How to locate your nearest Immunology Centre.

Type **http://www.ukpips.org.uk/find-an-immunologist/** into your web browser and follow the on-screen instructions.

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CARING FOR PRIMARY ANTIBODY DEFICIENCY (P.A.D.) PATIENTS IN THE COMMUNITY

A Leaflet for GPs

UK Primary Immune-deficiency Patient Support

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What are Primary Antibody Deficiencies (PADs)?

Primary Antibody Deficiencies are a group of rare conditions, some of which are caused by a genetic defect, but all resulting in an inability to produce functional antibodies. Primary Antibody Deficiencies range in their severity from mild to severe. The most common group of PADs is called Common Variable Immune Deficiency (CVID). There are approximately 5,000 people diagnosed with a PAD in the UK, but under-diagnosis means that these figures are probably not reflective of the true incidence. A patient suffering from any type of PAD will have difficulty in mounting a response to infection.

What is the Treatment for PADs?

The Clinical Immunologist with responsibility for your PAD patient will write to you to advise you about their specific treatment needs. However, these general guidelines may help to inform discussions with your PAD patient and their carers.

Immunoglobulin Replacement Therapy

Those with a serious PAD will need immunoglobulin replacement (IgG) therapy. This is usually administered via intravenous or sub-cutaneous routes. Many PAD patients, when established on IgG replacement therapy, choose to self-administer their IgG therapy in their own homes. They will have been given intensive training for this by the Clinical Immunology Staff at the Tertiary Centre. Many also choose to receive this treatment on immunology day units, or infusion units under the supervision of immunology nursing staff.

Prophylactic Antibiotic Therapy

PAD patients may require prophylactic antibiotics. These should be on the patient's repeat prescription to aid treatment compliance. These may be stopped during a breakthrough infection (although practice varies) when a full therapeutic course of antibiotics needs to be prescribed. If prophylactic antibiotics have been stopped, please resume them after the completion of the above.

Antibiotic Therapy

Breakthrough infections in this patient cohort may be difficult to treat and patients with any type of PAD will require high dose, timely and appropriate courses of oral antibiotics for all breakthrough infections. These must be prescribed for a course of between 10 and 14 days of high dose standard antibiotics. Decision about antibiotics should be made on the basis of results of previous microbiology or history of response to certain antibiotics. If unsure please discuss with an immunologist or your local microbiologist, who must be aware of PAD in your patient.

Pathology Screening

It is important that a sustained effort is made to identify the pathogens causing the infection/ infections and their sensitivity. This will aid treatment outcomes and promote antibiotic governance. PAD patients acquire pathogens which are similar to those encountered by patients with cystic fibrosis. The following tests should be requested depending on symptoms.

- Sputum**:** Culture and sensitivity (bearing in mind that low virulence organisms are often clinically relevant). Cough swabs may be sent for children or adults who cannot produce sputum.
- Throat Swabs**:** Viral PCR, immunocompromised panel culture and sensitivity.
- MSU**:** UTIs may rarely be related to PAD.
- Stool for C/S**:** Microscopy and viral PCR if indicated. Patients may have long standing norovirus or other enterovirus infection, giardia or bacterial infection such as campylobacter or salmonella.

Immunisation

- Live Vaccines**.** Patients with a Primary Antibody Deficiency **must not be given a live vaccine** without a discussion with the immunologist, since they are liable to develop vaccine-related infection or persistent carriage of vaccine strain. Close household contacts of PAD patient may be given most live vaccines such as the nasal 'flu vaccine, rotavirus vaccine, shingles vaccine. However, as they may transmit the virus to their family member with a PAD, the vaccinee and, where possible, family member and their immunologist should be informed, so that appropriate precautions can be taken. Precautions include hand washing/ delegating nappy changes for rotavirus vaccine and use of acyclovir prophylaxis if a post-vaccination rash appears after shingles vaccine. Live vaccines such as yellow fever vaccine and live/oral polio vaccine (no longer routinely available) should be avoided in contacts. Killed typhoid vaccinations should be used rather than the live oral vaccination. However, family members (but not immunodeficient patients) may receive MMR and BCG vaccines.
- Killed Vaccines**.** Whilst it is clear that PAD patients cannot generate effective long-lasting antibodies in response to vaccines, it is thought that they may be able to develop some cellular response. Some may also generate a partial or transient antibody response. Killed vaccines should be offered, particularly the annual flu vaccine.

What Treatment Protocols are required for PAD Patients in the Community?

- Protocols for self-administration of immunoglobulin therapy.
- Protocols for self-administration of I.V. antibiotic therapy.
- Protocols for investigation and adequate treatment of breakthrough infections.