday they will see me infusing clotting factor and they will notice that I am limited in physical activities like long walks in hilly countryside. And if something happens to me people should know what to do".

What was the reaction of your boyfriends?

"I had some 'loose' boyfriends before my present steady one. One of my first steady boyfriends was very nice, but too anxious and overprotective. I felt suffocated and eventually I ended our relationship. My present partner and I have a fifteen year-long monogamous relationship. My haemophilia was no issue for him. He gives me full support and we need few words. He is very understanding and actively looks for practical solutions. For instance on a city trip he finds alternatives for long walks and when needed he arranges for a wheelchair".

When you make love, are you afraid of getting a bleed?
"The only thing I fear is a bleed caused by anal penetration (being the receiving partner). So I never did and will not do this. I imagine that more persons with haemophilia who are homosexual are thinking about this. Furthermore I have no anxieties. In the unlikely event that I get a joint bleed, I consider it a calculated risk. I do anal penetration with a condom and do not have any problems. By the way, people may think that this is something that all homosexual men do as soon as they meet. But that is not the case. For me it was one of the later steps in the development of my relationship with my present partner".

Have you ever seen blood coming with your sperm?
"Yes, once and that frightened me. I consulted my physician and he diagnosed prostatitis (which can happen to any man). So I am not worried that it will happen during sexual activities".

Does pain in your joints or muscles limit your sexual activities?
"In the past I have had bleeds in my ankles, elbows and knee. In recent years, I have pain in my ankles while standing and walking, but that doesn't bother me during sexual activities. My elbows are not too bad and if one elbow hurts, I can always adapt my position and use the other arm and hand to stroke my partners' penis. And 'worst case scenario', with two elbow problems you can still use your mouth".

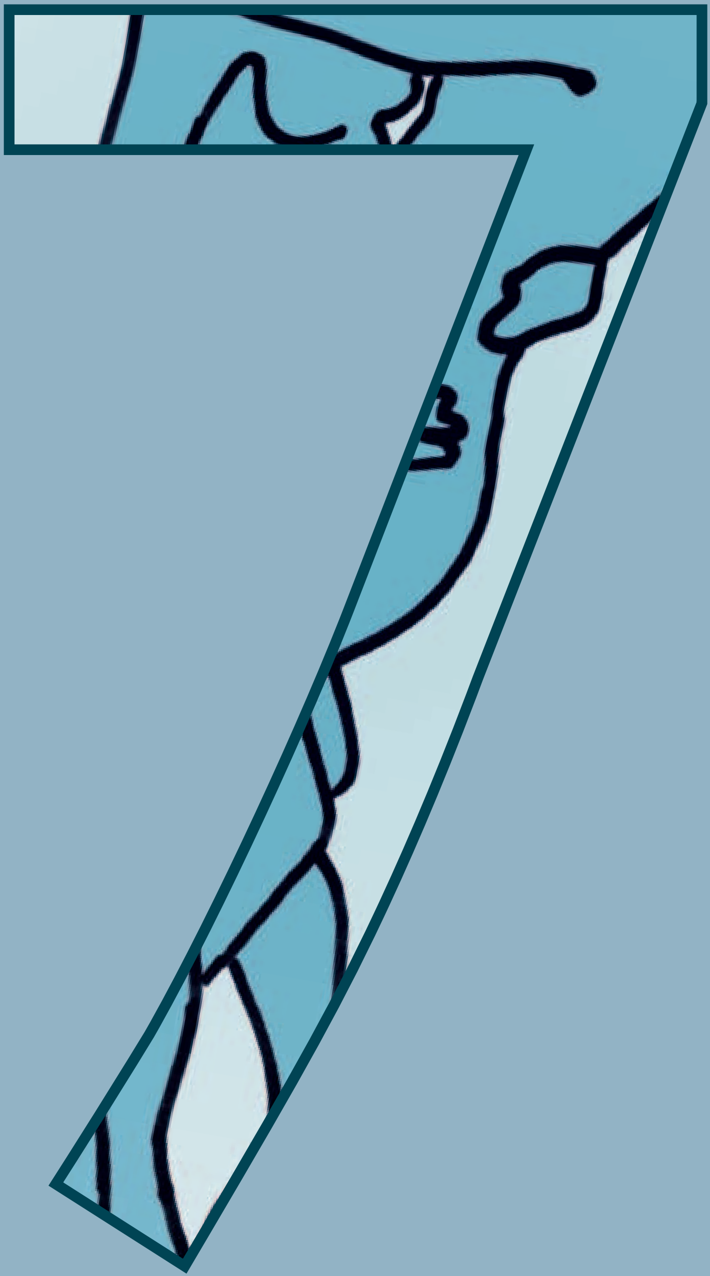
Do you have hepatitis C and did the medication for hepatitis C treatment have an effect on your relation?
"Yes, I am HCV-positive and HIV-negative. I told my partner straight away. At this moment I get medication, which gives me a tired feeling on day one and two. And of course if you are tired you are less inclined to engage in vigorous sexual activities".

Did the fact that you have haemophilia influence your homosexual development?
"No, my homosexuality and haemophilia are two non-related aspects. As a boy and adolescent I differed from the other boys in two ways. In the first place I struggled with my haemophilia because it

caused limitations, which made my life difficult. So I was busy coping with the consequences of my many ankle and elbow bleeds.

I was thinking about my feelings for girls and boys too during my puberty. But the timeframe was different from nowadays. I only came out years later when I was in my twenties. In those days society was less open and there were less role models. Nowadays boys can get so much information on television, internet and social media. Accepting that I was homosexual made my life much easier. A new world opened itself for me. It was really a big difference.

It is more difficult for me to accept the limitations caused by my haemophilia. I am working on that, because when I am able to accept my haemophilia like I did my homosexuality, life will become easier".



DANIELLE H.M. VAN ELST-LAURIJSEN AND
ANDY I.M. HOEPELMAN

Sexuality and infectious diseases

This chapter aims to raise insight in HIV, HCV, its impact and awareness about the effects of being HIV- or HCV-infected on sexuality.

INTRODUCTION

Over the last few decades the treatment of bleeding disorders has improved tremendously. Today the majority of haemophilia patients are expected to live a long and active life. The main breakthrough came when deficiencies in coagulation factors were identified and could be replaced by using products (plasma or clotting factors) derived from human blood.

Unfortunately, the treatment with blood products also showed negative consequences; in the Netherlands 17% of the patients with haemophilia were infected with human immunodeficiency virus (HIV) and 66% with hepatitis C virus (HCV) (Mauser-Bunschoten, 1995).

Since 1985, blood products are safe for HIV and since 1992 for HCV. However, at present, in our treatment centre, we still have many haemophilia patients in care who were infected with HIV and/or HCV before these years. Both co-morbidities can affect patients' sexuality.

HIV

Human immunodeficiency virus (HIV), belongs to the group of 'retroviruses' and was identified in the 1980s. HIV attacks the immune system and gradually causes damage, resulting in an impaired cellular immunity (with CD4+ cell count as a surrogate marker). Without treatment and care, an HIV-infected person is at risk for developing serious (opportunistic) infections and cancers, which a healthy immune system could fight off. Acquired immune deficiency syndrome (AIDS) is the final stage of an HIV-infection, and occurs if the body can no longer fight certain life-threatening infections.

With the introduction of combination antiretroviral therapy (cART) in 1996, it is possible to diminish the HIV-viral load (HIV-RNA) to an undetectable level in the blood. Consequently the immune system remains undamaged or is restored and able to act normal. Although cART does not eradicate HIV, with early diagnosis and taking lifelong medical treatment adherently, most HIV-infected persons do not develop AIDS and are expected to live a long and healthy life.

Currently, not HIV itself but cardiovascular disease, non-HIV-related malignancies and suicide are the most important factors in premature death (van Sighem et al., 2013).

HIV AND SEXUAL HEALTH

STIGMATISATION

Unfortunately, more than half of the persons living with HIV/AIDS experience stigmatisation after disclosing being HIV-positive. Stigmatising reactions include avoidance, exclusion, rejection, social ostracism, awkward social interaction, physical distance and being advised to conceal one's status. Stigmatising settings may vary from families, communities, friends, and sexual relationships to healthcare settings (Stutterheim, 2009).

The general population often relates HIV to being sexually obtained and promiscuity. Campaigns to stop spreading HIV increase the fear of sexual relationships with HIV-infected persons, can just lead to stigmatisation and can be perceived as insensitive, especially by people who aren't infected by HIV due to sexual contact (Parish, 2001).

Because of stigma and the fact that HIV can be transmitted sexually, HIV can have impact on one's partnership and sexuality. Questions like: 'When and how do I tell my sexual partner that I am HIV-positive?' and 'What if my sexual partner informs other people that I'm HIV-positive?' are difficult to answer and often discussed in the consulting room at the outpatient clinic.

Some HIV-positive persons even decide to avoid relationships for that reason, especially if they have been rejected before, due to disclosing their HIV-status.

SEXUAL DYSFUNCTION

However, for persons living with HIV/AIDS it is possible to experience a healthy and satisfying sex life. HIV does not necessarily cause sexual problems, but in practice more than half of these people experience sexual dysfunction (Collazos, 2007). Erectile dysfunction, with prevalence up to 67.1% is the most common sexual problem, even in men under 40 years of age and in those with a good immunological response (Perez et al., 2013). Erectile dysfunction is related to an increasing age and to anxiety, for example: fears of virus transmission, changes in body image, infection, stigma and condom use (Santi et al., 2014). Other common sexual problems are loss of libido, vaginal dryness and orgasm related problems (Santi et al., 2014).

Until now, the relation between cART and sexual disturbances remains unclear and studies show inconsistency in the influence of cART on sexual functioning. cART, particularly protease inhibitors, seem to some extent relate to sexual dysfunctions (Collazos, 2007).

INTERACTION WITH cART

The diagnosis and treatment of sexual dysfunction need a multidisciplinary approach in which specialists in both infectious diseases and sexual medicine are involved (Santi et al., 2014). First, a somatic cause, such as hypothyroidism or hyperprolactinemia, should be excluded. To treat erectile dysfunction Phosphodiesterase Type 5 inhibitors (PDE5i) can be prescribed, however with some caution, because PDE5i's (avanafil, sildenafil, tadalafil, vardenafil) may interact with cART. Therefore, when prescribed, interactions should be actively looked for. For example, protease and integrase inhibitors increase the exposure to PDE5i. Therefore the dose and the dose-interval of PDE5i need to be reduced. Caution is especially needed when PDE5i is combined with a ritonavir consisting regimen. On the contrary, non-nucleoside reverse-transcriptase inhibitors decrease the exposure to PDE5i. More information about drug-drug interactions can be found in ARV Guidelines (Panel on Antiretroviral Guidelines, 2013) or the HIV-ichart (Drug Interaction Charts, 2014).

PREVENTION OF SEXUAL TRANSMISSION OF HIV

TRANSMISSION OF HIV

HIV can be transmitted by blood, genital fluids (semen, vaginal fluids and rectal secretions) and breast milk. The risk of transmission occurs if one of these fluids enters someone's body, directly in the bloodstream, in case of unprotected anal or vaginal sexual contact or, however very rarely, unprotected oral sex.

Exposure to HIV does not necessarily lead to the transmission of HIV. The risk of transmission is influenced by the amount of the HIV-viral load, the exposure route, the presence of other sexual transmitted diseases (STD's) and specific genetic factors (Pebody, 2014).

POST-EXPOSURE PROPHYLAXIS

In case of possible HIV-exposure after a sexual risk event, such as unprotected anal or vaginal sex or in case of a ruptured or slipped off condom, post-exposure prophylaxis (PEP) can be prescribed. PEP is a treatment with antiretroviral therapy, which reduces the risk of seroconversion of HIV after a high risk event and is recommended for HIV-negative persons recently exposed. Based on the event, a risk assessment is carried out by an infectious disease specialist, mainly at the emergency unit of a HIV-treatment centre, to ascertain whether PEP is indicated. For PEP to be most effective, treatment should be started as soon as possible after the event and within 72 hours post exposure. After 72 hours PEP is not effective. PEP treatment lasts 28 days. During treatment, monitoring including blood tests will be carried out in a HIV-treatment hospital.

However PEP lowers the risk of HIV, some disadvantages are associated with the treatment. PEP needs to be taken adherently and serious side effects can occur. Consequently, a proper assessment should be made, before starting PEP (NVHB, 2014).

RISK OF HIV-TRANSMISSION

If taken consistently, antiretroviral medication inhibits HIV in its development, leading to a remaining undetectable viral load; an undetectable viral load means a lower risk of HIV-transmission. Antiretroviral drugs that inhibit viral replication reduce transmission of HIV by lowering the concentration of HIV in the genital tract. In 11 of 13 observational studies, in which antiretroviral therapy (ART) was provided to an HIV-infected index case, transmission of HIV to sexual partners was significantly reduced (Siegfried et al., 2010). The HPTN 052 randomised controlled trial done at 13 sites in nine countries showed that ART used in combination with condoms and counselling reduced HIV-transmission up to 96.4% in HIV-discordant couples (one HIV-positive the other not) in which the HIV-positive partner is on cART, in comparison to couples in which the HIV-positive partner was not on cART (see more on HPTN 052 in Grinsztejn et al., 2014).

The risk of HIV-transmission in steady partners in a serodiscordant relationship is considered 'extremely low' if (van Bergen, 2011):

- The person with HIV has sustained a viral load below the limits of detection (less than 50 copies/ml blood) for six months or longer as a result of cART and
- The person with HIV is adherent to his or her therapy and gets checked on a regular basis.
- Neither partner has any sexually transmitted infection (STI), or has any damaged mucous membrane (anus, penis or vagina).
- Both partners must also have no current risk of contracting an STI in the period since their most recent STI check-up.

Serodiscordant couples that meet the above conditions, can choose not to use condoms, however, they should always realise that the risk of HIV-transmission is low and is not zero. Therefore, the general advice still is to use condoms.

HIV AND REPRODUCTIVE HEALTH

With the introduction of cART the life expectancy of persons living with HIV/AIDS has increased dramatically, and turned HIV into a chronic instead of a lethal condition. Since HIV is a chronic condition, having children is a more common desire than it was before the cART era. This desire is strengthened by the diminished risk of HIV-transmission from mother to child and during sexual intercourse as an important effect of the treatment with cART.

'Family planning' has to be addressed once in a while by the HIV-treatment team (e.g. doctor and/or clinical nurse specialist). Patients and their partners should be well informed about both the low risk for HIV-transmission and the several possible options, which can be applied to fulfil their wish for a child. Based on the given information, a patient and his/her partner can make a reasoned decision at the right time.

In detail: HIV-positive women can give birth to a healthy non HIV-infected child. When cART is initiated before or early (usually after week 12) in pregnancy and the HIV-RNA has reached an undetectable level of less than 50 copies/ml blood before delivery, the risk of vertical transmission (transmission from mother to child) is extremely low (Panel on Antiretroviral Guidelines, 2013).
Discordant couples who are not willing to take the (low) risk of a HIV-transmission, the following options are open to discuss:

- For HIV-infected females with HIV-uninfected partners, the safest conception option is artificial insemination, including the option of self-insemination with a partner's sperm during the ovulatory period.
- For HIV-infected men with uninfected female partners, the use of sperm preparation techniques, in combination with either intra-uterine insemination or in vitro fertilisation, can be considered.

In the near future, periconception administration of antiretroviral pre-exposure prophylaxis (PREP) for HIV-uninfected partners may offer an additional tool to reduce the risk of sexual transmission (Panel on Antiretroviral Guidelines, 2012).

HCV

Hepatitis C Virus (HCV) is a single stranded RNA virus. Seven different subtypes have been described, denoted from 1 to 7, of which genotype 1 is the most prevalent. The genotype determines both the success rate and the type and duration of antiretroviral therapy (European Association for Study of Liver, 2014).

TREATMENT

The currently available treatment for HCV is based on pegylated interferon, which is linked to modest cure rates and many side effects, including depression (El-Atrebi et al., 2011). Interferon free therapy with direct-acting antivirals that have very few side effects and high cure rates will be available in the near future. This may even make eradication of HCV possible without intravenous drug use in countries such as the Netherlands.

LIVER DISEASE

Chronic HCV is one of the main causes of liver disease worldwide. The long term impact of HCV-infection is highly variable, from minimal changes to extensive fibrosis and cirrhosis with or without hepatocellular carcinoma (HCC). In addition of being a major cause of liver related mortality, chronic HCV-infection causes substantial morbidity due to disabling symptoms, such as fatigue and depression, as well as extra hepatic manifestations.

ROUTES OF TRANSMISSION

Up to the 1990s, the principal routes of transmission were blood transfusions, unsafe injection procedures and intravenous drug use (IDU). Currently blood products are screened for HCV, which has virtually eradicated transfusion-associated hepatitis C. In the developed world, new HCV-infections are related to unsafe medical or surgical procedures, IDU, tattooing or acupuncture with unsafe materials (European Association for Study of Liver, 2014).

HCV AND SEXUAL TRANSMISSION

Medical opinion varies considerably regarding the transmission of hepatitis C virus (HCV) through sexual contact. Regarding heterosexual transmission, the weight of evidence is that there is no increased risk of sexual transmission of HCV among heterosexual couples in regular relationships. This risk increases among persons with multiple sexual partners but this association may be confounded by increased likelihood of injection drug use with increased number of partners. There appears to be a real increased risk for women co-infected with human immunodeficiency virus (HIV) or other sexually transmitted infections (three times) and especially for HIV-infected gay men, who are having sex with one another, compared with HIV-uninfected men (5 times). HIV-infected gay men increase their risk of such transmission in association with practices that lead to mucosal trauma (multiple sexual partners, fisting, use of sex toys) and the presence of genital ulcerative disease (Tohme & Holmberg, 2010).

HCV AND SEXUAL HEALTH

SEXUAL DYSFUNCTION IN HCV-POSITIVE MEN

Although sexual transmission has been an area of research in HCV-positive patients, sexual dysfunction in these patients received less attention (El-Atrebi et al., 2011). The frequency of sexual dysfunction (SD) in men with chronic HCV-infection is currently not well studied.

Several studies demonstrated that sexual dysfunction is significantly more common in men with chronic HCV-infection than in control subjects without HCV. Men with chronic HCV have noticeably reduced sexual function in all domains (sex drive, erectile function, ejaculation, sexual problem assessment and overall satisfaction) (Danoff et al., 2006). In one study a strong association was seen between SD and advanced hepatic fibrosis or cirrhosis (El-Atrebi et al., 2011). Possibly biologic (such as diminished breakdown of female hormones in patients with cirrhosis) or virologic factors may be contributing to sexual dysfunction, but further research is required to underpin this. Until now, studies showed conflicting results about the role of depression in relation to SD (El-Atrebi et al., 2011).

As HCV-infected men experiencing SD have a significantly lower health related quality of life (Danoff et al., 2006), sexual dysfunction in HCV is of key importance for clinical practice.

SEXUAL DYSFUNCTION IN HCV-POSITIVE WOMEN

The frequency of sexual dysfunction in HCV-positive women is not very well studied yet, but seems to be even worse than for HCV-positive men (Soykan et al., 2005). The degree of impairment of sexual function in female HCV-patients is found to be higher than in patients with breast cancer (in both before and after therapy) (Elshimi et al., 2014). Women with HCV scored significantly lower in all sexual domains (desire, arousal, lubrication, orgasm, satisfaction and pain) (Elshimi et al., 2014).

Female Sexual Dysfunction (FSD) could be considered an extra-hepatic manifestation of the disease, as sexual function measurements of patients with HCV improved if a sustained virological response (permanent viral clearance) was achieved and decreased in those who relapsed (Elshimi et al., 2014). FSD might also be explained by the presence of other extra-hepatic manifestations of HCV, such as sicca-syndrome with the occurrence of dryness affecting the eyes, mouth and occasionally the vagina. Furthermore, being diagnosed with HCV is associated with psychosocial consequences, which can affect the risk of FSD. Further research is needed to conclude definitively, whether the presence of the hepatitis virus or the psychosocial factors are responsible for FSD (Elshimi et al., 2014).

HCV AND REPRODUCTIVE HEALTH

HCV-infected persons need to be informed about the consequences and risks of HCV and the current treatment on reproduction.

The current available antiviral therapy is contraindicated in pregnancy (Floreani, 2013). Ribavirin carries the risk for teratogenicity. Male patients taking ribavirin must take precautions to avoid pregnancy, during treatment and until seven months thereafter. Females in childbearing age need to use effective contraceptives during HCV-treatment up to four months after treatment is finished (European Medicines Agency, 2011). HCV-positive women are advised to treat HCV before pregnancy, because a successful treatment (sustained virological response) eliminates the risk of HCV-transmission from mother to child (vertical transmission) and reduces the risk of liver progression in mothers (Floreani, 2013). With the new direct acting antiviral agents (DAA's) coming to the market in 2014-2015 interferon free treatment for as short as six weeks will be possible. Therefore, treatment of all HCV-infected women who want to get pregnant is an actual possibility.

Vertical transmission is considered the main route of HCV-infection in children. The overall rate of mother-to-child transmission for HCV is 3-5%. Co-infection with HIV increases the rate of mother-to-child transmission up to 19.4%. Other important risk factors for vertical transmission are high viral load (defined as at least 2.5 x 10E6 HCV copies/ml) and invasive delivery supporting procedures. Caesarean section does not decrease perinatal HCV-transmission from mother to child, thus elective caesarean delivery should not be offered (Ghamar Chehreh et al., 2011).

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Annemarie de Knecht-van Eekelen and Lily Heijnen interviewed an older man with haemophilia. He is HIV-positive, caused by injections with plasma FVIII in the eighties. He has no relationship and lives on his own.

‘My parents never told me that I could die of haemophilia’

Did the fact that you are a person with haemophilia cause problems with starting a relationship?

“I never had a real relationship. I am not sure if that is because of my haemophilia or otherwise. I had two brothers with haemophilia, both of them died: one already young, he had a gum bleed; the other died of AIDS in 1990, he also had been infected with HIV through plasma products. This brother was married, so in itself haemophilia is not a constraint for getting in a relationship. But, there has been a girl who ended the relationship with my brother when he got a gum bleed.

My mother never told me what was wrong with me, she only mentioned that I had a ‘painful leg’, and the doctor said it couldn’t be cured.

In primary school, I managed relatively well with eight to ten bleeds a year. At high school I missed too many lessons because of bleeds, I had to repeat a class. It was a boys’ school, so at that age I had few contacts with girls. It was not easy to be accepted by my peers, I offered them to ride my moped, which was a way to get their attention.

A psychological test showed that I have a high IQ and I decided to continue my studies taking evening classes. I finished those, but I did not get a job, I needed psychotherapy to deal with my personal problems. One of my problems is the difficulty to get into contact with others”.

Do you practice masturbation?

"Yes. When I was an adolescent I was together with a couple of other boys and they showed how to do it. It was quite an experience even though that first time I didn't manage successfully. I have never been afraid of getting a bleed caused by masturbation. Once there was some blood in the sperm, I was worried and consulted a urologist. But there was nothing special to be found and it didn't influence the pleasure of masturbation. I don't need devices or porn to get an erection, my fantasies are good enough".

You are HIV-positive?

"Yes, but it doesn't influence my life. My parents never told me that I could die of haemophilia, so I was never afraid, it didn't affect me. And that is the same with being HIV-positive. It is just there and I live with it. In 1985 when I got to know that I had been infected, it was not yet known that you could die of an HIV-infection. When my brother died of AIDS, I didn't think of implications for myself.
If I would have a woman I would tell her that I am HIV-positive".

Do you experience influences of medication you take?

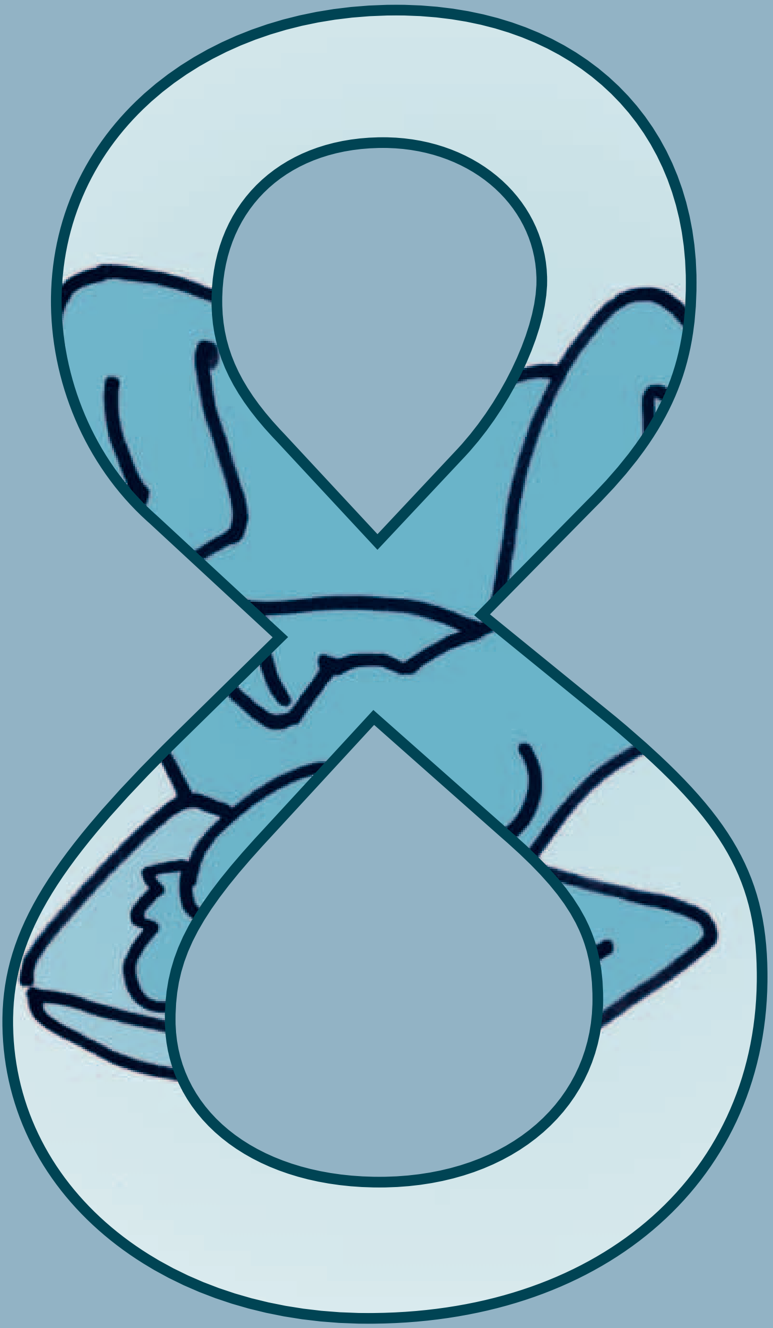
"My HIV-medication has no influence on the erections. I take pills to reduce my cholesterol level, Lipitor (atorvastatin-calcium), they have no influence on sexuality. In the past, when I took Cipramil, it is a psychopharmacon, I experienced libido reduction".

Did your childhood have its influence on your relationships?

"Yes, I think that my parents didn't want me. I was the last born in a large family; three of my siblings had died already. In those days the church forbade the use of contraceptives. If you did you wouldn't enter heaven and no sperm should be spilled, these were the rules. My parents never treated me as a healthy person. My difficulties in establishing a relationship arise from the fact that I wasn't appreciated for the person I was. Even my sisters didn't know what was wrong with me, no one informed them either. One of my sisters got married but she isn't carrier".

Do you want to have a relationship with a woman?

"Yes, I would. At present I work as a volunteer in a spiritual centre for personal growth where I do meet women and I must say that, in such a safe environment, I feel at ease, which makes it easier to confide in someone. So, perhaps ...".



RUUD BOS

Psychosocial impact of sexuality in patients with bleeding disorders

This chapter describes the emotional and psychosocial impact of sexuality in patients with a bleeding disorder and provides knowledge and tools for healthcare professionals on how to raise the subject and how to discuss sexual topics with patients in a professional way.

INTRODUCTION

Research literature shows that in people with chronic illness, physical disability or handicap the sexual functioning might be disturbed. That is obviously not perceived as such by each patient. However, some of the chronic patients suffer from physical limitations leading to physical, psychological, social or emotional problems in relation to their sexual functioning and the way they experience their sexuality. The healthcare professional may not sufficiently be aware of the fact that illness, physical limitations and disabilities can lead to the disruption of sexual functions. Therefore, it is often not a topic for discussion.

SPEAKING ABOUT SEXUALITY

For people with chronic illness, physical disability or handicap intimacy and sexuality are important aspects of the quality of life just as they are for healthy people of any age. Not speaking about sexuality might give a person a feeling of isolation. Assuming the patient has no questions about sexuality because he did not raise the subject is a false presupposition.

Many patients consider sexuality as an important issue and they do have questions, but they find it difficult to raise the subject. But those who don't ask, will get no answers. The healthcare worker should start asking and if the patient really does not want to talk about sexuality, he will indicate so.

Topics such as intimacy and sexuality need to be introduced, whether or not discussed by the patient himself, and should become an integral part of the patient's history, and medical and psychosocial coaching. The patient has to experience that sexuality is a serious and normal subject to talk about. Only then sexual problems can be prevented, and information and education can be provided. Counselling or referring to professional sexual care can be considered when regarded as necessary.

EMOTIONAL IMPACT OF SEXUALITY

For patients with a bleeding disorder sexuality is often a delicate balance of emotional and physical issues. How they feel may affect what they are able to like and do. A chronic disorder can have a major influence on the patient's sex life because of reduced joint functions, stiffness of muscles and joints, fatigue, pain, or loss of strength. Reduced endurance may limit sexual behaviour, positions and pleasure. Per-

Figure 8. Side position with partners facing each other. Advantage: This is comfortable for both partners and eye contact is possible.

haps the patient is not able to have sex and enjoy it as he once did before.

ANXIETY

Fear and feelings of uncertainty can be the cause of decreased sexual drive. When people experience anxiety (for pain) or uncertainty during sex they might avoid sex. This is often unconsciously. Rather no sex than be afraid. That's a very understandable and natural response. Also sometimes anxiety and uncertainty make people just have sex against their will, for example because they do not want to lose their partner. Fears can be:

- fear for haemospermia;
- fear that sex will cause a muscle (psoas) bleed, a joint bleed or a penile subcutaneous bleed;
- fear that sex will cause pain. Mostly women fear pain, but men with joint problems also can fear pain. Uncomfortable positions and vigorous strained muscles and joints cause pain and discomfort. The advice is to try to keep muscles relaxed and 'be kind to your knees';
- fear for transmission of HCV and/or HIV;
- fear for not reaching an erection or not remaining aroused. Men with a bleeding disorder often fear a possible bleed of the erectile tissues when they become too aroused. Women fear that they are not as aroused as their partner;
- fear of losing control over themselves. Some patients are afraid to let themselves go, especially during orgasm, because they do not have their body under control;
- fear of rejection. Many people are afraid of being rejected. Sex is an intimate activity. It makes people emotionally, physically and relationally vulnerable.

FATIGUE

Fatigue can be short lasting or chronic. In both cases fatigue seriously affects sexuality. Someone who feels very tired certainly has no desire to have sex. In general people are recommended to have sex during the time of the day when they feel rather well and have more energy, for instance in the morning hours. If the efforts are too strenuous a different approach or technique, which is less tiring, is advised. Find out which positions are possible. It does not always have to be an intercourse. One can enjoy sex in another less tiring way.

MEDICAL TREATMENT, PHYSICAL EXAMINATION, DRUG SIDE EFFECTS

It isn't only the disease and the associated emotional and social impact that affect intimacy and sexuality. Medication, an operation or prolonged hospitalisations have their influence. Some medications have side effects and decrease libido. For example opioids in chronic pain, anti-inflammatory medicines, and anti-depressants cause reduced urge and sexual feelings.

STRESS

Patients with a bleeding disorder face the same daily stress that affects people of any age. They may be more concerned about ageing, illness, retirement, and other lifestyle changes, all of which may lead to sexual dysfunction. Stress can cause people to get into a pattern of reduced sexual activity as it gives no fun and often fails when they are stressed or cramped. A warm bath or relaxation exercise might help. Some people just have more sex, not for entertainment, but rather as a distraction or to get rid of their worries.

DEPRESSION

A depression or not feeling comfortable in your own body affects a person's sex life, desire and fun. In particular patients on HCV-treatment experience such feelings. During a depression people have less spontaneous sex, but also less interest in finding time and energy to stimulate sexual desire. A low self-esteem often gives the idea of being neither cute nor attractive enough to entice the partner.

SHAME

There are different types of shame because of which people can't quite let themselves go and prefer to avoid sex. By not allowing sexuality to enter their life, they are protecting themselves. They might be ashamed because of their own sexual feelings, sexual fantasies or sexual orientation. It can be a result of their education: they were taught that these feelings and thoughts are wrong or sinful.

NEGATIVE BODY IMAGE

Patients with a chronic disease or handicap do have the risk of having a negative body image. That means that they are not satisfied with their appearance. Negative thoughts make a bolt, and they can no longer see what is good and beautiful in themselves. Feeling and thinking in this way affects the patient's sexuality immediately. Having sex can be confronting in that situation. They prefer to keep a safe distance to the partner.

Hinder in sexual contact

Feeling insecure about their body might hinder sexual contact. Having sex is with the whole body, so sex confronts them with what they dislike about their body. It can be difficult for the patient when the partner touches a part of his body or looks at it. Consequently arousal or intimacy is hindered. Concerns about the way the partner thinks about his body may distract the patient from focussing.

Keeping distance

Another consequence of having a negative body image is keeping distance to the partner and avoiding getting harmed by critiques about the body. However, keeping such distance is contradictory to arousal and intimacy. This feeling might lead to avoiding intimacy and sexuality.

Not knowing your body: less arousal

The third effect of a negative body image is not fully knowing your own body. Patients who dislike their (handicapped) body won't be interested to discover what they like and what is satisfying in sexual, physical contact. It's more difficult for them to let their partner know the way they like to be stimulated, which results in not becoming stimulated optimally. This causes feelings of uncertainty and decreases sexual fun.

PSYCHOSOCIAL IMPACT ON RELATIONSHIP

The impact of the disease or handicap on the relationship needs to be seen from the life or family perspective. It is obvious that young patients without a partner will have other problems than patients within a relationship. For adolescents with physical limitations, experimenting with sexuality, social roles and relationships might cause feelings of anxiety and uncertainty. Sometimes they receive less stimulation in developing a more independent life and have less opportunities to experiment because of an (over)protecting attitude of the parent(s). A chronic disease like a bleeding disorder raises questions for adolescents: are they attractive to others, can they rely on their body having sex, what are their chances to get a long-term relationship. Feelings of uncertainty affect positive self-esteem, cause feelings of shame or lead to social isolation. This hinders initiatives to establish social or sexual relationships. It might even lead to cutting off social contacts and avoiding sexual initiatives.

On the other hand, for others, a disease and handicap motivates them to prove that they are able to live a normal life.

ROLE CHANGES

A long-lasting partner relationship is what most patients or handicapped people are longing for too. Bleeding disorders, physical limitations or handicap can put a great burden on the relationship and family life. In case of a partner relationship, where the healthy partner is taking care of the patient, the relationship-balance might be disturbed. The partner / lover in a nursing role may lead to loss of equivalency in the partnership. In case the partner needs to take over tasks and responsibilities, the patient finds himself in a dependent role. This has its impact as the change of positions and roles implies loss of personal freedom for both the partner and the patient. Because of patient's daily fatigue, family life can be affected as there is less time to do things together as a couple. Patient and partner still feel connected, but there may be an emotional distance arising. The intimate partnership and sexual relationship can suffer from this increasing emotional distance.

A different situation is possible as well. The partners can get closer because of a disease or handicap. Taking care for the patient can create a new alliance between them. It is a challenge for both to find a new balance with possibilities for personal development. Getting rid of rooted social roles – being active, passive, and initiator – is advised to reach a new stable situation. It might be good to find conditions in which the patient does not feel being a patient for a while!

FEELINGS OF GUILT

Being confronted with a bleeding disorder or physical limitations puts a burden on the partner relationship. A lot of patients feel guilty about not being able to have sex during the time when their partners are so engaged in taking care of them. The feeling of guilt is understandable, but not justified. Diseases and handicaps are not chosen by the patients! Feelings of guilt can cause distressing moods, less fun in sex and relational problems. Talking about these feelings can help to cope with them.

FINDING A NEW BALANCE

The sexual desire can be changed by a chronic disease. In fact not everyone has the same need for a sexual relationship. But the disease can provoke existing relational problems and increase these. Sometimes the partner gets problems in reaction to the disease. These feelings are related to caring, fear, solidarity, grief or disappointment. Patient and partner are challenged to discuss these changes and find a new balance.

An active sexual life depends on what people do themselves, it does not always have to include intercourse. Intimacy can also be valuable: affection, hugging, kissing, touching, and spending time together are enjoyable. Give priority to the partner. Take time to enjoy each other's company and to understand the changes one is facing. Try different positions in bed and other times of day, like having sex in the morning when both are relaxed. Don't hurry: one may need to spend more time touching to become fully aroused. Masturbation is a sexual activity that many older people, with and without a partner, find satisfying. Talking to each other about one's personal needs and wishes is the key word.

EPILOGUE

This overview of the emotional and relational impact of a chronic illness on sexuality shows that a chronic illness has its impact not only on the patient, but also on the healthy partner, the relationship and the family life. In healthcare this implies the need for a more systematic approach in research and clinical practice to understand the interaction between a chronic patient – someone with a disability – and the partner and families. The quality of life of the patient also depends on the quality of life of his partner and social environment.

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RUUD BOS

How to discuss sexuality?

Talking about sexuality seems to have become more common in the Netherlands. Nevertheless, in healthcare talking about sexuality and sexual problems is anything but obvious. This chapter gives healthcare workers practical guidance on how to discuss the topic of sexuality.

INTRODUCTION

Many well-educated professionals do not know how to tackle the subject of sexuality, even when the patient mentions it. Their own taboos, embarrassment and other emotional barriers and lack of skills make professionals avoid the subject. The theme gets little or no attention in the training of professionals and if it is mentioned it covers knowledge and not the conversational skills about how to raise the subject. Intimacy and sexuality are considered as belonging to the private domain of people or these issues are perceived as commonplace topics with no interest for the healthcare worker. Curiosity is the wrong attitude, especially when it relates to your own patients. For patients, talking about sexuality might feel uncomfortable and embarrassing. The barrier to talk about sexuality is high for both professionals and patients. Avoiding the subject is obvious! Nevertheless patients with a chronic disease or handicap do find it meaningful to talk about their intimacy, sexual desires, needs and (im) possibilities.

WHAT TO DO IN THE TREATMENT ROOM?

Usually many professionals talk easily about sex when the patient himself starts the conversation. Implicitly the patient gives permission to deal with the subject and also points out what direction the conversation should take. Bringing up the topic is asking for a proactive attitude of the professional in a professional way. It is recommended to integrate intimacy and sexuality in the anamnesis or discuss it in the treatment context. For professionals working in a team, it is apparent that not everyone has to talk about the subject with the patient. A professional conversation needs an adequate context; it does not appear out of the blue. The right discipline, the right moment, the right purpose, these provide the proper context.

Preparing the interview with the patient one must know who takes part in the conversation. Does anyone of the team have a special relationship with the patient? Does anyone have a specific role or expertise? Is there a specific reason for the conversation related to any side effects of medications on sexuality? In these situations the treating physician is to speak with the patient. If there is a problem with joint constraints it might be an assignment for the physical therapist. Requests for assistance or advice related to the emotional and relational impact is the area of the nurse and the social worker.

Also, the purpose of the conversation is of interest. What do you want to achieve with the interview? Is there a request for help? What is the request for help? Is it about information or to solve a sexual problem? An interview may be required to clarify any potential contamination risks of existing infections with HCV and/or HIV. Talking about sexuality can also be aimed at the evaluation of possible side effects of medications.

STARTING THE INTERVIEW. THE FIRST STEP!

Many professionals fear that they might raise the topic of sexuality too quickly or abruptly. A gradual approach ensures an adequate interview structure in which sexuality fits naturally into the conversation. This supports the patient to feel more comfortable and to speak more easily. For example, start with talking about the relationship of the patient followed by the intimate aspects of it. Listing questions about intimacy and sexuality in the patient's history might be a good starting point. Another opportunity can be found in using results of research or other patients' experiences, allowing questions on sexuality to sound natural for the patient. For example start with a question like:

"Many patients with joint problems are experiencing changes in their sexuality. How is this for you? Have you noticed any changes since your own limitations?".

It may be helpful to acknowledge and name the taboo of sexuality. For many people sexuality is a sensitive topic and difficult to speak about. Dealing with this first and clarifying why the subject is important in this context will put the patient at ease. An example:

"Many people do not find it easy to talk about their sexuality. Nevertheless it is an essential part of their existence. Do you mind if I ask you a few questions about your sexual activities?".

If the team or the hospital organisation has developed policies on how to deal with sexuality it may support the professional in raising the subject. An example:

"In our hospital, we feel it is important to cover the topic of sexuality as part of treatment. Therefore I would like to ask you a few questions".

THE PROFESSIONAL APPROACH

People have their own norms and values: Ethnicity, culture, education, religion, media, all influence the development of these standards and values. Although the variety is huge, people tend to relate to one's own values and standards. It shows a professional attitude to take into account the values and norms of the other and respect the great diversity in human sexuality, even when this differs from our own ideas.

Transparency is of great importance. Every professional should have a clear answer to give to him/herself and his/her colleagues on the question of why and how he/she talks with the patient about sexuality and about what specific topics. Personal motives such as curiosity, personal commitment, love or own sexual needs can obviously not be a motive for such an interview.

It is also important to stick to the own discipline, knowledge and working area and otherwise refer to a colleague with other expertise such as a sexual therapist or sexologist.

The success of treating sexual problems depends on the underlying cause. The outlook is good for dysfunctions related to a treatable or reversible physical condition. Mild dysfunction related to stress, fear, or anxiety often can be successfully treated with counselling, education, and improved communication between partners.

TIPS AND RECOMMENDATIONS

- Start the conversation in the right context, for instance related to a side effect of medication, pain or in case of a joint dysfunction;
- Choose an approach as described above to start with;
- Accept for yourself a sense of discomfort and deal with it;
- Take your time to educate and inform; be aware of patient's fears and prejudices;
- Speak preferably with the patient and partner together;
- Adapt your language to the patient's personality;
- Be aware of a history of sexual abuse;
- Start talking about sexual topics with a medical question about sexuality;
- Leave room for diversity, do not fill in for the patient;
- You don't have to talk about all the details, discuss as much as you yourself find necessary;
- Be aware of your own professional limits; refer to a sexual therapist or sexologist when a problem is outside your range of knowledge or competencies.

EPILOGUE

A proactive attitude of the healthcare professional is needed to raise the topic of sexuality. The conversation about sexuality can make an important contribution to the quality of life of the patient with a bleeding disorder. However, it also improves the attitudes and skills of the healthcare professional. Training experiences show that integrating the theme of intimacy and sexuality in coaching patients in general and during the rehabilitation process in particular enhances the relationship between healthcare professionals and patients. It improves the quality of their own professional action. It is our advice to include intimacy and sexuality in the patient's medical history on a regular basis.

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‘I have never been afraid of a bleed’

‘My partner is very understanding and actively looks for practical solutions’

‘My parents never told me that I could die of haemophilia’

To commemorate the fiftieth anniversary of the Van Creveldkliniek, Centre for Benign Haematology, Thrombosis and Haemostasis at the UMCUtrecht, the Netherlands, this publication in the series *Haemophilia care and treatment* has been compiled. It addresses an issue that is often not discussed: sexuality. For patients and doctors, sexuality is not an easy topic to talk about and therefore it is frequently neglected. This is the case in general practices and in the interaction between patients and their specialist, so little is known about what might be a problem for a person with a bleeding disorder, let alone what might be a solution to the problem.

Our question for this book was: 'Is there something special about sexuality and people with a bleeding disorder that needs attention and are there ways to improve guidance and intervention?'. Research on sexuality and bleeding disorders is lacking. How haemophilia patients (i.e. males) adapt during sexual intercourse has not yet been studied.

The absence of lust and difficulties with erection or ejaculation are problems that increase in older age. Pain and invalidity in older patients may also have a negative influence on sexuality. The men with haemophilia who have been infected with hepatitis C (HCV) and/or HIV can have physical and emotional problems with sex, especially because of fear for virus transmission and side effects of the medication. Sexuality and von Willebrand Disease is also discussed as this disease may affect sexuality, especially in women who suffer from excessive menstrual blood loss.

Finding a way to deal with a problem starts with a diagnosis and no diagnosis is possible when people don't talk about sex. This book aims to improve openness and discussion between people with a bleeding disorder and the caregivers.

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