



World Primary immunodeficiency Week: Let's talk about PI!

22 April 2011 - Today, leading immune deficiency societies from every region of the world joined forces with patients, nurses and allied health professionals to launch the first World PI Week. This inaugural week will be celebrated from 22-29 April culminating in the World Day of Immunology on 29 April.

In support of the week, a dedicated Website has been created (www.worldpiweek.org) to serve as a catalyst and tool to support local stakeholders advocacy efforts throughout the week to raise awareness about primary immunodeficiencies, drive the earliest possible diagnosis and ensure optimal treatment of this devastating group of diseases is available to every affected patient.

Primary Immunodeficiencies (PI) are hereditary and genetic defects in the immune system that cause increased susceptibility to a wide range of infections, affecting the skin, the ears, the lungs, the intestines and other parts of the body. These infections are often chronic, persistent, recurring, debilitating, and in some cases, fatal.

It is estimated that **10 million** people suffer worldwide¹. However, in Europe, Primary Immunodeficiencies are classified as rare diseases. Current diagnosis levels suggest that 1 in 8-10,000 people have a genetic primary immunodeficiency that significantly affects their health. However, experts estimate that between 70–90% of PI remain undiagnosed, and the true incidence of some PI within the general population could be as high as 1:250-500 [Prof. L. Notorangelo and Prof. L. Hammarstrom, STOA, European Parliament, March 2004]. These figures speaks for themselves and yet a simple and relatively inexpensive blood test CBC (Complete Blood Count) can identify over 95% of the patients! The problem remains one of lack of awareness leading to the majority of patients being diagnosed too late, with healthcare practitioners writing off these recurrent infections as 'just another cold'. "Increased awareness for primary immunodeficiencies can save many lives of infants who can live a normal life after appropriate therapy. Children with recurrent infections or with severe unusual infections should be checked for defects in their immune system" declared Prof. Amos Etzioni, founding partner of World PI Week.

A recent peer-reviewed study documenting the financial impact associated with early diagnosis and management of PI in the USA found that each undiagnosed patient with an underlying PI costs the healthcare system an average of \$102,552 (€75,587.40) annually, while after diagnosis and treatment costs to the healthcare system averaged \$22,610 annually; yielding an average savings of \$79,942 (€58,9223) **per patient per annum**². Sharing her experience of a late diagnosis, Jose Drabwell, patient and Chairman of IPOPI stressed: "if diagnosis had happened many years ago, then I would not have suffered such prolonged episodes of illness, hospitalisation, unnecessary operations, needless X-rays, scans and lots more. Within three months of diagnosis my life changed! I started a miracle treatment of immunoglobulin replacement and now the world is my oyster".

- **Detecting the disease early** can save lives and avoid unnecessary vaccinations that can cause serious illnesses and even death for undiagnosed PI patients.
- **Detecting the disease early** can save patients from lifelong disabilities like hearing loss or lung scarring.
- **Detecting the disease early** can significantly improve the patient's quality of life, by reducing the usually numerous visits to the doctor and stays in hospitals. If treated early, the patient can get back to school or work.
- **Detecting the disease early** has huge benefits for the patients, their family and society as a whole

¹ U.S. Department of Health and Human Services. National Institutes of Health. PAR-08-206: Investigations on Primary Immunodeficiency Diseases (R01). <http://grants.nih.gov/grants/guide/pa-files/PAR-08-206.html>. Retrieved on June 30, 2009.

² Modell, F., Puente, D., Modell, V. *From genotype to phenotype. Further studies measuring the impact of a Physician Education and Public Awareness Campaign on early diagnosis and management of Primary Immunodeficiencies.* Immunologic Research. 2009. 44(1-3):132-49



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During World PI Week (22-29 April) thousands of organisers across the globe will join forces and **take these diseases out of the shadow**, by encouraging governments to put in place targeted campaigns to increase recognition of PI amongst the medical profession, parents, schools, day care centres, and nurses.

The World PI Week Partners thank Baxter and CSL Behring for supporting this first World PI Week and hope to gain support from other companies to enable the initiative to continue and become self-sustaining.

NOTE

World PI Week Partners:

ASID, African Society for Immunodeficiencies

CIS, Clinical Immunology Society

EFIS, European Federation of Immunological Societies

ESID, European Society for Immunodeficiencies

INGID, International Nursing Group for Immunodeficiencies

IPOPI, International Patient Organisation for Primary Immunodeficiencies

JMF, Jeffrey Modell Foundation

LASID, Latin American Society for Immunodeficiencies



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