

Conclusion

Being told that a long-standing diagnosis may no longer be correct and that your treatment may be changed or stopped, is very difficult to cope with. However, UKPIPS will continue to support you for as long as you need that support, even if you have been told that you no longer have a diagnosis of PID.

References

For references please contact UKPIPS

More Information

For further information about this leaflet or any PID issue, contact UKPIPS at:

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REVIEW OF CVID DIAGNOSIS

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UK
Primary
Immune-deficiency
Patient Support

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You have been told by your Consultant Immunologist that your diagnosis of CVID is being reviewed and this has probably left you feeling confused and bewildered. The purpose of this leaflet is to try to explain why this is happening, what implications it may have for your treatment and where to go to get more information and support.

Background Information

Clinical Immunology is a relatively young discipline. The first patient ever to be diagnosed with a Primary Immune Deficiency was a young boy in the 1950's. He had a very bright doctor caring for him, called Dr. Bruton, who realised that something was wrong when his patient kept getting ill despite the use of, what was then, the new treatment of antibiotics. Since this first diagnosis of a Primary Immune Deficiency (PID) many different types of Primary Immune Deficiency have been diagnosed and currently there are over 300 different types of PID. In the 1990's, Consultant Immunologists were diagnosing more and more adults with a late onset PID which affected the patient's ability to produce antibodies. At this time they called this disease Hypogammaglobulinaemia (HGG), but gradually they began to be able to divide patients with HGG into smaller groups, some having specific antibody deficiency, others having IgG subclass deficiencies and yet more, having deficiency in at least two types of Immunoglobulin (Ig's) - but always having a deficiency in IgG. They decided to call this group of conditions Common Variable Immune Deficiency (CVID) and created guidelines explaining to doctors how to diagnose a patient as having CVID. Being diagnosed with having CVID usually resulted in the patient being offered replacement immunoglobulin therapy, which replaced the absent or low levels of the patient's own IgG. Since then, more and more has been learnt about PIDs and how they need to be treated. The criteria

used for the diagnosis of CVID have been refined over the years, within increasing medical and scientific knowledge and the latest consensus is published by an organisation called the European Society for Immunodeficiency (<https://esid.org/Education/Diagnostic-Criteria-PID>). In patients who had CVID diagnosed many years ago and who are on replacement IgG therapy, it may not be possible to repeat all the tests currently used to diagnose CVID, without temporarily discontinuing Ig replacement therapy.

What will happen next?

Your Clinical Immunology Team will explain to you what tests they will be doing to try to discover how your immune system is working (or not). To do this, it may be necessary to take you off replacement immunoglobulin therapy. This is because the clinicians need to see whether your body is able to make adequate amounts of IgG that works in the way it should in order to keep you well. It will take 8-12 weeks for all the donated immunoglobulins that you have been given to leave your body and until this time, doctors cannot assess whether or not your own immune system is now able to keep you well. Your Clinical Immunology Team will support you through this period and will still be available to discuss your medical care and treatment options with your GP. It may be that you will still need to have prophylactic antibiotics and to have rescue antibiotics at home. It is very important that if you do develop infections during this time that you send samples for analysis to a pathology laboratory. Your Clinical Immunology Team will work with your GP to ensure that a protocol is in place which describes what you and all your medical advisors need to do if you start to become unwell.

What can the patient and their carers do during the process of review of their CVID?

It is important that a detailed daily record of your health is kept by you. This should include taking your temperature twice a day, making a note of how you are feeling - both physically and mentally - and recording any symptoms you may experience. UKPIPS has a Symptoms Diary you can use to help you to do this, or you can just note down your symptoms in an ordinary diary. It is important you have sample pots and swabs at home and that you can get samples **quickly** to your GP or Clinical Immunology Team as soon as you start an infection.

Start a folder in which you keep copies of all of your hospital letters, pathology results, including blood tests, your symptoms diary and any other information about your health. UKPIPS suggests that you keep this in date order, with your symptoms diary and a copy of your protocol at the front.

It is probable that the changes to your treatment and the news that you may not have CVID, after many years of being told that this was your diagnosis, will cause you worry and make you anxious. It is important you seek support from your family, GP and from patient support charities in order that you get the help you need in adjusting to this new situation. If possible, take someone with you when you go to your next immunology out-patient appointment. Ask them to help you to remember what the doctor says to you. Take a list of questions with you and ask the Consultant Immunologist to answer all of them. If the clinic is very busy and there is not enough time for you to obtain detailed answers to all your questions, ask the Consultant to answer them in a letter or an email and leave a list of your questions with them. You may find it difficult to think about what questions to ask and if this is the case, you can contact UKPIPS for help.

How long will the review take?

The review will finish when your Consultant Immunologist is certain that they have had the results for results for all the tests they needed to do, in order to decide whether you have CVID or not. However, this may take some time as they must wait until the donated immunoglobulin is no longer present in your blood. Every patient is different and it's difficult to be precise. However, as has already been stated, this process usually takes between 8-12 weeks.

The next step

Once the Clinical Immunology Team have had a meeting to discuss all your test results, you will be asked to come for an out-patient's appointment. At this appointment your Consultant Immunologist will tell you what has been discovered and whether or not you have CVID. If you have been found to have another PID, this will be explained to you. If it has been discovered that you do not have a PID, you will be told this and the tests that were done which revealed this, will be explained to you. You can ask for copies of all the tests results to take home with you.

Once your diagnosis has been explained to you, the Consultant will then explain what your new treatment plan will be.

If it has been discovered that you do not have a PID, you will be discharged from the care of the Clinical Immunology Team.

What can I do if I disagree with what has been said?

If you disagree with the diagnosis or the treatment decisions that your clinical immunology team have made, then you have the right to ask for a second opinion. If you choose to do this, UKPIPS can offer you advice and information on how to ask for this.