



Issue 8 2018

## Primary Immunodeficiency UK update

Dear PID UK member,

We hope you have had a wonderful summer and welcome back to our regular newsletters.

### Health Matters

The human papilloma virus (HPV) is a viral infection that is passed between people through skin-to-skin contact. In July 2018 the UK government announced that the HPV vaccine will also be given to 12 and 13 years old boys along with girls.

The HPV vaccine is effective at stopping girls from getting the type of HPV that causes most cervical cancers. Whilst boys do not have a cervix so cannot be affected by cervical cancer, HPV is linked to cancer of the anus, penis, mouth and throat so the introduction of the HPV vaccine for boys is a good thing.

Studies have shown that the vaccine protects against HPV infection for at least 10 years, although experts expect protection to last for much longer.

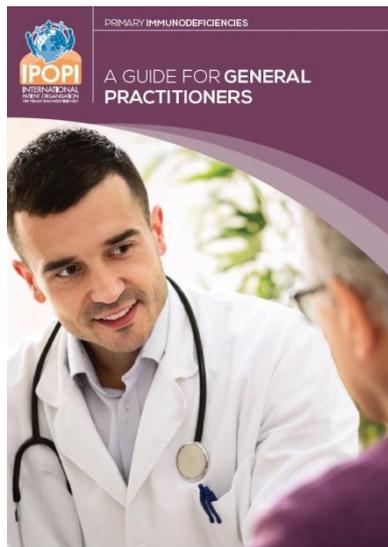
PID patients can receive the HPV vaccine. To find out more about the HPV vaccine visit the NHS [website](#)

### New genetic cause of PID identified



UK immunologists have pinpointed a new genetic cause of PID – *RIPK1* that causes severe immunodeficiency, arthritis and early onset inflammatory bowel disease. To date, four patients from three unrelated families have been found to carry mutations in RIPK1 (Receptor Interacting Serine/Threonine Kinase 1). Mutations result in altered working of a key cellular signal pathway and production of cell signalling molecules known as cytokines. The good news for those affected - RIPK1 is of medical interest as a drug target in other diseases and bone marrow transplant has been shown to resolve clinical problems. Read more at <http://science.sciencemag.org/content/early/2018/07/18/science.aar2641>

## GP booklet



A new booklet from IPOPI “A Guide For General Practitioners” is now available from PID UK.

The booklet explains how general practitioners can play a crucial role in identifying patients with primary immunodeficiencies and discusses key aspects of patient care.

If you would like a copy please email [hello@piduk.org](mailto:hello@piduk.org)

## Marian's story



In 2017, Marian our patient representative for the North – West joined the 100,000 Genomes Project.

She kindly let us interview her to find out more about why she signed up for the project and what she's hoping to gain from it.

You can read her interview [here](#)

## Life insurance

We've updated our website with [information on life insurance](#). We understand that this is a difficult area for PID patients. The content covers the questions you will be asked, declaring genetic testing and gives a list of companies who can be approached. We hope you find the information helpful.



## Update on the Prion Surveillance Study project

The [annual report](#) from the Prion Surveillance Study has been published. The Prion Surveillance Study has been running for over 10 years since it began in 2006. The study aims to find out whether there is evidence of the abnormal prion protein that is associated with variant Creutzfeldt-Jakob Disease (vCJD) in the blood and body tissues of primary immunodeficiency patients exposed to UK-sourced immunoglobulin between 1996 and 2000. vCJD is a very rare disease that causes

degeneration of nerve cells in the brain and spinal cord.

Approximately 175 primary immunodeficiency patients are thought to have been exposed to UK-sourced immunoglobulin between 1996 and 2000. Of these patients, 79 from 16 immunology centres across Britain have been involved in the study.

To date, no primary immunodeficiency patients have shown symptoms of prion disease, nor is there any evidence of prion infection in the tissues tested. These results are reassuring but it is very important that we continue to monitor patients over the long term because we know that prion disease may take many decades to develop after exposure.

## PID UK activities

Fiona Watt, one of our patient representatives for Scotland, took part in a NHS Scotland National Plasma Product Expert Advisory Group (NPPEAG) meeting. PID UK responded to a National Institute for Health and Care Excellence consultation on the evaluation of emapalumab in treating primary haemophagocytic lymphohistiocytosis (HLH). Susan, our Director, attended a meeting of the Public Health England oversight group on developing a pilot newborn screening programme for SCID.

## FAQs



We've updated all of our FAQs with queries we've recently received both online and over the phone.

Q. I'm worried that when my child transfers to adult services, I won't know what is going on and how to help/encourage him/her to take his/ her medication etc. [Can you advise?](#)

Q. I'm already under an Immunology team but would like to be as healthy as possible. Do you have any [tips to keep me well?](#)

Q. Do you have information on [travelling abroad with immunoglobulin?](#)

## Donations

Thank you so much for everyone who has donated to PID UK the past two months. We could not do the work we do without your generosity.

Thank you to the Price family who donated a portion of the entry fees of their open garden to PID UK. We are so grateful to you for wanting to support us.

## Fundraising

Want to fundraise for us? We'll help you any way we can whether it's with a sporting event or non-sporting. Email [events@piduk.org](mailto:events@piduk.org) if you are thinking of taking part in something.

Last month Jonathan (*pictured right*) took part in the Paisley 10k as injury forced him to cancel his participation in his previous race. Thank you so much for raising awareness and funds.



This month Chad (*pictured left*) took part in the Great North Run for PID UK as his son has XLA. We are so grateful for his effort and you can add to his fundraising [here](#).

Next month Victoria, her mum Claire and her sister, (*pictured right*), are all taking part in the Cardiff Half Marathon to raise money for PID UK and have raised an incredible amount already. If you want to add to their fundraising you can do here

<https://www.justgiving.com/fundraising/victoria-dyer>



### Changed your details?

If you've recently changed any of your details such as address or email please do let us know so we can update our database so we can stay in touch with you.



## Change of Address

With best wishes from,

Susan and Hannah  
**The PID UK Team**



Supported by an educational grant  
from Biotest (UK) Ltd

