Managing Primary Immunodeficiency During the Senior Years

August 4, 2022

Immune Deficiency Foundation

DISCLAIMER

Immune Deficiency Foundation (IDF) education events offer a wide array of educational presentations, including presentations developed by healthcare and life management professionals invited to serve as presenters. The views and opinions expressed by guest speakers do not necessarily reflect the views and opinions of IDF.

The information presented during this event is not medical advice, nor is it intended to be a substitute for medical advice, diagnosis or treatment. Always seek the advice of a physician or other qualified health provider with questions concerning a medical condition. Never disregard professional medical advice, or delay seeking it based on information presented during the event.

Immune Deficiency Foundation

Immune Deficiency Foundation

MISSION

Improving the diagnosis, treatment, and quality of life of people affected by primary immunodeficiency through fostering a community empowered by advocacy, education, and research.

VISION

IDF seeks to ensure that everyone in the U.S. affected by PI has a fully informed understanding of

1. the PI diagnosis that affects them,
2. all available treatment options,
3. the expected standard of care,
4. all their opportunities for connection and support within the PI community.

Questions?

https://community.primaryimmune.org/s/newask

800-296-4433

Get Connected Groups

https://primaryimmune.org/support-services

Virtual groups exclusively for individuals & families living with PI

IDF Forums

Coming soon to your home!

- *August 18: Managing Autoimmune Issues & Primary Immunodeficiency*
- *August 24: Lunch & Learn* - Activated PI3K Delta Syndrome (APDS)
- *September 1: COVID-19, What We Know Now*

IDF Education Meeting

Kansas City, here we come!

Saturday, September 10, 2022

9:00 am – 2:00 pm


PI Conference
PRIMARY IMMUNODEFICIENCY IN FOCUS
Immune Deficiency Foundation
OCTOBER 6–8, 2022

THANK YOU TO OUR SPONSORS

A WORD FROM OUR SPONSOR

Immune Deficiency Foundation

The community built by us, for us. All of us.

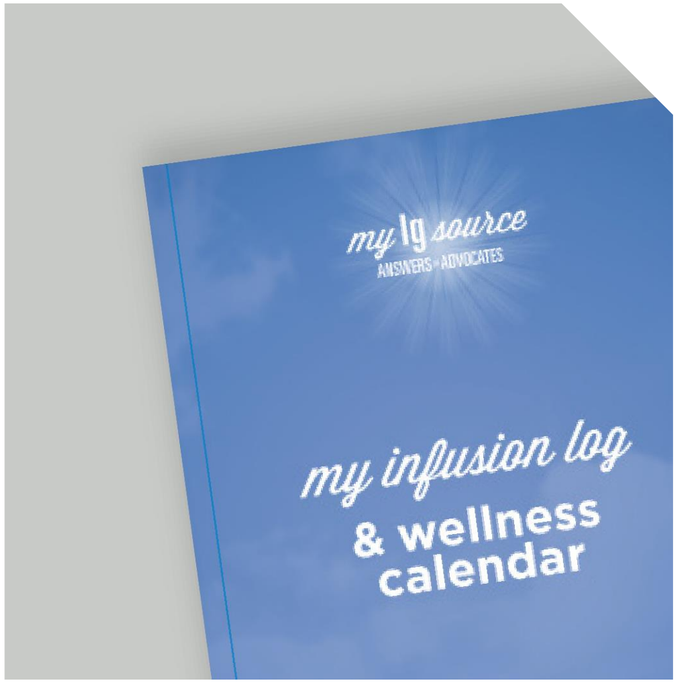
my Ig source | Takeda

Education is empowering.

Resources for people impacted by PI

You're not alone

MyIgEducation.com programs

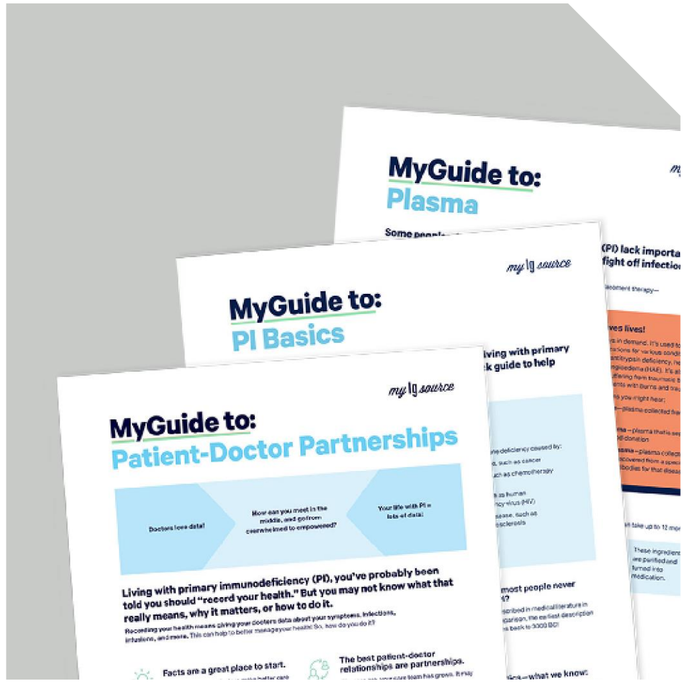


Infusion log & wellness calendar

my Ig source | Takeda

Education is empowering.

Resources for people impacted by PI



MyGuides

IGI therapeutic play kit

Patient Advocates

my Ig source | Takeda

Join the MyIgSource community.

This program is available to all patients and care partners, regardless of treatment. To enroll:

- Visit MyIgSource.com
- Call 855-250-5111

US-NON-7394v1.0 05/22

my Ig source | Takeda

A WORD FROM OUR SPONSOR

accredo®

Immune Deficiency Foundation

Accredo Health Group Inc., your specialty pharmacy

accredo®

Accredo Health Group Inc.

Who we are

- Best-in-class specialty pharmacy
- Structured around the patient
- Deep clinical focus
- Access to all IG brands

Who we serve

- Individuals with acute, chronic and complex medical or health conditions
- Urban as well as rural coverage
- Extensive experience serving Medicare and Medicaid patients
- Comprehensive payer relationships with over 3,000 individual contracts
- National coverage and payer relationships

What makes us unique

- Care model from condition-focused clinical teams
- 35+ immune-disorder pharmacists organized regionally
- 600+ field-based nurses
- National Customer Support Center (NCSC) staffed 24/7 answered directly by an Accredo-employed Registered Nurse

To reach our Accredo clinical team, call toll-free 866.820.IVIG (866.820.4844)

The Accredo Service Model

- Medications covered under both pharmacy and medical benefits
- Financial nurses who act as a resource for prior authorizations and appeals
- Coordination of benefits for rare and complex therapies, like infusion services and in-home nursing
- Broad access to third party copay assistance programs and alerts when charitable foundation funding is available, along with alternative funding options from other sources

~$7.9M

Secured in copay assistance for IG patients in 2021[1]

[1] Accredo data, 1/1/21 – 12/31/21

Managing Primary Immunodeficiency During the Senior Years

August 4, 2022

Immune Deficiency Foundation

Geriatric Primary Immune Deficiency
A Focus on Health Care Issues

Presented at the National Immune Deficiency Foundation Forum August 4, 2022

Roger H. Kobayashi MD
Clinical Professor
UCLA School of Medicine
National Consultant Immune Deficiency Foundation

A Humble Kahuna Lapa ‘au & Grandfather from Nebraska

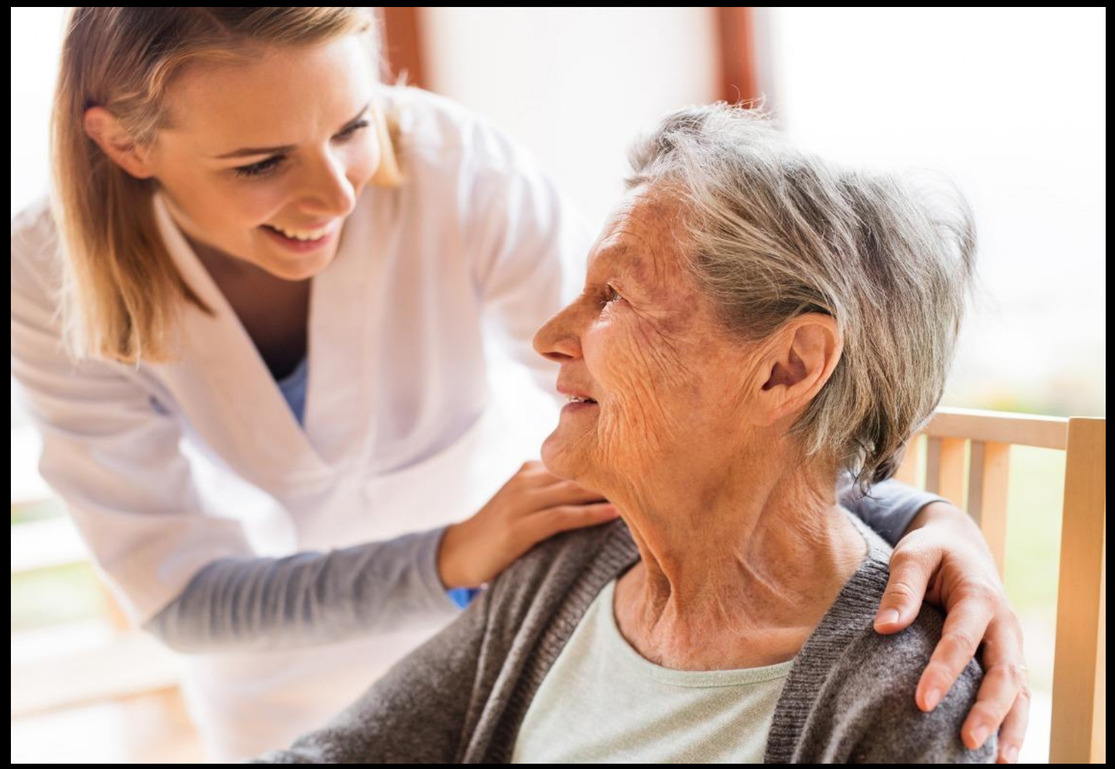
- Disclosures
- Clinical Immunologist 40 years
- Manage >320 patients on IgG.
- Clinical Professor: UCLA
- Lecturer [Hon]: Military Medical University Hanoi, Vietnam
- Grant support: Shire, Octapharma
- Consultant IDF, Octapharma.
- Reviewer: JACI-IP , Frontiers, Vox Sanguinis, others.
- Board Member: IfPA; National Biologic Physicians Working Group,
- Previously, Consultant for Bayer, Talecris, CSL/Sandoz, Baxter, ADMA, Shanghai Red Cross, American Red Cross

“Okage Sama De”

I am what I am because of you”

Japanese Proverb

Just as we cherish our children in America, so it must be that we cherish the elderly, for they are the foundation which made America the compassionate country it is and it is a debt and obligation that we can never repay.



“Kodomo no tame ni”

For the sake of the Children

Japanese Proverb

Bias of the Lecturer

- **I am not criticizing what has been done. I greatly appreciate & recognize the great strides made for the aged but we need to marshal our resources effectively & do more.**
- Geriatric PID is under recognized, under appreciated and constitutes a unique subpopulation of PID with unique needs.
- This group has many concomitant diseases [heart disease, cancer etc] which are problems in themselves but in combination with PID are much more difficult to manage
- The health care system makes it very difficult for seniors with serious, complicated diseases to interact with its bureaucracy, complexity & rigidity Our healthcare system is often not user friendly for the elderly. Particularly with the advent of internet communications.
- The elderly have a diminished and aging immune system and immune dysregulation may be a greater problem than in the younger population. This is underappreciated.
- The elderly's ability to self manage is overestimated and in addition to other health problems, they may have mobility problems, memory issues, frailty and diminished capacity to carry out treatment and instructions e.g. SCIG.
- In many instances, our health care system is configured for the convenience of the providers, hospitals, medical industry, insurance industry and technologists……..and less so for the patient.
- To paraphrase President Kennedy's famous inaugural address: our healthcare system should be telling themselves, **"Ask not what the elderly can do for you, ask what you can do for the elderly"...** The whole healthcare industrial complex should exist for the patient; not the other way around.

Outline of Discussion

- This discussion will review briefly health issues then focus on shortcomings of our approach to Geriatric PID:

  LACK OF RECOGNITION, UNDER APPRECIATION OF THE UNIQUE REQUIRENTS/ CHARAC - TERISTICS . SOCIAL ISOLATION, LACK OF MOBILITY, FRAILTY, COMMUNICATION.

- ***https://www.thedoctorschannel.com/view/a-call-to-action-to-address-unmet-needs-among-geriatric-patients-with-primary-immunodeficiency/
- PID is not rare in the geriatric age group **IN FACT ALL OLDSTERS MAY HAVE Immune Deficiency. Aging normally causes deterioration and loss of immune function.**
- Most adult PID patients have antibody deficiency➔but years to diagnose.
- Autoimmune diseases, inflammatory bowel disease or lymphomas may co-exist with PID, thus obscuring diagnosis.
- Older patients tend have complications/ co-existing diseases which impact and complicate PID

2015: A Concern About an Unmet Need

- 78 year old wonderful, kind woman with a long history of severe lung infections, chronic bronchitis, bronchiectasis, sinus disease referred by pulmonary specialists. Found to have CVID. Treated with IgG and appropriate ancillary medical management. Her husband, a saint, helped with SCIG and she began to thrive. At office visits, they were always cheerful and happy. Then the husband developed Alzheimer's and the roles reversed. She began to worry about him, she withered away and died.

- **This case bothered me and I began to wonder, "what's going on?"**

"How many elderly patients are we following, how many were managing themselves, was someone caring for them, did they have other diseases and how was that impacting their PIDD? What was their overall health status?"

How Common is Geriatric PID? It Turns Out We Didn't Know and No One Knows!

- Consortium of Independent Immunology Clinics – 22 private practice Immunology Clinics throughout the nation.

- **NONE OF US REALIZED THIS!**
- The CIIC follows ~ 2,000 patients ****778** registered were on IgG replacement therapy

Much to our amazement **40%** receiving IgG were over 60 years of age; only **11%** < 20 y/o

Another remarkable finding was that gender shifted from males in the younger age category to overwhelmingly females

- **2022 – in our clinic, *147 out of 283* on IgRT are over age 65 y/o**
- **2019** – Optum, a huge national infusion company had 500 geriatric patients on IVIG; success rate in switching to SCIG was only 10%. Now 70% on SCIG, but that creates a whole new set of problems.

Can You Imagine Our Surprise?

- 86 of 778 = **11% <20 y/o**
- 388 of 778 = 50% 20-60 y/o
- 304 of 778 = **39% > 60 y/o**

- **89%** on IgG replacement were adults [20 y/o or older]
- **40%** were older than 60 y/o
- More patients over *80 y/o* were on IgG infusions than under *10 years old*.

What an eye opener!!
Wow!!!

Critical Unanswered Questions

- What is the incidence of PIDD in the elderly? **No one knows**
- What type of PIDD do they have? **USIDNet now has the data**.
- How many are receiving IgG treatment? **No one knows**
- How many are receiving IVIG vs SCIG? **No one knows**
- Where are they getting infused?
- IVIG, ➔ **risk factors?**
- SCIG, ➔ **self infusing?**
- Is there a caregiver?
- **Are they doing it properly? Are they doing it regularly?**
- Do they have other diseases?
- Are those diseases such as heart disease, diabetes, kidney failure, arthritis, high blood pressure, visual impairment, mental impairment affecting their PIDD or their ability to treat and self manage their PIDD?
- **How does the aging process impact Primary Immune Deficiency?**
- **AS IT TURNS OUT, NO ONE KNOWS**.

“not even the Shadow knows”

Two Ends of the Spectrum

- **Immature:**

With infants and young children, the immune system is naïve, immature and developing => increased susceptibility to infections, poor response to vaccines, increased allergies.

- **Immunosenescence:**

With the elderly, the immune system gets “worn down”, doesn’t function as well, increased susceptibility to infections, poor response to vaccines, decreased tumor surveillance, increased auto-immunity, poor inflammation.

Immunosenescence

A tongue twister word which means your immune system is riding into the sunset

- Decrease in the number and function of T-cells & B-Cells
- T-cells ➔Decrease in thymic function; decrease in naïve T-cells.
- Decrease in T-cell diversity and repertoire
- T-cells become exhausted and don't respond well [like many of us getting fatigued from doing the same ol' job and dealing with bureaucracy]
- B-cells ➔ decrease in B-cell numbers, B-cell diversity, decreased antibody function, decreased affinity, decreased response.
- Increased memory cells, decreased naïve cells, cells become exhausted

- Increased infections, severity, poor respon-se, poor surveillance.

Results of an aging immune system

- Increased infections

e.g.Influenza, varicella, pneumococci, bad boy CoVID 19 [damned]

- Increased malignancy
- Increased autoimmune disease – one study in healthy older adults => 22% + RA 14% had elevated ANA
- Failure to make enough immune cells
- Structures which help cells differentiate and develop fail to do their job [thymus/lymphnodes]
- Cells that are made don’t work as well
- Cells don’t respond to signals and directions as well. [Sound familiar?].

What's the Long Term Prognosis for Seniors with CVID/PID?

- Depends upon the type of PID, concomitant illnesses & most importantly, presence of non-infectious complications.
- If antibody deficiency alone and infections, prognosis is great.
- However, if autoimmune or immune dysregulation abnorma-lities, then not as favorable.
- C. Cunningham Rundles 2012- ***mortality rate 11 times higher*** with N-inf vs. infectious symptoms.
- Blood 2012- 411 patients 19.6% died. Median age at death: F-44 M-42. Improved over last decade.
- PLOS 2021 Wilder: Faithful IgRT critical in improving outcomes.

Italian Retrospective Report: Biomedicines: 10: 635-647

M.G. Daniellei, C. Mezzanotte, J.U.Verga et al

- Study from a referral clinic in Central Italy Univ. Marche
- 78 patients with CVID followed average of 8.5 years
- Sixty five < 65 y/o 13 >65 y/o
- At diagnosis: Infections in 61% < 65 y/o and 69% > 65 y/o.
- Autoimmune diseases more common in younger [30% vs 18%]; cancer more common in older [38%]. ***However, they were only following 13 elderly patients.
- M/F ratio < 65 y/o 37%/63%
- M/F ratio > 65 y/o 15%/85%

- Autoimmunity: ITP, hemolytic anemia. Autoimmune hepatitis, Hashimoto's, arthritis, Sjogren's, vasculitis, vitiligo.
- Malignancies: lymphomas, M.M., G.I.
- Other neoplasias: stomach, pancreas, breast, skin, thyroid, bladder.

**** note other cancers occur in general population and increases with age.

What Are the Major Issues Geriatric PID Patients Face?

From Miller on

- Less ability to cope with infections particularly with diminishing immune function with age
- Increased incidence of autoimmune & inflammatory disorders: inflammatory arthritis, inflammatory diseases of the G.I. tract, cancer, diseases of the blood vessels [vasculitis], inflammation of the lungs
- Issues of mobility, frailty, memory loss, eye sight, hearing loss, social isolation, dependence upon caregivers.
- Concurrence of co-existent health diseases of aging: heart disease, kidney disease, hypertension, diabetes, osteoarthritis, cancer.
- Difficulty dealing effectively with a fragmented, highly complex, inconvenient , extremely expensive health care system. Monitoring critical!!!!!!

Rex Harrison: “Damned, Damned, Damned”

- Internet technology is wonderful but for the elderly “sounds Greek to me”
- Much of medical care and communication is internet based.
- Difficult to navigate even for nerds [try changing your reservations when you’re stranded at the airport or even ordering a Vietnamese sandwich at SFO] not to mention signing up for CoVID shots, critical interaction.
- What if you can’t get doctor’s messages, important lab reports, repondez si’l vous plais?
- So complicated, so Byzantine, so many organizations , so much information, so disorganized …. the elderly are often at a loss ….to their peril.

IVIG/SCIG?

- Safety
- Effectiveness
- Convenience
- Venue/Infusionist
- Caregiver
- Monitoring
- Patient Advocate
- Costs
- Confidence

How Shall We Proceed?

- **First: Recognize Geriatric Patients** [GPs] with PIDD: concomitant diseases & study the impact of these diseases on overall health
- **Need to figure out how to effectively communicate with GPID**
- **Identify & work** with family members or other responsible persons
- **Coordinate care** with the various specialty doctors; PCP's cannot because they are overwhelmed with work. ***Patient Navigators.***
- ***Coordination*** between **pharmacy services**, infusion services & physicians ***is critical***. ***Must be user friendly!!!!!!!!***

- **How** can we insure GPs are being treated consistently, safely & effectively?
- **How** can we insure that GPs are complying with treatment?
- **What help** is available to circumnavigate the morass of Medicare and other bureaucracies?
- Health care **costs** are prohibitive. IgG treatment costs $50,000 or more per year; combined with other medications, health care costs ….. **Staggering!**

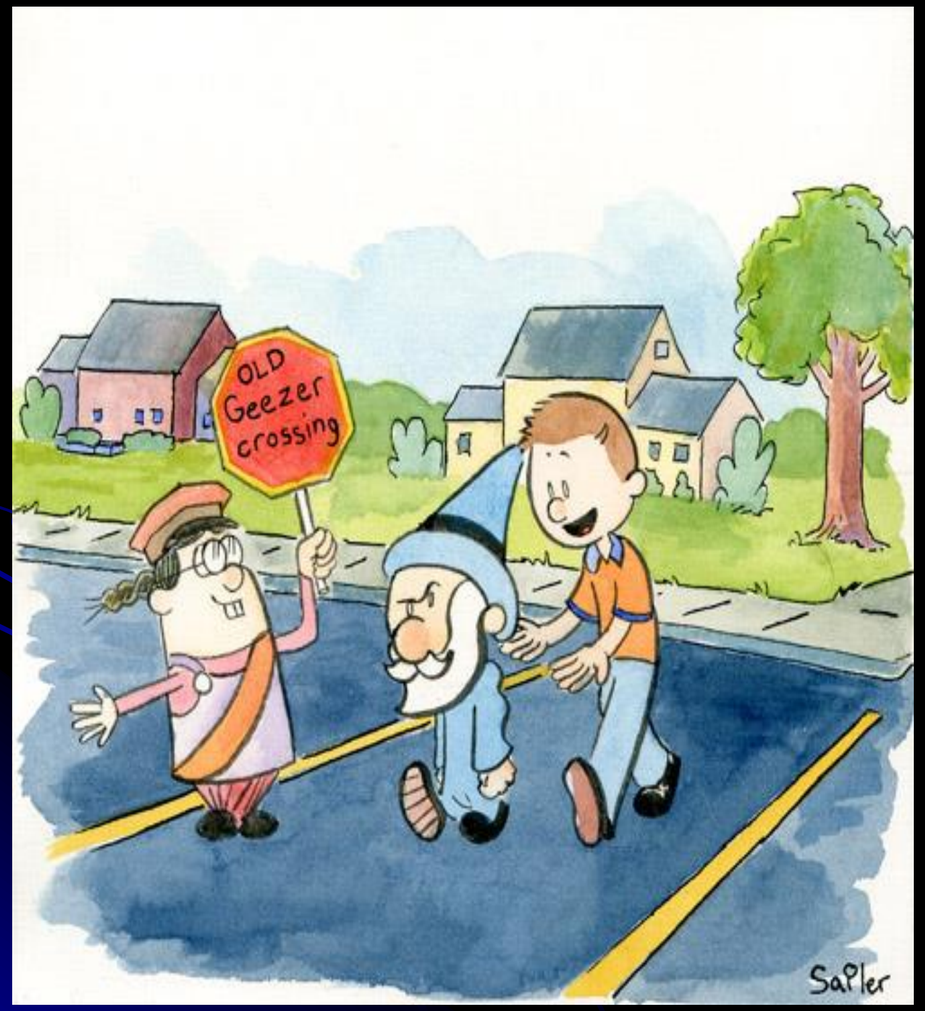
“Ho'okahi ka 'ilau like ana”

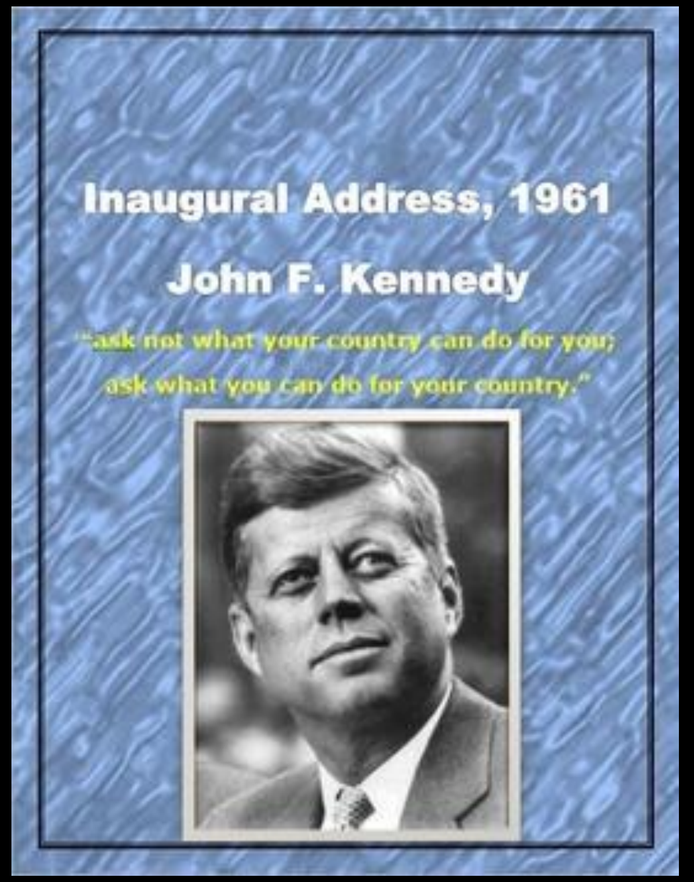
Put Your Paddle in and Join the Effort

- We need to recognize that GPs are a unique & growing group with special & unique issues.
- We need to coordinate care between pharmacy/ nursing/ physician and infusion services.
- We need to work with 3rd party payers to ensure coverage and support and KISS!!!!!?
- Anticipate complications
- Monitor. Patient Navigators plus Care Givers.
- Research must be done to understand & best help GPs
- We need to monitor & make sure GPs are cared for properly
- “Ho’okahi ka ‘ilau like ana”

Recognize, research & accommodate the unique needs of the Geriatric PID

- "ask not what the elderly can do for you. Ask what you can do for the elderly"





Let’s Help Our Geriatric PID’s “Rock & Roll!”

THANK YOU!

Roger H. Kobayashi, MD
Clinical Professor, University of California Medical School

Immune Deficiency Foundation

A WORD FROM OUR SPONSORS

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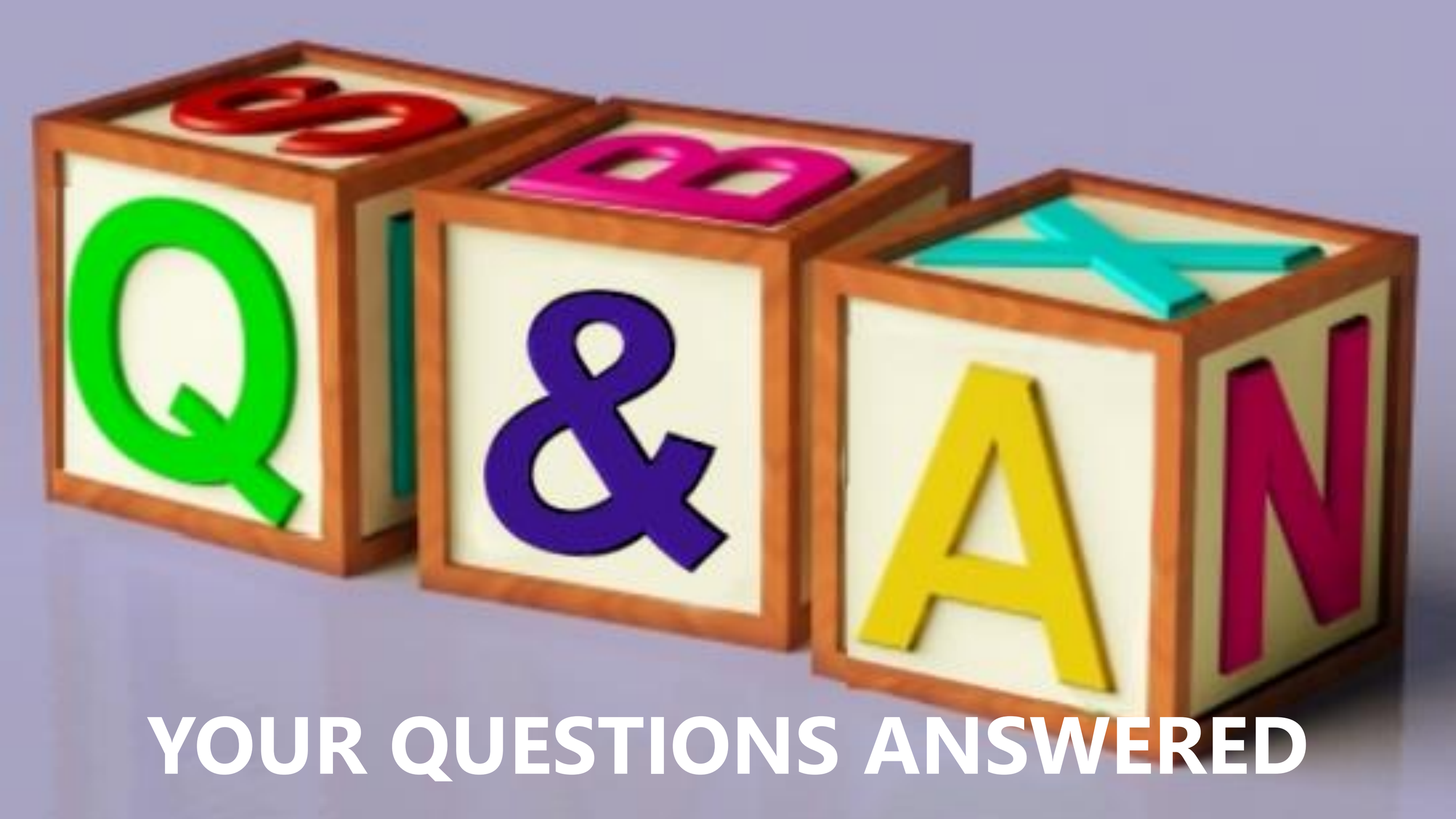

YOUR QUESTIONS ANSWERED

THANK YOU!

Roger H. Kobayashi, MD
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Laura Rohe, RN
Allergy, Asthma and Immunology Associates, P.C.

Immune Deficiency Foundation

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From all of us at IDF

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