



**West Midlands Immunology Patient Day
Sunday 29th September 2013
Crowne Plaza Hotel
NEC Birmingham**

Medical and scientific staff updated the audience on the changes in Immunology treatments and services, travelling and living with immunodeficiency and Immunology research in Birmingham.

Here we present a report on the topics we covered at our 2013 Immunology Patient Day.

Kindly sponsored by Grifols, BPL, Calea and Octapharma.

This report has been produced by [Primary Immunodeficiency UK](#) in collaboration with the [West Midlands Immunodeficiency Centre at Birmingham Heartlands Hospital](#).

Your immunology services



The Immunology team at Heartlands

Dr Aarn Huissoon and Dr Scott Hackett started the day describing the immunology services for patients in the West Midlands and the changes in service provision within NHS England through a new process called NHS Specialised Commissioning.

There are five immunology centres in the Birmingham region comprising Heartlands Hospital, City Hospital, Russells Hall, Stoke and services starting at the Queen Elizabeth Hospital. In total these services serve over 300 patients with primary immunodeficiency.

In contrast to previous years when decisions were made at a local level concerning funding and care provided, these services are now governed by national contracts under NHS England Specialised Commissioning. This took effect from the 1st October 2013. The aim is to improve equity of care and access to drugs for all patients within England. The NHS contract covering immunology services is set out by the Clinical Reference Group (CRG) Group B9 and can be found here www.england.nhs.uk/wp-content/uploads/2013/06/b09-spec-immun.pdf. Centres have to adhere to the standards set out in these contracts to receive funding.

Accreditation of centres and Birmingham Heartlands Hospital

An important and integral part to the contract is that the service provided by any centre is accredited. Accreditation involves a centre proving, through evidence and a site visit, that it reaches the standards set down by the professional body known as UK Primary Immunodeficiency Network (UK PIN).

The accreditation process concentrates on six main areas:

- Staffing levels required for immunodeficiency services
- Facilities required
- Organisation and administration of the service
- Quality of the clinical care provided
- Home therapy training where chosen by the patient and support for patients
- Audit, education and management of the service.

At present Birmingham Heartlands Hospital is one of six hospitals in England and the only hospital in the West Midlands to have gained accreditation.

'Never has the patient voice been more important'

Dr Huissoon stressed the important point that Immunology services need a patient voice. Local Area Teams want to engage with patients to ensure their voice is heard and their needs are met. They are keen to support the setting up of a local patient group for Heartland patients. The patient voice is also vital at a national level and for working within the Clinical Reference Groups that decide on the service contracts and policies determining what drugs are available to patients and under what circumstances.

What this means to you as patients

You should not see any immediate changes to your services as these changes are happening at a higher level. But what is certain is that the way healthcare is provided for is changing and will change more going forward. However, no one, including doctors and the commissioners themselves, has a firm understanding of what these changes might mean in the long-term.

Immunology research in Birmingham

Causes of immunodeficiency



Dr Scott Hackett talked about his collaborative work to get new genetic diagnoses in primary immunodeficiency based on measles vaccine susceptibility, severe skin and ear infections and severe chicken pox.

Research on Common Variable Immune Disorder (CVID)

Dr Gabriel Wong described his work on the causes of the most common primary immune deficiency known as Common Variable Immune Disorder, CVID. In CVID the immune system becomes unbalanced but what causes this is not known. Work funded by the Wellcome Trust is focusing on how different cell types with the immune system interact with each other in CVID. Currently more than 70 patients have been enrolled into the study. The ultimate aim of this study is to identify therapeutic targets that would tackle the non-infectious complications of CVID.

In collaboration with a major CVID centre in Freiburg, Germany, the Immunology team is looking at the outcome of interstitial lung disease (ILD) in CVID. Interstitial lung disease is the name for a large group of diseases that inflame or scar the lungs. The inflammation and scarring make it hard to get enough oxygen. The scarring is called pulmonary fibrosis. In many cases of CVID lung disease is driven by infection but in 20% of cases it is not. This multi-centre international research is focussed on identifying the non-infectious mechanisms that lead to ILD with the aim of gaining information that will lead to better treatments.



Clinical research and the UK PIN database

Dr Aarn Huissoon talked about clinical trials involving new products.

Dr Huissoon is also planning to enrol about five patients with Hereditary Angioedema (HAE) in a trial of a new treatment method to prevent attacks of swelling in this condition. Patients will need to stop taking conventional treatment to test this new product. The trial is likely to start in April 2014.

Birmingham Heartlands Hospital is also actively involved in the UK PIN database. This database contains the details of patients with primary immunodeficiency in the UK, and it is linked to a European (ESID) database. Its aim is to be a major source of information about the number of people affected by PIDs in the UK. Currently 2818 patients are registered. Patients have to consent to their data being entered and mechanisms are in place to ensure patient confidentiality.

The database gives information as to how many people are affected by different conditions, their outcomes, so aiding the development of health policy and as a major resource for research for patient benefit. The results from the UK PIN registry will be published soon. Dr Huissoon encouraged everyone with a PID to enrol on the database.

Gut problems in immunodeficiency



Gut problems are common in people with primary immunodeficiency and yet they are not well understood or studied. Dr Naveen Sharma, Gastroenterologist at Birmingham Heartlands Hospital, gave an update on this and his research.

The gastrointestinal (GI) tract is everything from the mouth to the bottom. Organs such as pancreas and liver and spleen are also involved.

Patients with CVID may present first at gut clinics so it is important that gastroenterologists know about immunodeficiencies. Gastro problems come second to chest problems in patients with CVID and current estimates indicate that 20-50% of people with CVID have gut problems. Classic symptoms are diarrhoea and loss weight. Because blood tests may not be helpful in patients with a weak immune system, endoscopy is used to investigate for gut diseases associated with CVID and other immunodeficiencies.

Patients with immunodeficiency may suffer more from gut infections including Giardia, but in some cases there is inflammation of the gut not related to infection. For this reason it's important to subtype the difference causes of gut problems in patients so clinicians can find better ways to treat it.

Sometimes liver problems also occur. This includes widening or dilation of the liver veins, or a condition called nodular regenerative hyperplasia. Sometimes enlargement of the spleen may occur. Patients need to be aware of these problems and their symptoms and report them to their healthcare teams.

How gastro problems are dealt with at Heartlands Hospital

The West Midlands Immunodeficiency Centre at Heartlands Hospital has an integrated combined clinic service involving the immunology health teams, a GI consultant and a dietician. These are held 3-4 times per year. This is one of only a few hospitals in the UK to run this joint clinic service.

Gut problems can really affect a person's quality of life so it's really important for patients to highlight any gut problems to their care teams.

Information on how you can access the clinic

If you think the combined GI/Immunology clinic might be able to help you, discuss this with your Immunology nurse or doctor. They will be able to arrange for you to have an appointment.



Your diet and immunodeficiency

Louise Dickie, Senior Dietician at Birmingham Heartlands Hospital gave a presentation on how getting help on your diet may help improve gut problems in immunodeficiency.

A range of gut problems can occur in CVID and other primary immunodeficiencies. These include inflammatory bowel disease (IBD). IBD includes ulcerative colitis, a condition affecting only the colon (large intestine) and Crohn's disease, which can affect the entire digestive system. Others problems can include coeliac disease where a person has an adverse reaction to gluten and irritable bowel syndrome.

What are the symptoms of gut problems you should look out for?

Symptoms of gut problems you should look out for are diarrhoea, nausea, vomiting, and bloating, extreme tiredness and unexplained weight loss. Low levels of iron, vitamin D and calcium can also occur. These symptoms are a result of malabsorption of the nutrients in food caused by changes in the structure of tiny finger-like projections in the intestines that are responsible for absorbing food. Characteristically these projections become shorter and flatter (like a flattened hairbrush) and less efficient at absorption.

Dietetic counselling can help

These conditions can be improved by changes to your diet. There isn't one specific diet that will work for everyone and what might work depends on the individual person and what their symptoms are.

A dietician will work through a range of options with you. These can include having a gluten free diet, an 'exclusion' diet working through exclusion of certain food groups, a low residue diet and changing the amount of fibre in the diet. Nutritional supplements may also be recommended e.g. iron supplements and calcium supplements.

Louise encouraged anyone affected by gut problems associated with their PID to inform their health team.

Plasma Therapies – from donor to patient



Dr Terry Woolley, of CSL Behring, gave a general overview of the production and safety processes involved in the purification of biotherapies, such as immunoglobulin, from plasma. Immunoglobulin can be used for replacement therapy in people with immunodeficiency. Dr Woolley explained why immunoglobulin therapy is important for patients with antibody deficiencies. Immunoglobulin binds harmful substances, neutralises bugs that cause infection and activates other immune cells so that they can do their work to help fight infection.

Producing immunoglobulin is a 6-7 month process with safety a high priority at every step. It is produced from either source plasma or plasma separated from whole blood donations from carefully selected donors.

CSL Plasma has a '4 Pillar integrated safety system' at CSL Behring. This covers:

- Selection of donors
- Manufacturing procedures

- Quality control
- Monitoring

Selection of donors

The careful selection of donors is key. Companies who produce immunoglobulin have to adhere to several regulatory requirements, in Europe this is regulated by the European Medicines Agency. In addition the quality systems are certified by the Plasma Protein Therapeutics Association (PPTA). All potential donors are screened before being allowed to donate plasma. This includes physical examination with vital signs temperature, blood pressure and pulse and completion of an in-depth questionnaire designed to assess health status, recent travel and risky behaviours. Each plasma donation is tested for presence of pathogens prior to its use in manufacturing. Only after donors have returned for a second donation and have again tested negative is the first donation released for processing. CSL Behring has 89 collection sites within the US and Europe with donors who donate regularly.

Manufacturing procedures

The screened donations are then pooled and segregated into batches. Virus inactivation and elimination procedures are integrated into the manufacturing process. This can involve acidic pH treatment to inactivate virus or 'nano' filtration to remove virus.

The manufacturing process ensures no batch-to-batch contamination so that if a batch were to become contaminated in this process its effects would not be wide spread. All the equipment used is scrupulously cleaned between each batch.

Donor centres and immunoglobulin manufacturers have very high standards of minimising the risk of infection getting into the immunoglobulin supply. Donor centres and manufacturers are inspected regularly and will be closed down if there is any hint of a problem.

Quality control

Each batch then undergoes extensive testing involving further checks for the presence of contamination, viruses and testing the quality and quantity of the immunoglobulin produced.

Monitoring

There are two phases of monitoring. One involves 'look back' and traceability so that each batch of immunoglobulin can be traced back from the donor to the patient it was used for and the second is monitoring of the patient who receive immunoglobulin by their doctors. For this your Immunologist will look out for any 'adverse' events when they treat patients with immunoglobulin. They will also either do annual hepatitis checks or save a sample of blood for infection testing. The make and batch of the immunoglobulin used and results of these tests are recorded in an Immunoglobulin dashboard. This is basically a database of information on immunoglobulin use in the UK. It is through this kind of surveillance that you can be confident that infections are not being spread by immunoglobulin.

Risk of infection

After Terry's presentation, the panel answered questions about infection risk and blood products. No one has ever caught HIV or Hepatitis B from immunoglobulins. In the 1990s, a small number of people caught hepatitis C from immunoglobulin. There have been no cases of infection being spread from person to person by immunoglobulin since the 1990s.

There are two theoretical risks from immunoglobulin. The first is from prion infection. Prions caused mad cow disease and then variant CJD in the 1990s, mainly in the UK. Prions have been spread from person to person by blood transfusions but never by immunoglobulin. But because of this theoretical risk, British plasma is still not used for making immunoglobulin.

It is not possible to predict whether new infections, which could be spread by immunoglobulin, will appear in the future. However, the immunoglobulin manufacturers and immunologists around the world are constantly on the look out for any problems such as this.

Some more frequently asked questions about immunoglobulin can be found on the PID UK website at www.piduk.org/whatarepids/treatment/immunoglobulinreplacementtherapy

Drugs in your backpack - travelling with immune deficiencies

Thinking of going holiday? 'All trips are possible. Discuss your plans with your consultant/nurse specialist a long-time before you travel', said Dr Scott Hackett.

His advice was to plan well ahead. Think about your immunoglobulin treatment and how long you plan to be away. It may be advisable to have a double dose of immunoglobulin before you go. Think about what vaccinations are needed and seek advice from your health team. Remember some may not be appropriate for you and in that case you may need written evidence of why you couldn't have them.

Don't forget about protection against malaria. This includes prophylaxis, use of bed nets and repellent sprays.

If you have chest problems you might need oxygen support while flying.

In case of infection whilst away it may be advisable to carry a dose of ciprofloxacin. Keep your medical supplies with you in the cabin. That way if your luggage is lost you still have the medicines you need.

It is also advised to carry a letter from your doctor to indicate what is wrong with you in case you fall ill. This should be translated into the language of the country you are visiting. Your health team can help with this.

While you are away use common sense to avoid infections through contaminated food and water.

Don't forget about travel insurance and remember it's essential to tell your insurance company about any pre-existing condition otherwise your policy will be invalid and you won't be covered.

You can find out what companies provide travel insurance for PIDs at the PID UK website www.piduk.org/whatarepids/management/travelling



'My trip to Borneo' by Tom Peel.

A highlight of the day was a talk from a patient, Tom Peel, who described how he was able to spend a in the Borneo jungle despite having an immunodeficiency.

The clear message was 'Don't let your condition stop you doing what you want to do. All things are possible.'

Tom spent six weeks in Borneo doing conservation work. In planning for his trip he

- Took advice from his health team
- Had a double dose of immunoglobulin

- Took strong antibiotics during his stay
- Took medicines to protect against malaria
- Carried a letter explaining his condition and why he hadn't had certain immunisations.
- Had medical insurance.

Also he said 'Don't forget if you are planning for a long trip you will need to arrange for treatment such as immunoglobulin therapy whilst you are away'.

Patient support groups: Midlands and National



*From left to right:
Susan Walsh from PID UK, Ann Price from HAE UK and Martin Parry and Margaret Bennett from the West Midlands Immunology Group.*

The West Midlands Immunology Group

Ending the day, Martin Parry and Margaret Bennett, patients at Birmingham Heartlands Hospital, talked about the launch of the new West Midlands Immunology Group (WMIG), a local support group for patients attending Immunology departments throughout the region.

Their website is at www.wmig.co.uk. Their aims are to establish a growing mutual support network to help patients locally and to link into national organisations for people affected by primary immunodeficiency.

Any one who would like to help is encouraged to get in touch: email contact info@wmig.co.uk.

National patient support groups



Susan Walsh,
Director of Primary Immunodeficiency (PID) UK spoke about the launch of this new organisation as a national support group.

Its aim is to be a powerful national voice for patients with primary immunodeficiency. She encouraged patients to visit the website www.piduk.org, comment on its content and to become members.

Ann Price of HAE (Hereditary Angioedema) UK spoke about the work of HAE UK to support all HAE patients in the UK. It is the leading source of information about HAE and provides advice and patient events to support those affected. For further information go to: www.haeuk.org/



This report has been produced by [Primary Immunodeficiency UK](http://www.piduk.org) in collaboration with the [West Midlands Immunodeficiency Centre at Birmingham Heartlands Hospital](http://www.birminghamheartlands.nhs.uk).