Capita

Condition Insight Report (CIR)

Primary Immune Deficiency (PID)

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Overview

Brief overview of the condition

People living with a Primary Immune Deficiency (PID) have conditions which impact on their ability to fight infection. All PIDs are counted as rare diseases and are covered under The Equality Act as a disability. PIDs are a lifelong condition and can be life-threatening. **It can take an average of 7 years to receive a diagnosis.**

What are the general term(s) used to describe the condition?

Primary Immune deficiency (PID), Primary Immunodeficiency or Immuno-dysregulation, or Primary Antibody Deficiency (PAD).

What is the condition usually called / any abbreviations used?

There are over 400 different types of PID. Some of the most common are:

-Common Variable Immune Deficiency (CVID)
-Selective IgA deficiency
-X-linked agammaglobulinemia (XLA)
-Hyper IgM syndromes
-IgG subclass deficiencies
-Transient hypogammaglobulinemia

and the well defined syndromes:

-Hyper-IgE syndromes
-Wiskott-Aldrich Syndrome

Presenting Symptoms

PIDs differ in the manner and severity with which they disable the function of the immune system. Different conditions have different impacts on peoples' daily lives because of the fluctuation in symptoms, any current infections, and the variety of treatments received. Some of the symptoms may include but are not limited to:

- Frequent bacterial, viral, fungal or parasitic infections, typically of the upper and lower respiratory tract. Infections *can* occur anywhere, as the immune system works throughout the body. Any infection requires robust and prompt treatments with high dose therapies or may otherwise become intransigent or life-threatening e.g. norovirus
- Pain (both muscle and joint)
- Fatigue (closer to exhaustion)
- Breathing problems like Bronchiectasis
- Inflammation and / or damage of internal organs (such as bronchiectasis or GLILD)
- Blood disorders (low platelets; low white cell counts; low lymphocyte counts)
- Digestive problems – diarrhoea and loose bowels are very common including irritable bowel syndrome (IBS),
- Associated autoimmune disorders e.g. type 1 diabetes; coeliac disease; lupus, rheumatoid arthritis, thyroid disease, adrenal failure, pernicious anaemia or Granulomatous Lymphocytic Interstitial Lung Disease (GLILD)
- Fibromyalgia – most people living with immunodeficiency have this, but do not necessarily get a formal diagnosis
- Allergies
- Anxiety relating to contracting infections or engaging with others who may be carriers and can lead to depression

PID patients do not "look ill" usually until they are in need of emergency care, and may be dismissed by medics because of this. The typical signs and symptoms of infections are created by a functioning immune system. Some patients will not cough since the rigorous physiotherapy they self-administer is aimed to prevent coughing. A chesty cough or dry cough is simply a symptom of another underlying condition, such as a secondary infection.

Many PID patients live with co-morbidities, often due to late diagnosis of their PID condition and this impacts significantly on their daily lives. Treatment for co-morbidities is a very complex balance of medications and treatments all of which can have significant side effects.

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?
For those having home infusions there are considerations around the hygiene implications to reduce bacterial, viral and fungal load and safe disposal of sharps. Some medications also require additional temperature regulated storage. Those with associated mental health conditions or those with cognitive restrictions may require support.	Pain and fatigue are significant (often debilitating and exhausting) and can increase with any physical activity or prolonged activity. People may have impaired cognitive function. They will experience difficulty with concentration, or in processing information, which will become more profound, as will their short-term memory. Tasks which require exertion physically and even mentally are likely to take longer, so it is critical this is fully explored.	Many immune deficient claimants have adapted to managing their own conditions (as a necessity since the condition is rare). They will establish their own adaptations in order to carry out simple daily activities. Understanding how they have adapted is critical and must be explored by the assessor to draw out the key points especially if not raised by the individual.	Claimants may not be aware they have an infection or virus, due to not getting the same indicators as would be expected which can result in very quick change from being apparently 'healthy' to needing emergency care. People living with a PID experience limited energy levels and must decide how they will use their energy. Attending a hospital appointment or any social event can result in two or three days at home in bed to recuperate.

Pertinent areas to explore regarding PID

How has the condition been over the past 12 months? What changes have they seen, over what period of time did they experience any deterioration, what has this meant for their functional ability now compared to previous?

For any impaired cognitive function: How consistent is their presentation, how do they manage their memory deterioration, are they orientated, can they plan tasks as they used to?

What is their level of pain/fatigue? Can they quantify this, do they use a scale and can they describe it? Can they share what a ‘typical’ day might be for them? What are their triggers? Whilst exertion is a main trigger, what amount of exertion is enough to cause further limitation

Where regular infections are reported: How often? How long do they last? How are they managed? Any rescue medication and/or therapy interventions including district nurse input during this time? Does treatment take place at home or elsewhere?

Where breathing difficulties are reported: what is the difference between symptoms at rest and what occurs during exertion. What level of exertion causes a change, recovery period and impact this has? Any recent lung function results?

Facts About Treatment

Treatments for PIDs range from **bone marrow transplant** for the most severe types, to **prophylactic antibiotic therapy**, with swift implementation of rescue antibiotics for those with the least severe PID.

In between these two extremes are the majority of PID applicants who are treated with **regular infusions** of replacement immunoglobulin therapy (replacement antibody infusions derived from plasma donations), together with **prophylactic antibiotics** and **additional backup antibiotics** for any breakthrough infections.

NOTE:PID patients do not become immuno-competent through replacement immunoglobulin therapy. This therapy provides a baseline level of background immunoglobulin (antibodies). The patient remains more vulnerable than their immunocompetent peers because they have no (or limited) ability to produce their own antibodies to fight bacterial, viral, fungal or parasitic infections, which can become intransigent.

Once their baseline infused dose of immunoglobulin is used up fighting an infection, or just coping with day to day life, the patient has nothing to fall back on to assist in recovery. PID patients catching a cold frequently go on to develop serious, secondary, bacterial infections of the upper respiratory tract, requiring additional antibiotic treatment. Many PID patients live with frequent, severe and / or intransigent infections, which impact on all areas of functional ability.

Sensitivities

What areas might they find difficult to mention or perhaps understate the impact of?

- **Fear of contracting an infection every day**.

This fear must not be underestimated. This can manifest as anxiety when encountering individuals carrying infection.

- **Extent of their illness**

Those with a PID do not look ill - their immune systems do not respond with the usual signs and symptoms of someone who is unwell. They may say they are 'fine' and completely underestimate the extent of their condition / symptoms.

- **Extent and impact of fatigue/exhaustion**

It is important to explore how they feel before, during and post a task to get a clear indication of impact.

- **Level of support provided by friends/family**

Please cross reference with any companions to add anything additional should they feel it is important to.

Watch Shelby talk about her experience of living with Specific Antibody Deficiency and hypogammaglobulinaemia HERE.

Customer Care

How is it best to ask about any sensitive topics and what are the common courtesies?

In general

- Offer a break, should they need one, to reduce fatigue
- Present verbal information slowly and clearly if the claimant appears to be having problems following
- Only present one question at a time. Repeat any questions if needed in a non-patronising manner
- Offer different ways to phrase a question. Be engaging by using open gestures. Images may be useful. Allow claimants to answer in written format if this is easier for them.
- Confirm that you have read through the persons background notes and further reading including the training outlined and signed off by DWP *(found in the resource section)*
- Verbal recognition of how difficult it must be to live with a PID and the fact that it is a hidden, rare, disability that nobody fully understands. Recognise this is an extremely rare disease and many professionals are unaware of it
- Ask the claimant to correct you if you say anything that indicates mis-understanding. Take corrections with grace.
- Practise active reflective listening. If someone explains an issue repeat your understanding back to them
- Explain the assessment process and what is required of the individual, including how long it may take and where the information will be going and why

During face to face interactions

- Always ensure scrupulous hand hygiene and do not expect to shake hands
- Ensure you have sanitising hand gel with you and use it before you introduce yourself. If this is not available, offer to wash your hands before starting the assessment
- Display social distancing – the claimant and their family will have had to practice this long before the coronavirus
- Confirm to the claimant that you do not have any current infections or virus’ and have not been in contact with anyone with a current infection or virus
- Ask where it is appropriate to sit – claimants may operate a ‘visitors chair’ system so that they maintain their own ‘clean’ chair environment
- Try to make any reasonable adjustments to noise, temperature and lighting if you are able – explore this with them prior to the assessment
- A home visit is preferable for a claimant who will be genuinely concerned about the risk of infection when travelling to and from or being inside an assessment centre. Do not just appear at the door - ensure the claimant is aware of your visit (and the purpose) well in advance
- Keep as much eye contact as possible allowing an individual to feel listened to and engaged. Be friendly and professional. The claimant may well feel threatened and misunderstood
- Do not try to sympathise E.g. ‘I know what you mean, my memory is terrible’. You have no idea what it is like to live with a PID

Functional Impact

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

Activity 1: Preparing food

Pain, fatigue, breathing restrictions and associated autoimmune disorders such as rheumatoid arthritis can impact on all aspects of preparing a meal. Some people may have difficulty with preparation, cutting items and other cooking elements. Many may need support due to burning items or getting distracted whilst cooking due to the fatigue experienced.

Remember in PIP…

We must explore STAR. How do they physically manage this task? Does it increase fatigue? How long does it take? How do they feel after? How long can they stand to cook? Can they lift pans and chop vegetables? Are they safe in the kitchen?

Activity 2: Taking nutrition

Some people with PID may have problems eating; finding eating too exhausting, and suffer from nausea.

Some might miss meals because they feel too unwell and have lost weight as a result.

Remember in PIP…

You need to establish if they are reliably taking nutrition. Firstly, can they physically cut food, bring it to their mouth and swallow, and secondly are they eating regularly. Do they require prompting to eat? If so, is this throughout the duration of the meal? Is there evidence of weight loss? Are there any recent incidents of choking?

Activity 3: Managing therapy and monitoring a health condition

Some people with a diagnosis of PID will be having Infusions delivered at home PID patients will also require an infusion buddy to be with them during infusions in case of anaphylaxis. Immunoglobulin therapies can vary substantially in duration.

Many people with PID, require physical therapy (lung clearance). Therapies such as nebulisation and chest clearance is considerable, typically requires a minimum of 30 minutes twice daily for either procedure.

Depression secondary to PID can cause non-compliance of therapies and other complex care plans.

Many claimants suffer with breathing difficulties requiring additional medications, nebulisers or both.

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

Claimants may find this task challenging and need to rest afterwards. Many will have days where they use wipes and dry shampoo for basic hygiene. Muscle pain e.g. from fibromyalgia, may lead to difficulty washing and combing hair due to limitation in raising arms or in some cases there may be no restriction at all.

Episodes of tiredness can come on suddenly. Levels of tiredness can also be highly variable. Some may use aids or have reduced the number of times they bathe.

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?

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Activity 5: Managing toileting needs and incontinence

High dose antibiotic therapy causes loose bowels and many have IBS so toileting can be a concern and focus for some and they may have resorted to using incontinence pads during times of bowel or bladder incontinence.

Some can be constipated and rely on laxatives due to some medication side effects.

Remember in PIP…

If the claimant is incontinent, how frequent is it and is this of one or both? Can this be managed with pads independently or do they need assistance to maintain hygiene? How do they transfer on/off the toilet?

Activity 6: Dressing and undressing

Muscle pain e.g. from fibromyalgia, may lead to limitation in raising arms or in some cases there may be no restriction at all. Episodes of tiredness can come on suddenly. Levels of tiredness can also be highly variable.

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

When communicating, some may find it increasingly difficult to communicate as they become more cognitively fatigued.

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

Consider any impact of their cognition changes.

Remember in PIP…

Lived examples are useful to provide a picture of function or restriction. What can they read on a daily basis? Can they understand what they have read? Do they use any aids or assistance to overcome their restriction?

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Activity 9: Engaging with others face to face

Avoidance of infection can mean voluntary self-isolation during cold, flu and norovirus season. The social isolation that can be caused by an inability to mix with friends and family can lead to depression. Those with associated mental health conditions may find it difficult to engage socially and either avoid them or rely on others for support.

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?

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Functional Impact

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

Activity 10: Budgeting

Claimants often report what is termed ‘brain fog’ - short-term memory issues, lack of concentration or mental clarity, or the inability to focus on a task.

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

Claimants often report what is termed ‘brain fog’ - short-term memory issues, lack of concentration or mental clarity, or the inability to focus on a task. Anxiety to go out due to the fear of being made ill by coming into contact with a virus or bacteria can make an individual very tense and cause difficulty completing a journey. In some cases it can cause such overwhelming anxiety that it can cause panic attacks and for them to avoid journeys all together.

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

Walking (both indoors and outdoors). Most immunodeficiency claimants will find walking and climbing stairs a challenge, either due to breathing problems or from exhaustion. Muscle and joint pain may also contribute to walking limitations which can be exacerbated by stairs and sloping ground.

Remember in PIP...

STAR is imperative, especially with conditions that may cause fatigue and breathlessness. 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

https://primaryimmune.org/about-primary-immunodeficiencies/specific-disease-types

PDF PID Awareness Distance Learning

PDF What is PID

PDF What is PAD

PDF What is CVID

PDF Why do I still get Infections

PDF The Spoons Theory

INTERNAL

- Desktop Aid – Fatigue, Activity 12, Activity 6
- CPD on Fibromyalgia and Chronic Fatigue Syndrome

Version control

Version	Date	Signed off by	Comments
1.0	20.10.20	Dr Shah Faisal	New document
1.1	29.03.21	Rebecca Noble	Links to ukpips.org.uk factsheets added to replace embedded documents
1.2	06/07/2023	Rebecca Sparks	Page added around pertinent areas to explore regarding PID and facts about treatment. Amendments also made to what questions to ask to establish functional restriction secondary to their condition.