Irish Haemophilia Society

Ageing & Haemophilia

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We are now living in a time where the first generation of people with haemophilia will have to confront the challenges of ageing. Life expectancy figures for people with haemophilia from various studies vary from a median of 63 to 75 years and many people with haemophilia can expect to live beyond their 70th or even 80th year. This is very reassuring and a testament to the improvement in comprehensive care and availability of factor replacement therapy for people with haemophilia over the past decades.

The normal challenges and causes of significant morbidity and mortality which confront an ageing population will now also have to be faced by people with haemophilia. Cardiovascular disease, cancer, renal disease and diabetes are some of the challenges which may await people with haemophilia. These conditions are well characterised and have proven treatment pathways but these may be complicated by the underlying haemophilia or co-morbidities associated with haemophilia. The pathway to monitoring, diagnosis and treatment for diseases of ageing is one which will have to be worked out between the haemophilia services and general practitioners.

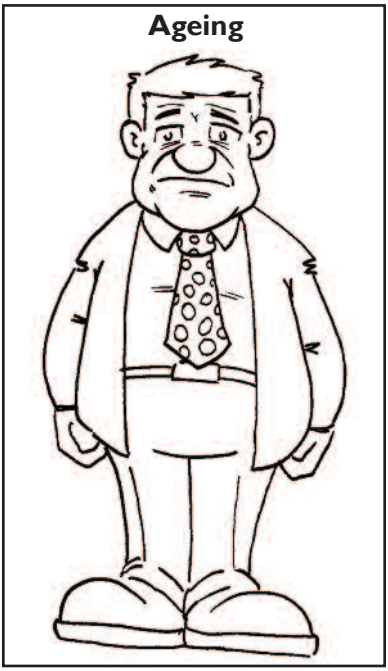
We are committed to taking a strong role, on behalf of people with haemophilia, to ensuring that the most effective pathways for monitoring, diagnosis and treatment for conditions associated with ageing are put in place. This starts with information. This booklet contains a broad overview of medical issues associated with ageing and haemophilia. I have no doubt that these challenges will be faced by people with haemophilia with their usual resilience, fortitude and proactive involvement. We hope that the information in this booklet will be useful in assisting you with information which will help to maintain your health in later life.

While no one wants to dwell on the degenerative effects of ageing, and the problems they may bring, being prepared can go a long way towards alleviating these challenges.

Brian O'Mahony
Chief Executive

AGEING

Ageing is the term used to describe the decline of physical ability (loss of hearing, sight and mobility), appearance (such as wrinkles and loss of hair) and mental agility (ability in retaining and processing information, old or new) that is experienced as we get older. This inevitable process happens at different speeds in different people, and for different reasons. Medical assessment can quantify ageing by measuring heart, brain and kidney performance among others.

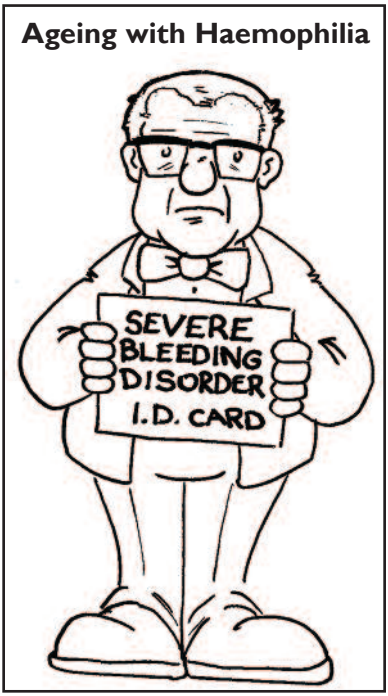


AGEING AND HAEMOPHILIA

Joint disease

Prophylaxis with factor concentrates has been shown, if started early enough, to reduce the effects of haemophilic arthropathy. However, many adults with severe haemophilia did not have the benefits of prophylaxis as children and therefore have significant joint disease and other factors associated with repeated bleeding into joints such as deformity, muscle weakness and impaired balance. These can cause difficulty with mobility and result in pain on a regular basis. Also, the combination of joint disease and advancing age can leave people with haemophilia at an increased risk of falls and social isolation due to reduced mobility.

Maintaining joints in the best possible shape as we get older requires a co-ordinated approach.



Using appropriate pain relief for arthropathy when required. Options that are common in people with haemophilia are:

- Paracetamol (used cautiously in patients with concomitant liver disease).
- COX-2 inhibitors (these are anti-inflammatory drugs that have minimal effect on platelet function). **Note: These can cause bleeding in the stomach and it is important that their use is monitored carefully and that they are used only following a discussion with your Haemophilia Treatment Centre.**

Other methods of pain relief, such as Rest-Ice-Compression-Elevation (RICE), splinting and walking aids (e.g. crutches, canes, orthotics etc) are reliable and can provide some help when carrying out daily activities. Also, light daily exercise and physiotherapy are important with joints that have severe or repeated bleeds. Joint bleeding accounts for more than 90% of all serious bleeding events in persons with severe haemophilia. This risk of bleeding persists throughout life, and the potential benefits of prophylaxis in some form may be experienced by all age groups. The use of prophylaxis in adulthood has increased in recent years. It may be recommended to continue, restart, or even start prophylaxis on an ongoing basis or a short term basis for a number of reasons. This will help to:

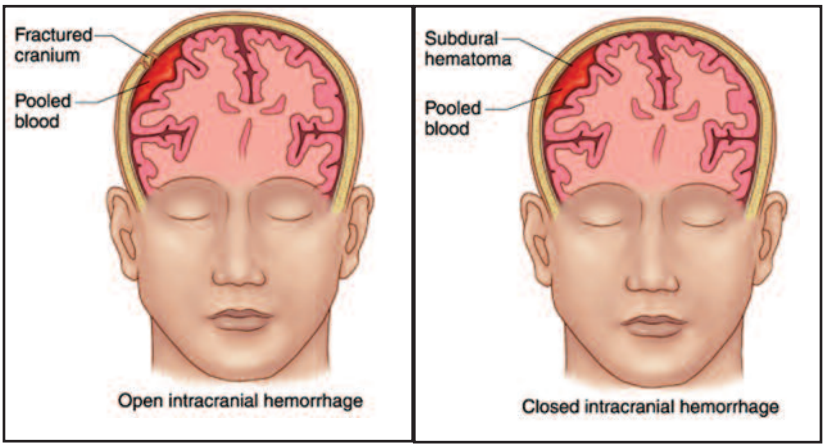
- Prevent bleeds.
- Preserve joint function or slow the progression of joint damage.
- Relieve pain associated with bleeding and/or synovitis.
- Allows for gentle exercise and reconditioning.
- There is some evidence that bleeds can increase after the age of 50 and prophylaxis will reduce the likelihood of this happening.

A Synovectomy can also be used to maintain joint function and/or reduce pain in target joints. Steroid injections and total joint replacement are other interventions for relieving severe pain and disability. The results of hip and knee joint replacement surgery in people with haemophilia, under 55 years old, are similar to the general population. It is important to note that people with haemophilia, in particular those with HIV, may be at increased risk for infectious and non-infectious complications after surgery.

Being overweight can put extra strain on your joints, which can lead to further problems with arthritis, so trying to maintain an ideal body weight is important for preserving joint health.

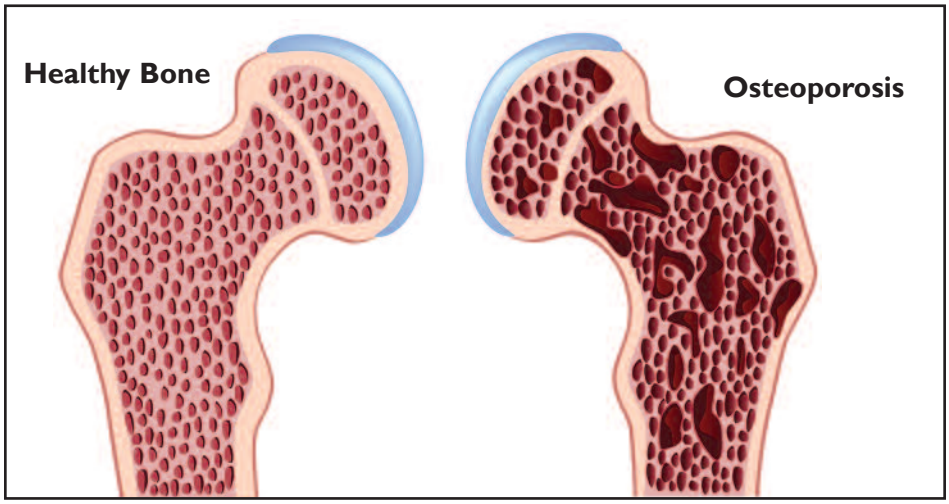
Inhibitors

There is a small increased risk of developing inhibitors as a person with haemophilia ages. It is more common in people with haemophilia A and generally the risk does not increase with haemophilia B. These can occur after receiving factor concentrates for surgical procedures and should be monitored regularly during and after surgery. The management of inhibitors in older people with haemophilia is a challenge. This is not only because of bleeding, but also the complications of alternative treatments for inhibitors, as they may cause clotting.



Intracranial Haemorrhage

Intracranial Haemorrhage is the third leading cause of mortality in people with haemophilia, after HIV and Hepatitis. It also has the highest mortality rate of any haemophilia related bleeding. Thirty percent of people with haemophilia, over the age of 50, are reported as dying as a result of Intracranial Haemorrhage. The majority of these were not as a result of trauma, such as fall. It can sometimes start as a headache. If you have had some sort of trauma to the head or are experiencing a severe headache (without a history of migraines) take a 100% factor rise and contact your Haemophilia Treatment Centre immediately. You may be required to keep factor levels up for a number of days after bleeding and prophylaxis may be suggested.



OSTEOPOROSIS

There may be an increased risk of osteoporosis in individuals with haemophilia. As a result this may make the replacement of joints more problematic. Another aspect of ageing is that although prophylaxis may prevent haemophilic arthropathy, it is unlikely to have an impact on the most common type of arthropathy in older individuals, i.e. degenerative or osteoarthritis.

The best prevention is to build up peak bone density by 35 years of age. However, if osteoporosis has already developed it is possible to prevent further deterioration and reduce the risk of fractures by:

• Exercise: Weight-bearing exercise may help to retain minerals in the bone. Activities such as walking, hiking, swimming and pilates are low impact and may improve bone density and lower the risk for developing bone disease.

• Lifestyle changes: Remove risk factors such as smoking and moderate alcohol and caffeine intake. A diet containing calcium, phosphate and Vitamin D will help to improve bone strength.

• Medication: A specialist doctor may advise firstly taking calcium and Vitamin D supplements, but if the fracture risk is significantly increased other drugs are available.

• Steroids: Should be avoided if possible.

• Surgery: May be required, especially if a joint is damaged.

SLIPS, TRIPS AND FALLS

Slips, trips and falls can happen to anyone, but they are more common and more significant as we get older. This is even more important in people with haemophilia due to repeated bleeding, joint deformity, muscle weakness, and impaired balance. About 10% of falls in people with haemophilia result in serious injury. Many falls are preventable and injuries from falls can be minimised. Physiotherapy and an active lifestyle are important for the preservation of joint function and improving your quality of life. They also play an important role in reducing the risk of falls. Inactive or unfit people tend to have poorer balance and weaker muscles, and can be unsteady when walking. Over time this actually makes you more likely to fall because your muscles get weaker, your joints stiffer and your balance gets worse. Physical activity can:

- Improve balance.
- Improve muscle strength and flexibility.
- Keep bones strong.
- Increase energy levels.
- Help with sleeping problems.
- Help to control blood pressure, blood sugar levels and weight.
- Help you to feel good about life.

What you can do:

• Be physically active every day. Whatever your age, aim to do at least 30 minutes of activity, at least 5 times a week.

• The activity should make you breathe a bit faster and your heart pump a bit harder, but you should still be able to talk while doing the activity.

• Exercises and activities that make you both stronger and improve your balance, lower the risk of having a fall.

• Some examples are Tai Chi, dancing, or group exercise programmes that include balance and muscle strengthening.

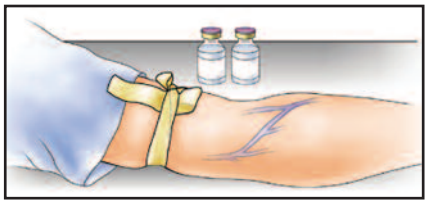
• Talk to a physiotherapist about what balance and strengthening exercises or activities will best suit you, and how to manage any pain you might have.

• A physiotherapist can also advise you about whether a walking aid, such as a stick or frame, would help you. Using a walking aid can increase your steadiness and confidence to walk more. It is important that walking aids are properly adjusted for you, and that they are properly maintained (e.g. replace worn stoppers).

• If you have arthritis, being active helps to control pain, weakness and stiffness.

• If you are unable to walk, it is still important to get outside and socialise. Aids such as mobility scooters should be considered.

VEIN ACCESS



A child with a bleeding disorder is well educated to understand the importance of the role of their veins as they get infusions of prophylaxis. As we grow older we begin to see changes in our body. Our skin can wrinkle and sag, our hands become less steady and our eyesight can deteriorate. All of these make infusing more difficult. Therefore, the importance of good vein care is vital.

Some examples of how to maintain good veins include:

• **Vein Rotation**

Find three or four veins that are easy to access and rotate between them when administering factor. If you have time to take your factor, use the more difficult vein. Never rush when giving an infusion. You may find you are much quicker when using the easy or "reliable" vein, but if you overuse this vein, it won't be reliable for too long.

• **Arm Exercises**

Keeping fit and healthy helps strengthen muscles and joints which can help develop good veins.

• **Vein Exercises**

The staff at the haemophilia treatment centre can help with some exercises to help build veins, but you can try the following: Secure a tourniquet to your arm and squeeze a fist or a rubber ball in your hand. Repeat this twice a day every day for no longer than 5 minutes at a time.

If you are in hospital for infusions or blood tests, ask the nurse/doctor not to use your "reliable" vein. Staff are trained to access more difficult veins and this will provide some rest for your reliable vein.

It is important to know what not to do:

• Do not infuse into an area that is swollen, inflamed or sore to touch.

• Do not put pressure on the injection site until the needle has been removed.

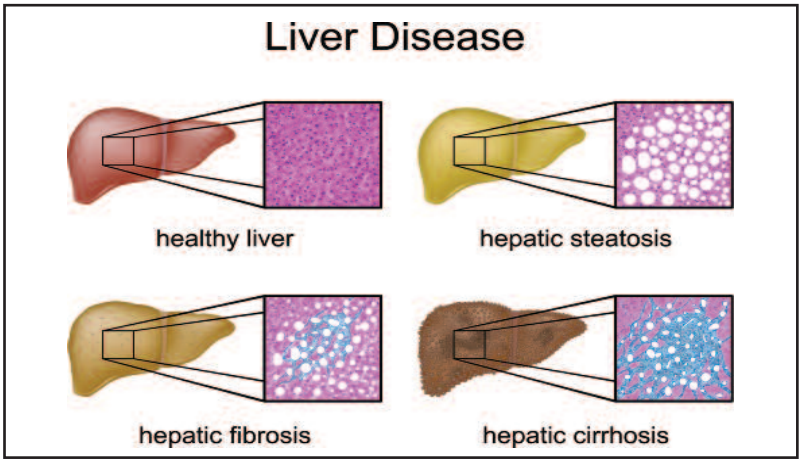
VIRUSES

HIV

As a result of Highly Active Antiretroviral Therapy (HAART) people with HIV infection are living much longer with a better quality of life. However, the ageing process whether on long-term HAART or not is still not well understood. What is known is that HIV infection causes individuals to age prematurely due to the way the virus affects the body. The use of long term HAART has meant that HIV-related conditions are now less common as the virus is suppressed. However, this new increase in life expectancy has resulted in other HIV related complications,associated with ageing, becoming more common and occurring sooner in people with HIV than in the general population. Therefore, more attention needs to be paid to age related symptoms sooner than might normally be expected. In this booklet the increased risks associated with the effects of ageing and HIV are briefly mentioned in the relevant sections.

Hepatitis C

Hepatitis C (HCV) is the major cause of chronic liver disease, liver failure and liver transplantation, and the leading cause of death in people with haemophilia. Approximately 20–30% of people with haemophilia with chronic HCV develop cirrhosis. The likelihood of cirrhosis developing increases rapidly after 15 years of infection. Further development to early stage liver disease, and eventually liver failure, increases with age. The risk of end-stage liver disease and liver failure is increased by between 4 and 8 times in those co-infected with Hepatitis C and HIV, compared with those with single mono-infection of Hepatitis C. Alcohol should be avoided completely by those with Hepatitis C.

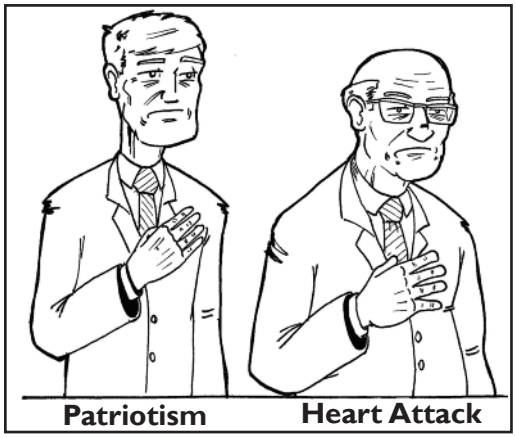


Management of Hepatitis C

A liver biopsy is still the gold standard for determining the impact of HCV and damage to the liver. It is also useful in guiding treatment decisions. Factor replacement therapy for invasive procedures, such as biopsies, will be required and should be done in a hospital with a Comprehensive Care Centre. In Ireland the recommendations for monitoring HCV in people with haemophilia are as follows:

	Non-cirrhotic or Fibrosis stage unknown	Cirrhosis, Pre-Cirrhotic or Bridging Fibrosis
Hepatitis C PCR Positive & HIV Positive	• 3-4 monthly review in Co-infection Clinic as directed & review in Hepatology as recommended • Liver ultrasound as indicated by physician • Liver function tests & blood test for alpha-fetoprotein 6 monthly	• 3-4 monthly review in Co-infection Clinic & review in Hepatology as recommended • Liver ultrasound 6 monthly • Liver function tests & blood test for alpha-fetoprotein 6 monthly • OGD every 2-3 years and more frequently, if varices identified
Hepatitis C PCR Positive & HIV Negative	• 6 monthly review in Hepatology • Liver ultrasound as indicated by physician • Liver function tests & blood test for alpha-fetoprotein 6 monthly	• 6 monthly review in Hepatology • Liver ultrasound 6 monthly • Liver function tests & blood test for alpha-fetoprotein 6 monthly • OGD every 2-3 years and more frequently if varices identified

CARDIOVASCULAR DISEASE



Cardiovascular disease (CVD) is the leading cause of death in the developed world and is the most common cause of death in Ireland, accounting for 36% of all deaths. The two biggest contributors are ischemic heart disease (IHD) and stroke. IHD is the result of reduced blood supply to the heart muscle, usually through the small arteries around the heart. This is due to plaque building up in one area in the arteries and reducing the blood flow in the arteries (atherosclerosis). This can result in chest pain (angina) and if left untreated a heart attack. Stroke occurs when plaque builds up on the small arteries supplying blood flow to the brain.

Haemophilia has attracted a lot of attention in this area, probably due to the high mortality rate associated with CVD, and the dilemma posed in the treatment of CVD that requires the use of drugs to prevent clotting (antithrombotic agents, e.g. Aspirin or warfarin).

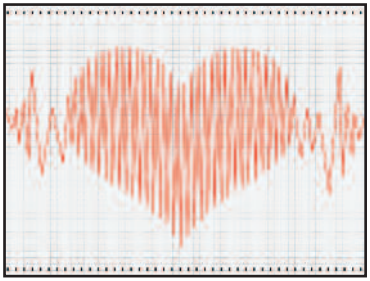
Am I at an increased risk of CVD?

This is a new area for a lot of people with haemophilia and the information is limited. For years, it was thought that people with haemophilia could have a significantly reduced risk of CVD. While some reports suggest that this is true, others indicate little or no preventative affect due to haemophilia. It may also be possible that if there is a preventative affect, there may be a reduction in it with the use of prophylaxis. There is no clear explanation as yet but this area is constantly being explored and more information is on the horizon.

However, if a severe deficiency of factor VIII or IX does offer some protection against IHD, then it may be worth a discussion with your haematologist about any intensive replacement therapy you may require for surgical procedures, and controlled continuous infusion may have an advantage in this scenario.

In individuals with HIV, with or without haemophilia, there is an increased risk to be found independently of IHD. This may be due to the effects of some anti-retrovirals or immune inactivation.

Is the management of CVD different for people with bleeding disorders?



CVD is a significant challenge because of the need to consider the risk of bleeding when using anti-clotting therapy. While information is relatively limited at the moment, the general principle is to correct the clotting factor deficiency by using factor replacement and then treating the patient as closely as possible to standard protocols for CVD.

There are some specific recommendations for haemophilia. They suggest; avoidance of thrombolytic therapy and use of bare metal stents (small scaffolding placed at the point of blockage to prevent a total closure of the artery) and the use of prophylaxis during dual anti-platelet therapy. It is important to note that these recommendations are largely based on opinion as the number of people with haemophilia being treated is minimal at this stage.

As we have an ageing population, there will be an increase in the need for heart surgery such as bypasses or replacement heart valves. Cardiac Bypass has been performed safely in people with haemophilia but requires careful planning and management. Heart valves can be coated in a material that prevents clotting. It is suggested that heart valves without this coating be used in individuals with haemophilia.

Reducing your risk

You can reduce your chances of getting a stroke or heart disease by making certain changes in your lifestyle, especially not smoking and controlling high blood pressure. If you have high cholesterol, lowering your cholesterol levels may also reduce your risk. Your doctor may tell you to change your lifestyle.

TOP 5 TIPS FOR AVOIDING CARDIOVASCULAR DISEASE

1. Stop smoking.
2. Eat a healthy diet.
3. Be physically active.
4. Be a healthy weight.
5. Relax – take time out for yourself.

CANCER

The overall risk of developing cancer, of any type, increases with age especially after sixty. Cancer is a broad term that encompasses over 200 different diseases, banded together because they are all caused by cells that have started to grow out of control. It is estimated that one in three individuals will develop cancer during their lives and the risk for many cancers increase as you get older.

Am I at an increased risk of cancer?

There is no clear connection between haemophilia and an increased risk of cancer. There are a number of conflicting reports on the topic. For people with haemophilia the answer to this question will become clearer in the future.

There are two situations where there is an increased risk in those people with haemophilia who have HIV and/ or HCV.

Firstly, in relation to HIV, the immune system plays a fundamental role in protecting the body from cancer cells by killing cells which contain mutated DNA. However, with age the immune system becomes weaker and more cancer cells can slip through the immune system's surveillance. HIV attacks the immune system, making it less able to fight off diseases and therefore increases the risk of developing cancer. In the past, people with HIV infection typically got three types of cancer, Kaposi's sarcoma, non-Hodgkin's lymphoma and cervical cancer in women. As people with HIV are living longer, they are developing more cancers that are related to ageing rather than to HIV infection.

Secondly, the risk of hepatocellular carcinoma (HCC) is significantly increased in chronic Hepatitis C infection, and as a result, is currently the leading cause of death in people with haemophilia. This risk increases further if the person is over the age of 45 years, was an older age at the time of infection, or has cirrhosis.

Is the management of cancer more problematic in individuals with bleeding disorders?

In general, the management of cancer in a person with haemophilia should be similar to an individual without haemophilia. There may be some medical issues such as the need for factor replacement therapy for invasive procedures (e.g. biopsy or surgical procedures) and prophylaxis may also be needed when platelet counts are low (thrombocytopenia).

Other issues may arise as a result of chemotherapy or radiotherapy. There may need to be monitoring in relation to bleeding as a result of tumours shrinking. This can be prevented with the use of factor replacement therapy. This should be monitored at the Haemophilia Treatment Centre in liaison with your Oncologist.

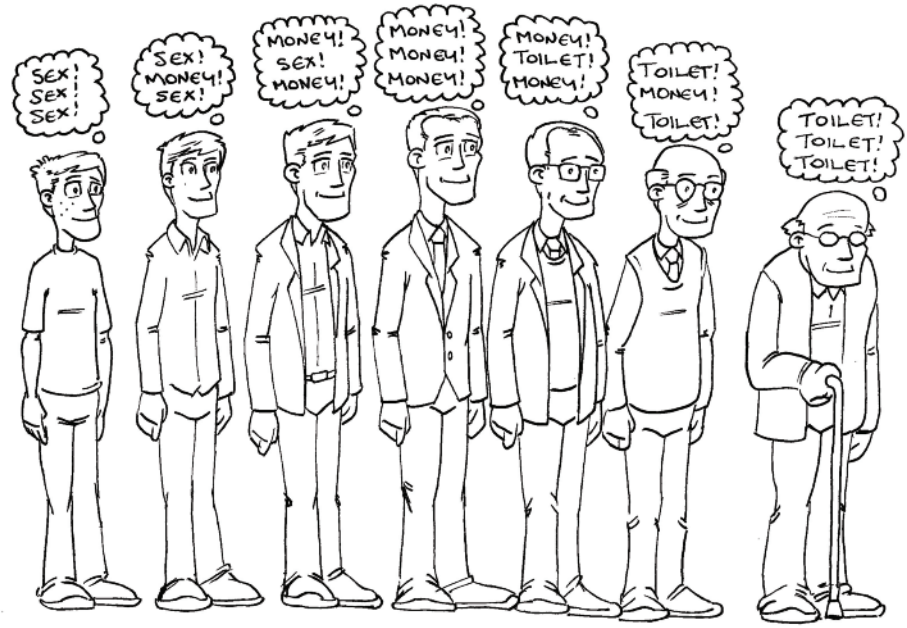
Reducing your risk

It is not always easy to avoid some of the risks to your health, but there are some very high risks that you can partly or totally avoid. There are many ways that you can reduce the risks to your health. To reduce your risk of cancer. Here are some important points to remember:

THE TOP 5 TIPS TO REDUCE THE RISK OF CANCER

1. Stop smoking.
2. Eat a healthy diet.
3. Be physically active.
4. Be a healthy weight.
5. Take care in the sun.

Being aware of early signs of cancer and getting treatment can lead to better survival rates. Have regular check-ups with your GP. Check your body routinely for lumps, moles or bruising that does not heal so that changes can be picked up quickly.



RENAL DISEASE

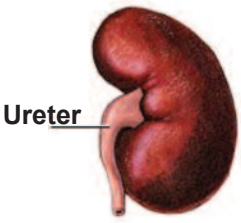
Chronic kidney disease (CKD) is the slow loss of kidney function over time. In the early stages, there may be no symptoms. The loss of function usually takes months or years to occur. It may be so slow that symptoms do not appear until kidney function is less than one-tenth of normal. If chronic kidney disease is diagnosed at this stage there may be a need for dialysis or a kidney transplant. Chronic kidney disease is another important age-related medical issue and is known to become an increased risk factor over the age of 70.

Am I at an increased risk of CKD?

Although the information is still limited, people with haemophilia may have a higher risk than the general population of chronic kidney disease. Risk factors may be inhibitors and kidney bleeds. HIV is also strongly associated with acute and chronic kidney disease.

NORMAL KIDNEY

* Healthy Function
* Proper Size
* Low Urine Protein



KIDNEY DISEASE

* Granular Surface
* Decreased Function
* Smaller Size
* High Urine Protein

Is the management of Chronic Kidney Disease different for people with bleeding disorders?

If kidney disease is more common in people with haemophilia and as is already happening, a population at advanced age emerges, it is likely that more cases of end stage renal failure will be seen in the near future.

The management and monitoring for haemophilia and renal disease is the same as the general population with renal disease. This includes referral to a nephrologist, blood pressure monitoring and treatment, control of cardiovascular disease risk factors and laboratory monitoring. There will be a need for factor replacement therapy for invasive procedures (e.g. biopsy or surgical procedures).

For the treatment of CKD, Peritoneal dialysis may offer advantages for people with haemophilia as factor replacement therapy is often only required for the insertion of the peritoneal catheter, but not for subsequent dialysis procedures. However, this may not be suitable for those with chronic liver disease or HIV disease because of the risk of infection and the concern of peritoneal haemorrhage.Hemodialysis has also been used successfully but may require both the administration of factor concentrate and anticoagulation with heparin during dialysis.

TOP 5 TIPS FOR REDUCING RISK OF RENAL DISEASE

1. Stop smoking.
2. Reduce alcohol intake.
3. Eat a healthy diet.
4. Be a healthy weight
5. Be physically active.

DIABETES

Diabetes mellitus (MEL-ih-tus), or simply, diabetes, is a group of diseases characterised by high blood glucose levels that result from defects in the body's ability to produce and/or use insulin. Type 2 Diabetes, often called non-insulin dependent diabetes, is the most common form of diabetes (90% - 95%).

People with type 2 diabetes produce insulin; however, the insulin their pancreas secretes is either not enough or the body is unable to recognise the insulin or use it properly. This is called insulin resistance. When there isn't enough insulin or the insulin is not used as it should be, glucose (sugar) can't get into the body's cells. When glucose builds up in the blood instead of going into cells, the body's cells are unable to properly function.

The simplest and quickest method of diagnosing diabetes is to measure for glucose in the urine. Undiagnosed type 2 diabetes may cause damage to a number of body systems as follows:

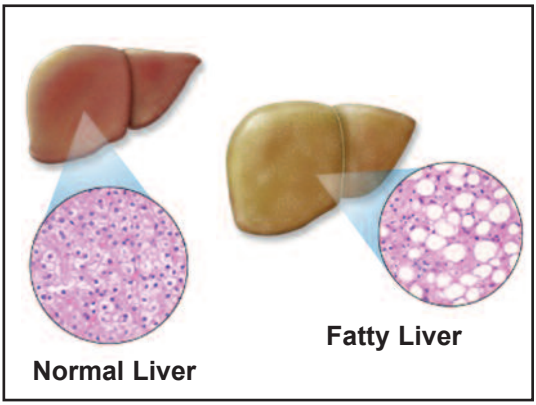
- Dehydration.
- Diabetic coma.
- Other Damage: Over time, the high blood glucose levels may damage the nerves and small blood vessels of the eyes, kidneys, and heart.
- Erectile dysfunction and impotence.

Symptoms of type 2 diabetes

- Excessive thirst.
- Frequent and increased urination.
- Increased fungal infections.
- Slow healing of wounds, including small cuts.
- Blurred vision.
- Tiredness, which may be significant.
- Fluctuations in weight.

The prevalence of diabetes in haemophilia is not well documented. If treatment with insulin is required subcutaneous injections can be administered without bleeding complications.

In relation to Hepatitis C, research shows that there is a higher prevalence of diabetes among persons infected with HCV, up to 3 times more likely than those without HCV. Chronic Hepatitis C may contribute to the progression of diabetes. Diabetes mellitus has been reported to be more prevalent in patients with HCV compared to those with hepatitis B and those with other types of liver diseases. Fatty liver disease is also associated strongly with the development of type 2 diabetes.



Type 2 Diabetes is also common in HIV infection and ageing. HIV medication predisposes the individual to the development of diabetes and certain drugs are implicated more than others; however the more recently developed medications tend to have a lower risk for the development of diabetes. Development of any of the symptoms listed on page 19 should be discussed with your doctor as soon as possible.

TOP 5 TIPS TO REDUCE THE RISK OF DIABETES

1. Eat a healthy diet.
2. Be physically active.
3. Be a healthy weight.
4. Choose the right fats.
5. Stop smoking.

1 in 4 people have mental health problems. Think of your three closest friends... if they seem ok, then you may be the one!

MENTAL HEALTH

There are many factors that can lead to depression, such as retirement, bereavement, loss of mobility, changing your home etc. Retirement can lead to a loss of self-worth and not feeling an active member of society. The reduction in income can also force unwanted lifestyle changes. Bereavement is particularly hard to bear - not only have you lost a loved one, but your lifestyle may completely change. A loss of health or mobility may mean getting out less or becoming more dependent on others. If you have to move home, for instance into a smaller home or into a care home, you may find it hard to cope with the loss of friends and independence. There may also be additional issues that may be associated with haemophilia such as problems with joints and mobility or an inability to self-infuse.

Symptoms of depression may include a loss of appetite, insomnia, weight loss or gain, lack of motivation and little energy. It's normal to feel one or two of these at any time, but if you are affected by a combination of them, talk to your GP about diagnosis and treatment. In relation to haemophilia, any psychosocial and / or physical support you may require should be discussed with your Haemophilia Treatment Centre.

There are steps you can take to help cope with these life changes. Make sure you have activities and hobbies in place when you retire, so that you are not suddenly faced with long, empty days. Use the support of family, friends and local services if you are looking after a partner. Keeping physically and mentally active will help reduce the risk of illness and disease.

If you, or someone you know, needs support, someone to talk to or is in crisis:

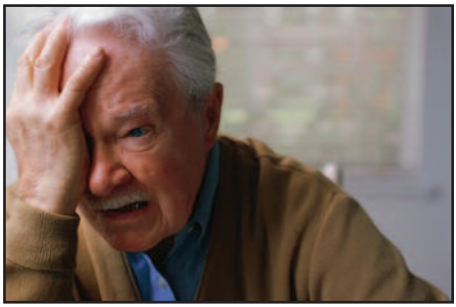
- Contact your local GP or family doctor.
- Contact your GP out-of-hours service.
- Go to or contact the Accident and Emergency Department of your nearest hospital.

Dementia

Memory problems and dementia are a common concern for many people as they grow older. Dementia is the progressive loss of the powers of the brain. There are many kinds of dementia but the most common are Alzheimer's disease and vascular dementia. What all types of dementia have in common is that they damage and kill brain cells, so that the brain cannot work as well as it should.

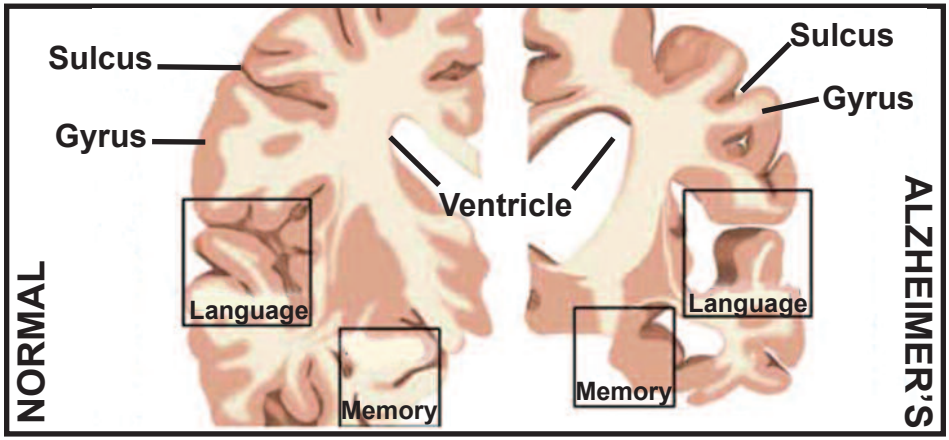
Alzheimer's disease

Alzheimer's disease damages individual brain cells one by one, so that the brain can't work as well as it used to. There may be a genetic factor in some cases of Alzheimer's but this does not mean that someone whose parent had Alzheimer's will automatically develop the disease.



Vascular dementia

This is a result of problems with the blood supply to brain cells. For example, some people have tiny strokes (or infarcts) which damage small areas of the brain.



Symptoms

Every person with dementia is different. How their illness affects them, depends on which areas of their brain are most damaged. One of the most common symptoms of dementia is memory loss. Most people's memory gets worse as they get older but when someone has dementia they may forget the names of family members, they may burn pans because they have forgotten them or forget whether they have eaten lunch. They may repeat the same question again and again and not know they are doing it. They may lose their sense of time, which day it is or the time of day. They may forget where they are, or get lost even in a familiar place. People with dementia may often be confused. Their ability to think can be damaged. They may make odd decisions, find it hard to solve problems and handling money may become difficult. Dementia can also cause personality and behavioural changes.

Confusion or forgetfulness does not mean someone has dementia. Other conditions such as infections can cause similar problems, so see your GP for advice. If your GP finds no reason for the symptoms, they may want to refer you for an assessment by a dementia specialist. Some treatments may be available which may help some people with the symptoms of some forms of dementia (particularly Alzheimer's disease), and research is progressing all the time.

There is no known way to prevent dementia. However, there are actions that you can take to reduce your risk for dementia and, in some cases, slow the progression of the disease. These factors include:

1. Regular exercise.
2. Healthy diet.
3. Mental stimulation.
4. Quality sleep.
5. Stress management.
6. An active social life.

Some factors may decrease the effect of the damage by developing more connections between the remaining brain cells, rather than preventing damage. With more connections between brain cells, function can be maintained longer despite damage to the brain.

LIFESTYLE

Throughout this booklet you will have seen several "Top 5 Tips" to prevent whatever condition it may refer to. You will hopefully have noticed that the top tips are quite similar for most conditions. In this section, we will look at how the majority of these tips can be achieved.

NUTRITION

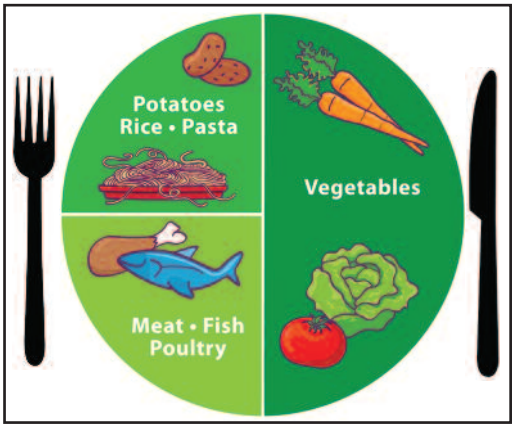
Having a healthy diet is essential for a healthy lifestyle. Make sure your diet is full of fruit, vegetables wholegrain foods and pulses. Wholegrain foods include brown bread, brown rice and brown pasta. Pulses include peas, beans and lentils. These foods help with keeping your energy levels up without piling on the weight. The great thing is they also make you feel as though you are full and may be less hungry.

Each day aim to eat:

- 5 or more portions of fruit and vegetables.
- 6 or more portions of high fibre foods such as brown bread, porridge, brown rice and pasta.

Meat

You don't need to miss out on the meat option. Lean red meat is rich in nutrients like protein and iron and is ok in small portions. Have a look at the mix of food on your plate. The ideal mix is one-third or less of protein (meat, chicken or fish) and two-thirds of vegetables and pasta or rice.



Other options for protein in your diet are eggs for breakfast, and chicken, turkey or fish for lunch some days of the week. With vegetarian options being more widely available in restaurants, why not try a meat free dish the next time you are out, or you can go a step further and try a meat free day from time to time.

THINGS TO WATCH OUT FOR IN YOUR DIET

• TAKE AWAY FOODS

Foods like sausage rolls, pastries, cakes and ready-made meals are best avoided. They often have high levels of fat in them.

• SALT

People tend to eat a lot more salt than they need. Cutting down can reduce your risk for a number of diseases. Try adding less to your food during cooking and at the table. Use things like black pepper, herbs, lemon juice, garlic or spices to flavour your food instead.

• HIDDEN SUGAR AND SALT IN PROCESSED FOODS

The food may not taste sweet or salty but it doesn't mean they aren't packed with them.

SMOKING

It is never too late to quit smoking. After quitting, your risk of getting a number of diseases, including lung cancer, falls as long as you remain a non-smoker. There are a number of options available to help you quit smoking. Nicotine Replacement Therapy (NRT) is available from pharmacies or through your GP. Used correctly, it can be very successful in easing the cravings from nicotine. There are many types so make sure to use the right one for you. You can discuss your options with your pharmacist. You can also discuss these and other methods with your GP.

Quit plan

• List your reasons for quitting.

• Set a date to stop.

• Tell all your friends and relatives, as they may support you. Some people prefer to keep it secret for a while. That's ok too.

• Getting someone to quit with you can help. You will reinforce each other's willpower.

• Clear the house, car and your pockets of any packets, papers, matches or lighters.

• Reward yourself regularly with the money you have saved.

• Ask your friends not to smoke around you. People accept this far more readily than they used to.

• Cravings usually last for 3-5 minutes. If you get a craving use the 4 D's to help you get through it – Drink water, Deep breathe, Distract yourself, Delay grabbing a cigarette for 5 minutes.

You can also call the NATIONAL SMOKERS' QUITLINE on CALLSAVE 1850 201 203. Stop smoking advisors are on hand with confidential advice and support. They will also send you a copy of the Quitting Booklet, a positive and supportive guide to quitting smoking.

ALCOHOL

Alcohol, in moderate amounts, can help people relax and it is a part of most social occasions. Drinking too much alcohol can lead to deterioration of liver or heart function, thinning of the bones and impairment of brain function, especially memory and co-ordination. Many of these deteriorate with age as well. Alcohol is also high in calories. It is difficult to give alcohol up completely, and although this would be the recommended option, generally reducing alcohol intake is a good idea. Try switching to soft drinks, non - alcoholic beers and wines or shandies during a night out. Having some glasses of water in between drinks will help your body to process the alcohol.

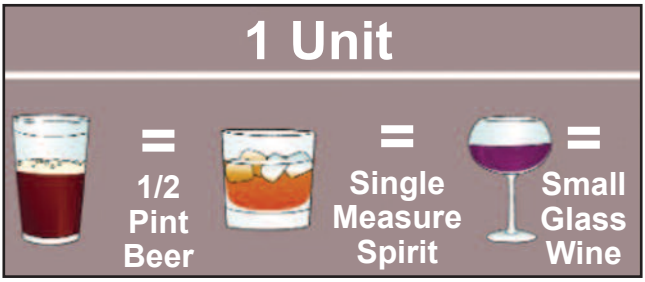
Unfortunately not everything improves with age!

If there are other underlying issues with viruses, such as Hepatitis C or HIV any alcohol intake will have even greater effect on the function of all of these in particular the liver, and ideally alcohol should be totally avoided. Heavy alcohol use in people with haemophilia who have Hepatitis C has been demonstrated to increase the progression to severe liver disease more than 5 fold.

How much is recommended?

Aim for no more than 2 standard drinks per day, with some alcohol-free days.

(1 standard drink = 1/2 pint beer; a small glass of wine; 1 pub measure of spirits.)



EXERCISE

Exercising the mind

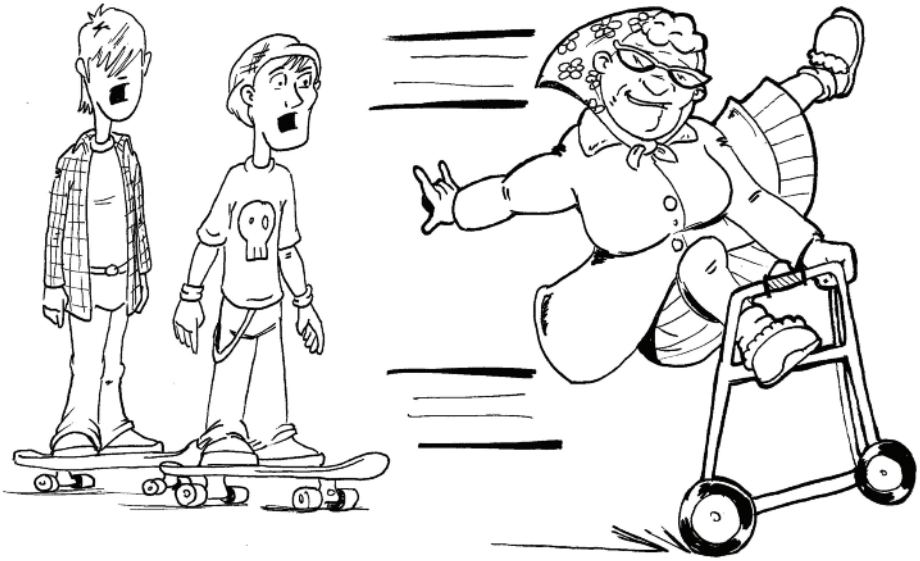
Many activities help keep your brain mentally agile: crossword puzzles,chess, bingo, learning a language or taking a cookery or computer course. It is also possible to train your memory to some degree, for instance by making lists, or using certain 'trigger' words to jog your memory. Meeting new people may be all the mental stimulation you need. For those of you who like technology there is a wide range of games, puzzles and activities for the mind available on phones, computers and other devices. Check your local newspapers and websites to see what's going on in your area. It may seem daunting to go somewhere different or try something new, but it really is worth taking that first step.

Exercising the body

Keeping physically active has a huge number of benefits: it helps to reduce the risk of conditions such as diabetes, heart disease and stroke, it strengthens your muscles and aids mobility. It also encourages a healthy appetite and good sleeping routine. It can reduce the risk of falls, osteoporosis, stress and depression. It stimulates the brain, keeping brain cells alert. Thirty minutes of physical activity each day is what is recommended to keep you active. If you are already active for 30 minutes a day, you could step up the effort.

Physical activity doesn't mean having to join a gym. You could take up a dance class, walk to the shops instead of taking the bus, or tackle the garden, all of which will be keeping your muscles toned and your joints supple. Even if you are not able to be active when standing up, there are lots of seated exercises you can do - you can try chair based exercises. Don't forget local centres and organisations offer a range of activities like walking clubs, swimming, aqua-aerobics or dancing. If you are unable to get out, using a Wii© Fit can be very beneficial in improving balance and muscle strength.

Most people with haemophilia will have joint damage, pain and /or poor balance. If you have difficulty with mobility you should discuss exercise options with your haemophilia centre or physiotherapist. You should begin with easy, range-of-motion exercises and low-impact aerobics to improve your joint strength and fitness and then you can build from there. It may be useful to talk to your haemophilia centre about going onto prophylaxis for a short period when you are starting new activities.



BE ACTIVE!

BE PHYSICALLY ACTIVE FOR AT LEAST 30 MINUTES EACH DAY.

Anything that makes you breathe a little deeper and your heart beat a little faster is ideal. A good example is a brisk walk.

You can spread the activity out over one or more sessions in the day.

This will help you avoid getting a spare tyre and keep the engine ticking over.

Fat but fit?

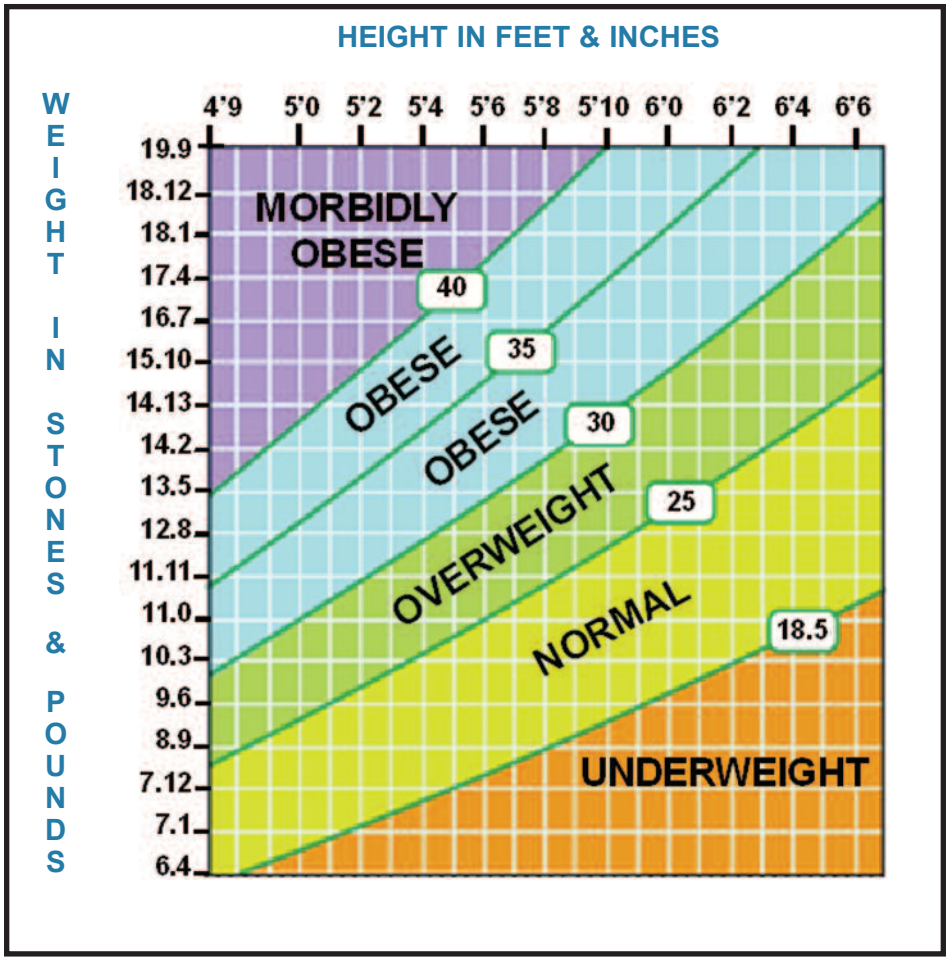
After age 45, the average person loses around 10% of their muscle per decade. This is about one-half a pound of muscle each year. However the body usually gains this amount back in body fat. Because muscles burn a lot of calories compared to fat, the total number of calories needed goes down. So eating the same amount of food as we age can lead to weight gain even if you are active and fit. Extra fat still can cause problems as it increases your risk of diseases such as high blood pressure and heart disease. So, are you a healthy weight?

There are two ways you can check this out.

- Measure your Body Mass Index (BMI)
- Measure your waist.

BMI Measurement

Draw a line across from your weight and down from your height. Where these two lines cross is your BMI. Aim to be at the lower end of the normal BMI range. If you are underweight or overweight, it is important to see your doctor. A medical check-up and advice on diet and lifestyle may be needed. Remember, BMI is not always an accurate measure if you are an athlete or very short in height. A high BMI can be associated with significant limitation in joint mobility, pain, increased risk of developing target joints. If you have limitations, such as poor joint mobility, that restrict daily activities, a physiotherapist may be able to suggest appropriate alternatives. In some cases, referral to a dietician may be useful.



WHAT DOES YOUR BMI MEASUREMENT MEAN?

Less than 18.5
You are underweight, which can lead to some health problems.

18.5 - 24.9
You are a healthy normal weight.

25 - 29.9
You are overweight, which increases your risk of cancer, cardiovascular disease and other conditions.

Greater than 30
You are obese and your risk of a number of conditions is greatly increased.

To measure your waist line
Find the top of your hip bone. At this point, measure around your waist. Make sure the tape measure is snug but not marking your skin. Take the measurement at the end of your normal breath.

If your waist measurement is over 37 inches or 94cm you are at increased risk of cancer.

SEXUAL HEALTH

People often assume that sexual intimacy disappears as you get older. The truth is that desiring sex and having sexual relations doesn't diminish as we get older. It's an inevitable fact that, however young we feel inside, our bodies change physically as we get older.

With haemophilia, there may be other physical limitations that reduce the desire for sexual intimacy as a result of joint restriction, pain and/or balance issues. There are also a number of options that are available such as adapting positions or sexual aids that can assist in sexual intimacy. You should discuss these issues with your Haematologist and/or Physiotherapist at the Haemophilia Treatment Centre.

Remember, safe sex isn't just for younger people - all age groups can get, and pass on, Sexually Transmitted Infections (STIs). Condoms can help to protect against STIs, so talk to your new partner about using them.

HOUSING MATTERS

Home Adaptations

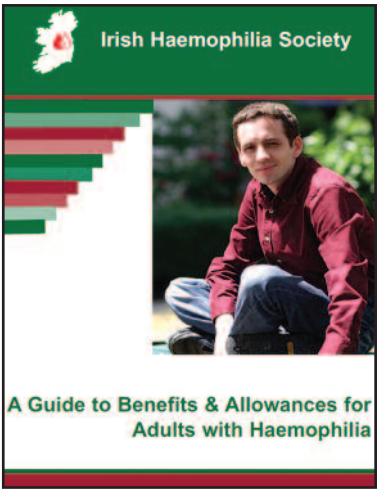
As we age, our homes should be updated to accommodate our changing needs. Home adaptation or modification can provide a better living space so older occupants may continue to live in the comfort of their own home. The aim in modifying your home should always be on increasing and improving the following:

- Self-sufficiency/self-reliance.
- Mobility.
- Safety.
- Security.
- Comfort/convenience.

The best time to start thinking about home modification to meet the needs of aging is long before the need actually arises so that the work can be carefully planned and budgeted. Ideally, people in their fifties and sixties should prepare their homes for later installations while they are doing routine home improvements and repairs. This also allows for easier adaptations in the future without the stress that can be associated with home improvements and also help the adaptation to blend in to your home. An example is extra wide doorways that can accommodate a wheelchair but this can also be an attractive feature in any home. Strong supports can be installed behind tile walls when updating bathrooms to later accommodate grab bars. Before any changes in the home are considered, a thorough room-by-room assessment of the surroundings should be made. This should include consideration of the individual's current and future needs.

Occupational Therapists are trained to identify dangers and make recommendations to alterations in your home to ensure your safety, so consider a consultation. Conversions and extensions can be expensive but health board grants are available. Conditions include an Occupational Therapist's report and means test.

For older people there are schemes whereby the local authority re-houses people in "independent living" secure units or apartments and purchases their house at a reduced price. These units are carefully designed and often include wardens and personal alarm systems. You should contact Citizen's Information or the Irish Haemophilia Society for more information on these and other entitlements and benefits. You should also consider where you live now may not be suitable in the future when you may be less mobile, unable to drive or at a distance from support such as family, services or your GP.



Modifications to your home do not have to be expensive or require a lot of work. Some simple, helpful and cost effective options are listed below:

Kitchen
- Rearrange dishes for easier access and less reach.
- Remove throw rugs.
- Purchase easy grip utensils.
- Replace round door handles with lever handles.
- Replace appliance knobs with easy grab knobs.
- Change cabinet handles and drawer pulls to types that are easier to grasp and remove magnetic clasps if possible.
- Turn down the hot water heater to a safe level.
- Use a folding step with a rail to reach higher objects.

Living Room
- Remove all throw rugs.
- Rearrange furniture to open walk area.
- Add risers to furniture.
- Add extension leads to raise electrical outlet heights.
- Remove the clutter.

Stairs
- Remove carpeting to prevent trips and slips.
- Add extra handrails.
- Add reflective strips.
- Increase lighting.

Bedrooms

- Raise the bed for ease of use.
- Change the doorknobs to lever-style handles.
- Install grab bars near the bed.
- Remove or add furniture, depending on need.
- Remove the clutter.

Bathrooms

- Add grab bars.
- Only use rugs with rubber backing.
- Replace fixtures.
- Add a seat to the shower.
- Raise the toilet or add a raised seat to the toilet.

Hallways

- Add more lighting, with a switch at both ends.
- Add a handrail.
- Place a chair at the beginning or end of a long hall for resting.

Garages

- Use reflective tape on edges of garage door openings for better gauging or space.
- Hang a tennis ball to gauge the stopping spot for your car.

Entryways

- Replace doorknobs with lever-style handles.
- Replace old stiff locks with new easy turn styles with large knobs for easy gripping or keyless entry locks.

Outside living areas

- Build a ramp that is discreet.
- Add motion detectors at the front and back of homes to light up walk areas and to help with safety of resident and ward off mischief.

Additional changes

- Add an amplified phone with large buttons.
- Remove all the unnecessary clutter and get organized. This can prevent falls and increase the safety of the occupant.

REDUCING THE RISK OF FALLS AT HOME

• Being fit and active can help to reduce your risk of having a fall by maintaining muscle strength. Try gentle exercises to improve balance.

• Dehydration can cause dizziness and increase the risk of falling. Make sure you drink plenty of fluids: 6-8 decent sized glasses of water a day. Dehydration can start before you feel thirsty so it's important to drink regularly even if you don't feel thirsty.

• Make your home clutter-free and easily accessible, especially stairs and pathways.

• Light your home brightly by using high wattage bulbs.

• Wear flat, well-fitting shoes.

• Get help carrying heavy objects.

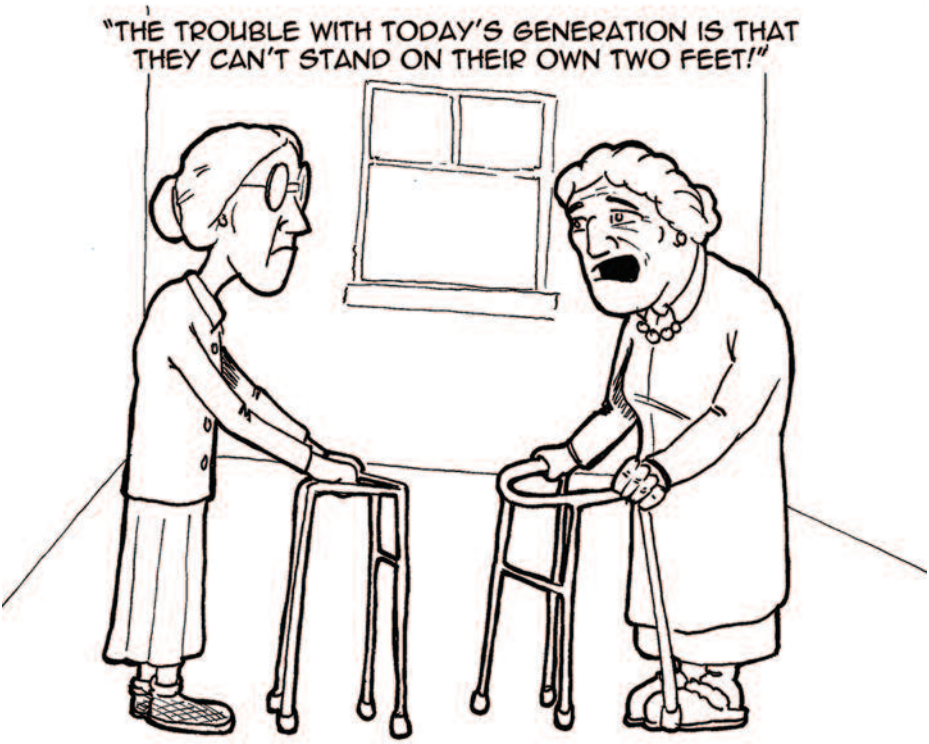
• Considering installing a shower seat and grab rails if necessary.

• Evidence shows that once an older person has had a fall, they are more likely to fall again.

Tell your GP or nurse if you have had a fall.

Make an action plan of what to do if you fall:

- Call for help.
- Keep warm, covering yourself with anything available and try to keep moving as much as you are able.
- Rest before you try to get up if you are able to do so.
- Carry you mobile phone on you as much as possible.
- Consider a pendant alarm (Lifeline) to get help quickly.



GETTING THE BEST FROM YOUR GP

When you make an appointment to see your GP, whether it's for a routine check-up or for a new health issue, there is more to do than just call the office to schedule the appointment. Planning ahead and being prepared can help lower your stress and make your visit more productive, both for you and your doctor.

Firstly, make sure you have your basic identification with you, such as a medical card or health insurance card. It can be easier to have things organized ahead of time.

Decide how you will get to the GP on the day of your appointment. Bring a folder with items you may need for your appointment like new test results you have received since the last appointment, money for taxi's or parking etc. It's easy to forget the most important things during a GP visit especially if you are feeling unwell on the day.

If you make a list before you go, you'll have many of the answers for your doctor. You should make notes on:

- When did it start?
- How did it feel?
- Did this ever happen before?
- What have you done about it so far?
- Are you on any medicines at present?

Don't be afraid to say you may need certain accommodations. For example, if you cannot get onto the examining table by yourself, there should be an adjustable height table in the room or staff to assist you.

Don't be afraid to ask questions if you are not clear on what has been explained to you. It is important for your own self-management that you understand what you need to do if you have a problem and why you need to do it. If you require new medications, ask about side effects or if they will interact with the medications you are already on. If you require tests for a diagnosis, ask why they need to be done, when the results will be back and when you need to return for the results.

In relation to haemophilia, ask that letters from your consultant be copied to your GP and you. This will help your GP to keep up to date with your haemophilia care. It is important that your GP is aware of all other conditions and having a good relationship with your Haemophilia Centre will only benefit your care.

There are a number of reasons you may need to switch GP such as your current GP is retiring, you have only used your Haemophilia Centre, or you are moving home. The Golden Pages should also have a list of GP's in your area or you can check the internet but asking your friends or neighbours can be the best way. Ask your GP about consulting your Haemophilia Centre and what you may need from your GP.

Finally, we hope you have found the enclosed information not only informative, but helpful, in planning and preparing for a long and healthy future. As our knowledge of haemophilia and ageing develops we will keep you informed by periodically updating this book. As bearers of a rare and complicated medical condition, we must acknowledge the members of the medical profession, and pharmacists, who have helped us survive thus far.

Old age is often referred to as 'The Golden Years' - Let's enjoy them!

This booklet is dedicated to our friends who didn't get to share our journey.

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