Cleveland, OH
Be Your Own Healthcare Advocate

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Topics

▪ Learn!
  • Basic Facts about PI

▪ Live!
  • Living with & Managing PI
Primary Immunodeficiency Diseases
Acronym Confusion

PI
PID
PIDD
You Look Good!
How Do I Tell Others About PI?
Learn!
Facts about PI
What Are Primary Immunodeficiency Diseases?

- Primary immunodeficiency diseases (PI) are a group of more than 350 rare, chronic disorders in which part of the body’s immune system is missing or functions improperly.

- While not contagious, these diseases are caused by hereditary or genetic defects.
Who Does PI Affect?

- Approximately 250,000 people diagnosed with PI in the U.S. (1 in 1,200), and thousands more go undetected.

- Some disorders present at birth or in early childhood, but they can affect anyone, regardless of age or gender.

Source: 2006 IDF National Prevalence Survey
What Are the Symptoms of PI?

- Severe
- Persistent
- Unusual
- Recurrent
- Runs in the Family
How Is PI Diagnosed?

- Medical and family history, physical exam, blood and immunoglobulin level tests and vaccines to test the immune response may be included in the diagnosis process.

- **Over half** (52%) of individuals with PI are 30+ years of age when diagnosed.

- Average length of time between onset of symptoms & diagnosis is **15.3 years**.

Source: 2012 IDF National Patient Survey
How Is PI Treated?

- Depends upon specific diagnosis
- Various treatments available:
  - Antibiotic Therapy
  - Antifungal Therapy
  - Gamma Interferon
  - Gene Therapy
  - Granulocyte-Colony Stimulating Factors (G-CSF)
  - Hematopoietic Stem Cell Transplant (Bone Marrow Transplant)
  - Immunoglobulin (Ig) Replacement Therapy
  - PEG-ADA Enzyme Therapy
Live!
Living with & Managing PI
YOU Are the Coordinator of Your Care
Who Are the Members of Your Team?

- Family
- Friends
- Doctors
- Nurses
- Psychologist
- Pharmacist
- Specialty Pharmacy
- Social Worker
- Insurance Company
- And More…
"Before anything else, preparation is the key to success."

~ Alexander Graham Bell
Understand Your Rights

- Medical Information
- Choice of Healthcare Provider
- Involvement in Healthcare Decisions
- Fair Treatment & Non-discrimination
- Privacy & Protection
- Grievances
Save Important Information!

- Diagnosis information
  - Who made your diagnosis?
  - When?
  - Based on what data?
    - Treatment history
    - Lab data

- Documentation
  - Records regarding this information
  - Don’t assume someone else has these data
  - Consider using IDF ePHR

This information will help you manage treatment and insurance issues!
Keeping Everyone on the Same Page

- At every appointment request copies of everything for yourself and copies of everything to be sent to all providers.

- Track medications, allergies, infections, etc.

- Store pamphlets, medication and side effect inserts, and important papers and phone numbers in one place such as a file, notebook or IDF ePHR.
Immunoglobulin (Ig) Therapy

- **More than 4 out of 5** individuals with PI report being treated with immunoglobulin (Ig) replacement therapy, which is the standard of care for most antibody deficiencies.

- You have options when it comes to Ig therapy.

- Work with your healthcare provider to decide what works best for **YOU**!

Source: 2012 IDF National Patient Survey
Ig Therapy: Know Your Options

- **Product**
  - There is not a generic Ig product.
  - Find a product that works for you.

- **Mode of Administration**
  - **IVIG** – Intravenous
  - **SCIG** and **fSCIG** – Subcutaneous, Facilitated Subcutaneous
    - Multiple options for pumps, needle sets and tubing exist.
    - Consult your care team to ensure you have the correct combination.

- **Site of Care**
  - IVIG administered by an RN in the home, infusion suite or hospital
  - SCIG/fSCIG administered by the patient/caregiver at home
Ig Therapy: Know the Basics

Information to Know

- Ig product name
- Ig manufacturer’s name and patient assistance programs
- Amount infused, frequency and rate
- Name and dose of pre and post infusion medications

Take Action

- Report infusion related side-effects to your healthcare provider.
- Track Ig product lot numbers and all infusion related information
  
  **IDF ePHR: [www.idfephr.org](http://www.idfephr.org)**
- Register with the Patient Notification System for product recall info.
  
  **[www.patientnotificationsystem.org](http://www.patientnotificationsystem.org)**
NEW: IDF Guide to Ig Therapy

- Developed for patients and caregivers to help increase understanding of Ig replacement therapy.

- An increased understanding puts patients and caregivers in a better position to make informed decisions regarding care.

- These informed decisions will in turn improve overall health.

Available at No Cost to You: Stop by the IDF Table to Pick Up Your Guide!
The Appointment
The Question…

And how are you doing?
Notes for an Appointment

- Your questions.
- How you have been feeling (physically and emotionally).
- Changes in your body.
- Your worries and concerns.
- Issues related to treatments and side effects.
- Insurance and benefit coverage that could affect healthcare choices.
What Did They Say?

- Take notes during the appointment.

- Consider bringing a supportive family member or friend with you to appointments. That person can write down the information you receive and help you ask questions.

- If there is no one to who can go with you, ask your healthcare provider if you can record the meeting.
Communicating with Your Healthcare Team
I’m Sick – Now What?

- Prepare for when you get sick
- Develop an **illness protocol**
  - Ask your immunologist:
    - “How will I get my questions answered between scheduled appointments?”
    - “When and where I should go when I get sick?”
  - Prepare for possible emergency room and/or urgent care visits
    - Have your immunologist contact information and/or letter from your immunologist
    - Have basic health information (such as IDF ePHR)
  - Go the extra mile to prepare for ER/urgent care visits
    - Print or have access to IDF handbook chapter on your diagnosis
    - Have copy or digital access to IDF Diagnostic & Clinical Care Guidelines
Signs of a Communication Breakdown

- You don’t feel like you are part of your care team.

- Healthcare team members aren’t open to discussing your questions, concerns or problems.

- You are leaving an appointment with unanswered questions or a lack of information.
Improving Communication

- Be prepared with notes.
- Ask questions.
- Talk to other members of your healthcare team.
- Get a second opinion.
Good Communication Will Help You...

- Find out about current information related to your PI and treatment.
- Participate in decisions about your medical care.
- Better manage your care.
- Make the most of the time you have with your healthcare team.
- Reduce stress by making sure your questions are answered.
- Feel confident that you are getting the best healthcare.
IDF Is Here for YOU!

800-296-4433

www.primaryimmune.org/ask-idf