Condition Insight Report (CIR)

Haemophilia, Haemarthropathy & Infected Blood

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Overview

What is the condition usually called/ any abbreviations used?

Brief overview of the condition.

Haemophilia is a genetic bleeding disorder in which the blood fails to clot normally. Blood clotting is the process by which protein factors and platelets activate one another to form a plug at a site of bleeding, such as a torn blood vessel, to prevent further loss of blood from the site. Over time the initial plug forms into a scab and healing of the torn vessel occurs. Clots are formed by the activation in the blood of special proteins called clotting factors, these factors activate one another in a sequential manner or 'cascade' that results in a blood clot over a few minutes and this stops bleeding. When a clotting factor is missing, only present in low amounts or in an ineffective form the cascade cannot complete. An ineffective clot is formed so bleeding continues. This bleeding most frequently occurs internally.

Bleeding can occur into muscles and joints, both with and without injury. People with haemophilia don't bleed any faster than everyone else, but they will bleed for longer, as they don't have normal amounts of clotting factor. Treatment is aimed at reducing spontaneous bleeds.

If you require any further detail on Capita guidance for these cases. Please access the in-depth document HERE

Types of Haemophilia

Types of haemophilia

There are two types -:

- **Haemophilia A** – this is the commonest type of haemophilia. In this condition clotting factor VIII (8) is abnormally low or absent.
- **Haemophilia B** – this is the less common type of haemophilia. In this condition clotting factor IX (9) is abnormally low or absent. It's also called Christmas disease.

Both types of haemophilia have the same symptoms and are inherited in the same way. Treatment is different depending on which clotting factor is missing. Specialist blood tests are needed to measure the clotting factors to show whether factor VIII or factor IX is affected and how much is missing.

People with haemophilia born from the late 1990's onwards are most likely to have benefited from modern treatment regimens and very few will have any significant impairment related to bleeding complications. This is because they will have been treated with prophylactic clotting factor treatments to prevent serious bleeds from occurring. However, the cohort of people born before the late 1990s will not have had access to this treatment.

All of those with severe haemophilia and some of those with mild and moderate haemophilia will have haemarthropathy: joint damage due to repeated bleeds into joints.

Hepatitis C and HIV affecting some claimants with Haemophilia

Some people with Haemophilia may report they have contracted Hepatitis C, and/or HIV from contaminated blood transfusions and clotting factor products. It is, however, pertinent to suggest that the HP MUST consider, that each of these conditions will have a range of symptoms which themselves can greatly impact on function, but in combination with haemophilia, can result in potentially significant/ severe functional difficulties.

Scale of severity….

Haemophilia is classed as severe, moderate or mild depending on how much clotting factor is missing

Mild haemophilia

People born with mild haemophilia may not have any symptoms for many years or they may have some symptoms, such as bruising easily. The condition usually only becomes apparent after a significant wound or surgery, or a dental procedure such as having a tooth removed. These events could cause unusually prolonged bleeding.

Spontaneous bleeding may be seen in some people with mild haemophilia.

Moderate haemophilia

People with moderate haemophilia are affected in the same way as those with mild haemophilia, but also bruise easily. They may also have symptoms of internal bleeding within their joints, particularly if they have a knock or a fall that affects their joints. This is known as a joint bleed.

The symptoms usually begin with a tingling feeling of irritation and pain in the affected joint – most commonly the ankles, knees, and elbows. Less commonly, the shoulder, wrist, and hip joints can also be affected. The pain may quickly progress to a severe level, even if factor infusion is given promptly.

If a joint bleed isn't treated, it can lead to:
- more severe joint pain
- stiffness
- the site of the bleed becoming hot, swollen, and tender

Severe haemophilia

The symptoms of severe haemophilia are similar to those of moderate haemophilia. However, joint bleeding is more frequent and severe. People with severe haemophilia have spontaneous bleeding and this may occur frequently without prophylactic treatment. This means they start bleeding for no apparent reason

For example, this could be:
- nosebleeds
- bleeding gums
- joint bleeds
- muscle bleeds

Without treatment, people with severe haemophilia can develop:
- painful arthritis**(haemarthropathy)** which may require replacement surgery
- joint deformity
- soft tissue bleeding
- serious internal bleeding

Haemarthropathy

Haemarthropathy is a severe type of arthritis caused by bleeding into the joints due to haemophilia

- Multiple large joints may be affected by arthritis from childhood onwards.
- If there is joint damage, mobility is likely to be restricted because of pain related to haemarthropothy in the upper body (such as shoulders elbows and wrists) and the lower body (hips, knees and ankles for example),
- Joint replacement of individual joints will relieve pain from arthritis and prevent further bleeding into that joint but will not improve mobility or range of movement. This is because other joints are affected.
- If flexion deformity of the knee or hip is present, then mobility is especially likely to be reduced; a flexion deformity effectively shortens the affected leg and affects gait. In someone with multiple damaged joints this places further strain on other joints and increases the risk of bleeding when walking. Remember that upper limb pathology will also have an impact on the ability to mobilise. Arthropathy affecting the elbows is common and affected shoulders and wrists are not uncommon. This will affect the ability to use walking aids and other daily living activities.
- Once the joints are damaged, changes in joint anatomy and inflammation mean they are more likely to have bleeds into them from mild joint trauma associated with, for example, walking short distances. Bleeds can also occur spontaneously.

As always, it is vital that HPs consider the reliability criteria (safety, repeatability, in a reasonable timeframe and to an acceptable standard) when assessing PIP claimants with this condition.

Symptoms of bleeding into joints

- Sensations of joints feeling warm, tingly, 'bubbly 'or stiff can be felt before any external signs become evident
- Joints may become warm, swollen and difficult to straighten
- Pain can increase as bleeding into the joint continues
- As blood is not intended to be inside the joint, it has a damaging effect. Once damage has occurred to the joint, bleeding may happen again spontaneously or on minimal use and more frequently. In addition, any damage to the joint has the potential to be permanent. This is known as a 'target joint' and is a complication of haemophilia
- Joint bleeding causes an irreversible form of arthritis, known as **haemarthropathy or haemophilic arthropathy**
- Significant haemarthropathy in cases of mild haemophilia is very unusual, but not impossible

Haemarthropathy assessment scenario
Poor Vs strong information gathering

HP: So, you have haemophilia and arthritis?

Claimant: Yes

HP: Which joints are affected?

Claimant: My shoulders are not too bad, but my elbows and knees are very bad.

HP: What symptoms do you get?

Claimant: Pain which gets worse when I'm walking, or if my elbow catching on anything. I'm always bumping it on something. They can swell too, so I must be careful.

HP: How many good days do you get? What can you manage on a good day?

Claimant: Oh, well, not many, probably one or two days a week. I don't manage much more; I just feel the pain is a bit better.

HP: Right so what about the bad days?

Claimant: I'm clumsy which is not good when you have a condition like mine, so my joints get sore and swollen if I bump them on anything. Most days are not great. I'm always sore. I've been told I'll probably need knee replacements at some point.

What we know so far:

- The questioning is very superficial
- The HP does not appear to have made the link between Haemophilia and that haemarthropathy is a complication of this condition. The HP is not asking about bleeding into the joints, what happens and how often it occurs.
- Has the HP considered the risk of spontaneous bleeds – are they occurring? How often? What is the impact? What activities caused them? This would **NOT** help the HP produce a high-quality report

HP: So, you mention arthritis, but I want to check if that's linked to your haemophilia?

Claimant: Yes, it is, it's all part of it. I'm told it's a complication.

HP: Do you get bleeding into your joints?

Claimant: Yes, not all the time, but it does happen so I need to be careful.

HP: What would trigger you to have a bleed?

Claimant: It can be something obvious like having a fall, or even if I bump myself against something like a table. Sometimes I can get a bleed and I haven't had any injuries or falls. They scare me as they can come on any time.

HP: Can I ask you more about the unexpected bleeds first, then I'll ask some more questions about the other bleeds?

Claimant: Yes.

HP: Thank you. So how often do you get these unexpected bleeds at the moment?

Claimant: I've had 2 in the last 6 months.

HP: And has your doctor made any suggestions about what might have caused them?

Claimant: Well we discussed what I was doing around these times, and I wasn't doing anything unusual. It was my right hip then my right knee which were affected. Something about my joints being bad affects how I walk which puts me at risk of more bleeds I think.

HP: Right so how long ago was the last bleed and where was it?

Claimant: 3 months ago, in my right knee. I'm still recovering from that.

HP: Can you tell me how many good days you get each week at the moment?

Claimant: Maybe one or two at the most.

HP: What makes it a good day for you?

Claimant: If the swelling is a little less and I don't need to take all the painkillers.

HP: What about bad days each week – how many of those do you get?

Claimant: The rest of my days tend to be bad. I'm usually in a lot of pain and struggle to get around and do what I need to do each day. I can't look after myself independently with things like dressing and showering and that upsets me.

HP: In your daily routine, have you ever experienced any joint bleeds? For example when washing, dressing or cooking?

Claimant: Yes. My most recent bleed when was getting in and out of the bath. I have to sit to dress, but still have had bleeds I the elbows and one in my wrist before when cooking.

<u>How does this probing compare?</u>

This example shows us that the HP has made the link between their haemophilia and the arthritis, and that they understand bleeding into the joints is a factor they need to consider. The danger of not making the link for the Case Manager is that they are left wondering if the HP appreciates the risks of bleeding when doing everyday movements, and whether that together with reliability has adequately been considered when choosing descriptors.

Customer Care & Sensitivities

- People may under report their functional restriction or not want to discuss the detrimental impact of being given infected blood during a transfusion
- This must be addressed sensitively and with empathy
- You must remember that if someone has had treatment with a contaminated clotting factor, they may have HIV or Hepatis C and this may be difficult for them to discuss

REMEMBER: The PIP assessment is a functional assessment. We do not need to be unnecessarily probing around sensitive areas unless we feel it will have an impact of function i.e the impact on someone's mental health secondary to receiving infected blood

The psychological impact on people affected by contaminated blood

'We believe that a lack of long-term specialist psychological support for victims of the contaminated blood scandal is putting lives at risk.'-The Haemophilia Society

The Inquiry heard from infected individuals who highlighted fear of infecting partners as a huge barrier to forming lasting and meaningful relationships. Some 'unattached' witnesses underlined this fact by stating that they had not had any or many

The infection can also have a negative impact on self-perception and self-esteem, and a loss of one's sense of self. (Yang et al., 2016)

'The Infected Blood Inquiry has meant that many people infected and affected by the contaminated blood scandal have been re-traumatised by their memories of that devastating period in their lives. We believe that as the inquiry goes on, the pressure on peoples' mental health will increase. Evidence from clinicians and politicians and other decision makers has been particularly traumatic.'-The Haemophilia Society

The Inquiry heard that very few infected or affected individuals were offered any counselling or psychological support. Yet the evidence reviewed suggests that many HIV/HCV infected people would benefit from psychological support /counselling

Psychological impact continued

It is imperative that you explore the psychological impact secondary to infected blood. Mental Health should be explored as a separate condition, be it depression, anxiety, post-traumatic stress disorder (PTSD) or any other condition.

There has been research and evidence to support the detrimental impact on someone's Mental Health of receiving infected blood as per the earlier slide.

This may not always be the case, and not all claimants with haemophillia will have infected blood, so ensure you assess each case individually.

Possible areas to explore

- **Diagnosis date and who diagnosed** (REMEMBER: lack of diagnosis does not necessarily mean lack of functional restriction)
- **Any treatment or input received for their mental health**. The NHS have specifically funded talking therapies for related trauma, however, there may have been a multitude of other interventions they have received
- **The functional impact.** How does their MH impact their day to day activities? Do they lack motivation? Do they experience anxiety? Do they have flashbacks? Do they socially isolate?
- **With all cases where the claimant suffers with a Mental Health condition, suicide and self-harm must be explored sensitively.**

Clinical Considerations in the PIP process

Initial Review

In all cases of **moderate** or **severe** Haemophilia being reported in the questionnaire or AR1 form, the HP at initial review should request further evidence from the Haemophilia Treatment Centre, as they assess these people every 6 months (which includes musculoskeletal assessment) and they will be aware of and able to detail physical restrictions.

In some cases, this information may already be available to view on PIPCS. This information may then enable the HP to progress to complete a paper-based review (PBR).

F2F assessment

We must <u>NEVER</u> carry out a musculoskeletal examination (MSK) on people at risk of spontaneous bleeding. This includes all those with moderate and severe haemophilia and those with mild haemophilia with a history of spontaneous bleeding and/or haemarthropathy.

This is due to risk of spontaneous bleeds occurring, even when doing everyday movements.

Reliability

What specific areas should be covered to ensure a complete, reflective report?

SAFETY	TIMELY	ACCEPTABLE STANDARD	REPEATEDLY
Do they have any symptoms which could cause a safety consideration?	For any activities where restriction is reported how long does it take them to complete these activities? Has how long it takes them changed over time?	How have they adapted to completing tasks over time – is this different to what might be considered ‘normal’?	Are they able to repeat a task as often as required? Is this the same every day?
Someone with this condition may suffer from joint bleeds and consequently the ability to safely complete activities must be adequately explored.	A musculoskeletal impairment may result in someone reporting they can perform a task, but extra probing may be needed to establish that in fact they only managed it because they were going at a slower pace	Establish whether the person experiences any pain or discomfort when performing tasks. Some people may push through an activity whilst in a great deal of pain or discomfort. Some people can experience an increase in their pain levels after treating themselves with clotting factor, which may further impact on their function – the impact must be explored and made clear for the Case Manager	If someone has joint pain and stiffness this may mean once they have completed an activity, they would be unable to physically move on to another activity or repeat this as many times as is reasonable throughout the day.

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 1: Preparing food

The elbow is often affected, and other upper limb joints may be affected. Range of movement such as restrictions of flexion; extension; reduced pronation and supination and reduced dexterity can all have a large impact on ability. They may struggle to chop and hold/lift pans. They may also experience fatigue and difficulties standing for prolonged periods.

REMEMBER: There is a risk of joint bleeding during this activity if haemarthropathys in the upper limbs.

Furthermore, we must consider the psychological impact of their condition and how this may impact their motivation to complete this activity.

Remember in PIP…

Can someone prepare and cook a simple meal for one? How long can they stand in the kitchen? Can they chop and peel vegetables? Can they lift pans? Does completing this activity increase breathlessness or fatigue? How do they feel after the activity? How long does it take to recover? Do they have pain or are they at risk? Are they motivated?

Activity 2: Taking nutrition

If there is joint damage secondary to their condition, they may struggle to cut up food and, in some cases, may need assistance. If there is severe joint damage, they may struggle conveying mood to mouth.

They may also report reduced appetite secondary to Hepatitis C.

If someone reports lack of motivation secondary to their condition this must also be considered here if it affects their motivation to reliably take nutrition.

Remember in PIP…

Can they chew, swallow and bring food to their mouth? Can they physically cut food? Do they have any aids to assist such as adapted cutlery? Can they eat solid food? Do they have any risk of choking? Can they complete this activity in a timely manner?
Do they need prompting to eat?

Activity 3: Managing therapy and monitoring a health condition

Moderate to severe joint destruction may affect the ability to manipulate medication packaging, the handling of medications and importantly, may affect the ability to inject the clotting factor.

They may also report motivational restrictions because of their mental health secondary to their condition which must be adequately explored.

Remember in PIP…

Therapy is only considered within the scope of the activity if received in the home environment. If they do have therapy at home, do they require assistance? If so, why and how long does this take?
Regarding their medication, can they manage this themselves? Can they administer their own medication? Can they remember to take it and physically remove it from packaging? Do they need prompting?

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 4: Washing and Bathing

Some people with joint bleeds may struggle to get in and out of an unadapted bath and/or shower and wash their body due to joint pain and limited movement. They may also be impacted by fatigue.

You must consider the limited range of movement they may have secondary to their condition.

Furthermore, we must consider the psychological impact of their condition and how this may impact their motivation to complete this activity.

Remember in PIP…

You must explore how someone is managing the activity. How do they get in and out of the bath? Do they have any aids? Can they wash their entire body? How long does it take and how do they feel after? Does this activity increase their symptoms? How long does it take to recover? Do they need assistance with upper or lower body?

Activity 5: Managing toileting needs and incontinence

Bleeds leading to joint pain and stiffness may limit someone's ability to reliably sit and stand from the toilet as well as clean themselves after using the toilet.

Remember in PIP…

You must explore how someone gets onto and off the toilet and how they clean themselves? Does doing this make them more breathless? We do not consider ability to mobilise to the toilet. Also explore if someone has incontinence on majority of days and how they manage this.

Activity 6: Dressing and undressing

Joint bleeds may lead to difficulties related to joint pain, stiffness and limited movement which impacts someone's ability to reliably dress.

If balance problems are reported, find out what impact they have on ability with this activity area. How does the person manage? Is any help needed and if so, what does it involve?
Furthermore, we must consider the psychological impact of their condition and how this may impact their motivation to complete this activity.

Remember in PIP…

You must explore someone's ability to dress and undress within the scope of STAR. How long does it take them? Can they dress upper and lower body? Does this increase symptoms of fatigue or pain? Do they require assistance or use any aids? Can they dress reliably whilst seated?

Activity 12 Moving Around

Remember in PIP…

Moving around can be limited where there is joint destruction. The risk of spontaneous bleeds is increased in severe disease, even doing everyday movements. Use of mobility scooters or wheelchairs may be reported (although remember that if shoulder, elbow and wrist joints are affected by haemarthropathy, this may not be possible).

Continued mobility problems may be reported after joint replacement, this is because unlike in ordinary arthritis, multiple joints are affected. There may also be soft tissue damage and flexion deformity from bleeds into muscles around joints as well as bleeding into joints.

You must explore STAR when asking about someone’s ability to mobilise. It is not sufficient to simply indicate a distance or time someone can walk for.

Firstly establish when someone is walking, do they require an aid? If so, what aid? Was this prescribed or self purchased?

How far can they walk and how long does this take? How does walking make them feel? Does pain/fatigue increase each time they walk? When they walk a second time, does pace reduce or distance reduce? If they experience fatigue, how do they feel later in the day? How many times could they repeat the distance they walk? How long does it take to recover from mobilising?

Lived examples are also extremely beneficial. How do they manage up and down the stairs? How do they manage with chores? How do they manage their shopping? How many aisles can they walk and how long does it take? If they go to their GP surgery or hospital, how far is it from the car? How long does it take them to get to the required department and how far is this?

Functional Impact

If there is evidence of the condition causing a detrimental impact on their mental health, you must focus on exploring the below adequately

Activity 9: Engaging with others face to face

The psychological impact of the condition must be considered across relevant activities. There may be some anxiety or low mood secondary to their condition which could impact their ability to reliably engage with others.

Remember in PIP…

If they do report restrictions engaging, who can they engage with on the majority of days? How do they engage with unfamiliar people? If they report support is required to engage, who can provide this and why?

Activity 11: Planning and Following Journeys

The psychological impact of the condition must be considered across relevant activities. There may be some anxiety secondary to their condition which could impact their ability to reliably go out of the home without support.

They may also suffer from co-existing conditions that impact their ability to plan and follow journeys.

Remember in PIP…

Physical restrictions are not considered within the scope of the activity. Consider someone's ability to plan and follow a journey in relation to potential sensory or cognitive issues or the presence of OPD. How do they manage both familiar and unfamiliar journeys? Can they plan a journey? Could they manage a diversion? If anxiety is reported, does this meet the threshold of OPD? If so, could they leave the home on the majority of days with support?

Functional Impact

Although the condition may not directly impact the below activities, you must consider any co-morbidities that may cause functional restriction:

Activity 7: Communicating verbally

Remember in PIP…
Can someone express and understand both basic and complex verbal information? Who do they speak to? Can they use a mobile phone? Do they have any cognitive or sensory impairments that may impact their ability to complete this activity?

Activity 8: Reading and understanding signs and symbols

Remember in PIP…
Can someone read and understand basic and complex written information? What do they read? Can they read a text message or a book? Did they gain any qualifications in education?

Activity 10: Managing budgeting decisions

Remember in PIP...

You must explore someone's ability to manage both basic and complex budgeting. This would include how they manage their household budgets. How do they plan for future purchases? Would they understand change to expect in a small transaction? Is there area impacted by low mood or any cognitive impairments?

Additional reading or other resources

EXTERNAL

NHS Choices Haemophilia – symptoms

NHS Choices-Haemophilia – treatmentshttps

The Haemophilia Society-Understanding haemophilia

Guidelines for the management of acute joint bleeds and chronic synovitis in haemophilia-

Specialist Psychological Support Campaign | The Haemophilia Society

Psychological support provided by the Inquiry | Infected Blood Inquiry

England Infected Blood Support Scheme | NHSBSA

INTERNAL

Haemophilia-Haemarthropathy-and-PIP-Assessments