Condition Insight Report (CIR)

Ehlers-Danlos syndrome

Version 1.0

Made in collaboration with Ehlers Danlos support UK

Overview

What is the condition usually called/ any abbreviations used?

Ehlers-Danlos syndrome (EDS)

Can also be known as:

- **Hypermobile Ehlers-Danlos syndrome (hEDS)**
- **Benign joint hypermobility syndrome**
- **Joint hypermobility syndrome (JHS)**
- **Ehlers-Danlos syndrome – hypermobility type**
- **Ehlers-Danlos syndrome type 3**

Brief overview of the condition

Ehlers-Danlos syndrome is a genetic connective tissue disorder which affects the collagen within the body. Collagen is a protein which is the main building block of the body providing strength and support in ligaments, tendons and cartilage and as we are made up of such a high percentage of collagen, the condition can impact the whole body.

There are thirteen different types of EDS, with hypermobility being the most common and the others being; classical, vascular, kyphoscoliotic, arthrochalasia, dermatoparaxis and tenascin X.

Presenting Symptoms

As collagen is present throughout the body, people with EDS tend to experience a broad range of symptoms, most of them less visible than the skin and joint differences.

These are complex syndromes affecting many systems of the body at once, despite this EDS is often an invisible disability.

Symptoms commonly include, but are not limited to,

- Long-term pain
- Chronic fatigue
- Joint problems (including frequent dislocations and weakness)
- Dizziness
- Palpitations
- Bowel & Bladder problems
- Digestive issues (such as heartburn, nausea and vomiting, constipation and/or diarrhoea)
- Psychological disorders (such as depression and anxiety)
- Hypermobility
- Problems with internal organs, such as mitral valve prolapse (in the heart) or organ prolapse

Such problems and their severity vary considerably from person to person, even in the same type of EDS and within the same family.

Hypermobility spectrum disorder (HSD) is a closely linked condition with the same management and joint issues/chronic pain suffered by those with EDS.

Common Treatments

Medication

- **Analgesics:** Over-the-counter or other stronger prescription pain relieving medications may be prescribed for muscle or joint pain.
- **Nonsteroidal anti-inflammatory drugs (NSAIDs):** Relieves pain, decreases inflammation, and reduces fever.
- **Anti-hypotensive agents:** Medicines to keep your blood pressure a little low would be prescribed, as a precautionary measure to prevent damage to blood vessels.

Therapy

- **Physical therapy**: Exercises may be recommended to strengthen muscles around the joints to prevent dislocation.
- **Pacing therapies**: Often utilised to teach the person how to preserve and manage their energy levels.

REMEMBER: The absence of any of the above, does not necessarily mean the absence of a restriction. Each claimant should be assessed individually.

Watch a video HERE about the spoon theory which aims to explain the impact of fatigue

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?
Postural tachycardia can be common with people suffering from EDS. This may mean postural changes cause a loss of consciousness secondary to increase in heart rate, and consequently impact someone's ability to complete activities safely.	If someone reports fatigue, their ability to complete activities in a timely manner may be impacted. How long is it taken someone to cook, wash or dress? Is it more than double the time taken by someone without a disabling condition?	When someone has a longstanding condition, they may have adapted to their restrictions in day to day life to a certain extent. It is your role to explore if they are completing activities to an acceptable standard within the scope of the PIPAG.	Fatigue can be a common symptom of EDS and make it difficult for people to repeat activities to an acceptable standard. If someone reports they can complete an activity, you must explore how long it takes and how they feel after. Could they repeat it as many times as is to be reasonably expected throughout the day?

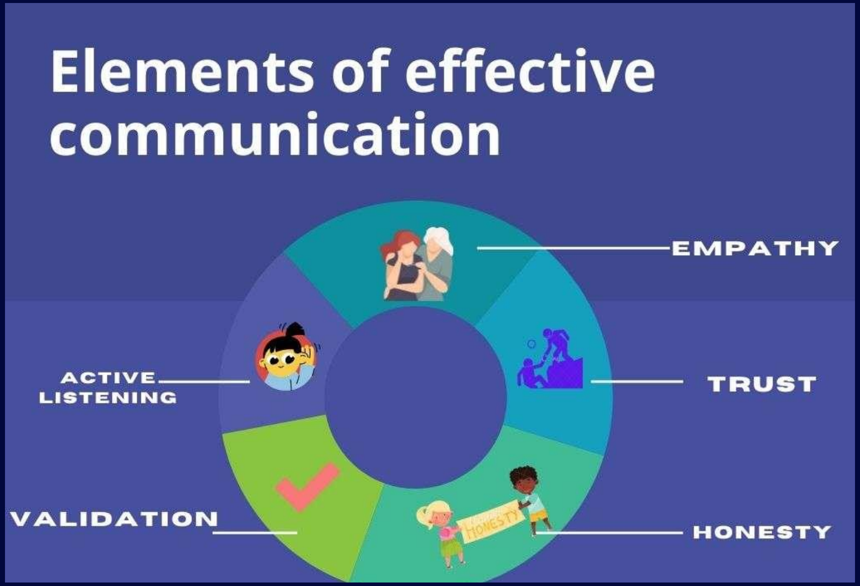
Sensitivities & Customer Care

- Be aware that EDS is an **invisible condition-**Individuals often look well and because they are hypermobile are deemed to have good movement when this is not the case as it can cause dislocations and pain.
- People may look well but are suffering from pain and other symptoms.
- Appreciate the **fluctuating nature** of the condition. The day of the assessment may be a good day, or the energy used to complete the assessment may have a detrimental impact on the claimant later in the day.
- **Physical examinations regarding movement are not accurate or appropriate** due to hypermobility being a primary symptom of the condition.
- They may find it difficult to talk about sensitive and personal topics such as **bowel problems,** among others.
- They may also fail to mention their **psychological suffering** to avoid being labelled as mentally ill when their condition is rooted in physical issues. People with EDS are often labelled by medical professionals as having a mental health condition rather than a physical condition due to the lack of knowledge about EDS and how it manifests and is diagnosed.

Empathy
It is imperative that the people you are assessing feel heard and listened to.

Trust
Trust can be obtained by ensuring you are open with the claimant and have adequately prepared for your assessment to be able to ask relevant questions throughout.

Active listening
Repeating back what has been reported at appropriate intervals and ensuring you don't ask the same questions multiple times shows active listening.



Validation
Validation can be verbal or nonverbal. It helps people feel heard and respected. Assessments can often be stress inducing for claimant's and such validation is important to ensure they feel comfortable to continue.

Honesty
You must ensure your assessment report is an accurate representation of what the claimant has reported. Although not a verbatim report, advice to the Department must be clear and concise.

Functional Impact

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

Activity 1: Preparing food

People who have EDS may experience, pain, fatigue and joint instability leading to dislocations. This may all impact their ability to prepare a meal to an acceptable standard.

Remember in PIP…

We must explore **how** someone cooks. Can they stand for the duration of cooking? How long does it take and how do they feel after? Do they require assistance? Can they safely peel and chop vegetables? If they experience fatigue, how long does this take to pass? Can restriction be overcome with aids, or do they require assistance? Do they have pain or joint dislocation? Also, if they experience PoTS, how is this managed?

Activity 2: Taking nutrition

If someone experiences digestive problems secondary to their condition such as nausea, vomiting or heartburn, this may impact their ability to take nutrition.

Joint pain may also impact their hands and consequently their ability to cut food and bring it to their mouth.

Remember in PIP…

Dietary restrictions are not considered within the scope of the activity. However, you must explore if someone needs prompting to eat. If so, is this prompting throughout the duration of the meal? Also, can they physically cut food, bring it to their mouth and swallow?

Activity 3: Managing therapy and monitoring a health condition

People who have a diagnosis of EDS may be having therapy such as physiotherapy or pacing therapies. They may also be prescribed multiple medications to help with joint swelling or pain.

Remember in PIP…

Duration of therapy is not considered. However, any assistance that is required within the home environment, if clinically consistent, must be considered and probing take place around times.
Also, can they manage their own medication? Can they remember to take it and physically remove it from packaging?

Functional Impact

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Activity 4: Washing and Bathing

People who have EDS may experience, pain, fatigue and joint instability leading to dislocations. This may all impact their ability to wash and bathe to an acceptable standard.

Also consider if someone has PoTS if they can safely wash and bathe. Is there any risk of loss of consciousness or falls?

Remember in PIP…

Can they wash and bathe majority of days? Can they reach all areas of the body? How long does it take them? Do they sit down, if so, why? Can they get in and out of an un-adapted bath? Can they overcome restrictions with aids, or do they require assistance? Do they experience pain or increase fatigue after the activity?
Also, if they experience PoTS, how is this managed?

Activity 5: Managing toileting needs and incontinence

You must consider if joint pain impacts their ability to it and stand from the toilet and reliably clean themselves. Do they have dislocations doing this activity?

Also, if they experience digestive issues causing diarrhoea, does this lead to incontinence?

Remember in PIP…

Mobilising to the toilet is not considered within the scope of the activity. Do they have incontinence? Can they manage this themselves? Can they sit and stand from the toilet and clean themselves after? Do they have doing this? Do they utilise any aids?

Activity 6: Dressing and undressing

People who have EDS may experience, pain, fatigue and joint instability leading to dislocations. This may all impact their ability to dress and undress to an acceptable standard.

Remember in PIP…

Sitting down is considered acceptable within the scope of the activity. Does this reduce their fatigue? How long does this activity take? How do they feel after? Can they dress their upper and lower body? Do they have grip, can they bend? Do they experience pain?

Functional Impact

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

Activity 7: Communicating Verbally

Although EDS itself is unlikely to cause a functional restriction in this area, please consider any co-existing conditions that may impact their ability to communicate.

Remember in PIP…

A claimant must be able to both express **and** understand verbal information. Retention is not considered, however, they must be able to comprehend what is being said to them. Do they communicate on the phone? Do they speak to friends and/or colleagues?

Activity 8: Reading and understanding signs and symbols

Although EDS itself is unlikely to cause a functional restriction in this area, please consider any co-existing conditions that may impact their ability to read.

Remember in PIP…

Within the scope of the activity complex written information is considered more than one sentence. Can they understand what they are reading? Do they use any aids?

Activity 9: Engaging with others face to face

Someone who has a diagnosis of EDS may have psychological symptoms secondary to their condition. This may include depression and anxiety and consequently impact their ability to want to leave the home or reliably engage with others.

Remember in PIP…

Prompting can be considered as social support. Does the claimant have other conditions that may impact their ability to engage? If so, who can they engage with? How do they feel engaging with unfamiliar people? Who can support them to engage? If someone specific, why is this?

Functional Impact

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

Activity 10: Budgeting

Although EDS itself is unlikely to cause a functional restriction in this area, please consider any co-existing conditions that may impact their ability to make budgeting decisions.

Remember in PIP…

A person needs to be able to make simple and complex budgeting decisions. How do they manage their bills? Can they plan for a future payment? Can they manage their own food shopping?

Activity 11: Planning and following a journey

Although EDS itself is unlikely to cause a functional restriction in this area, please consider any co-existing conditions that may 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?

Activity 12: Moving Around

Pain, fatigue, hypermobility and dizziness are all common symptoms of Ehlers Danlos syndrome and are likely to impact someone's ability to reliably moving around.

Remember in PIP…

STAR is imperative, especially with conditions that may cause fatigue. How far can they walk? How long does this take? What pace do they walk at? Can they repeat this? Is this at the same or a reduced pace? How many times could they repeat it? How long does it take to recover? Do they need an aid?

Additional reading or other resources

EXTERNAL

The Ehlers-Danlos syndromes (EDS) GP Toolkit

What is EDS? – The Ehlers-Danlos Support UK

EDS-Family-Friends-A4-Poster.pdf (ehlers-danlos.org)

Spoon Theory – YouTube

The Ehlers-Danlos Support UK – Support for people touched by the Ehlers-Danlos syndromes

INTERNAL

Desktop-Aid-Activity-12

Desktop-Aid-Fatigue

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