TV-110-030420-90

SYLLABUS AND COURSE GUIDE

A Multidisciplinary Approach to the Diagnosis and Optimal Management of Primary Immunodeficiency: The Latest Evidence and Best Practices

A Free, 90-Minute CME/CNE/CPE/MIPS/ABIM MOC/ABP MOC Live and On-Demand Activity

Premiere Date: Wednesday, March 4, 2020

12:00 PM - 1:30 PM ET (live)

Credit Expiration Date: Thursday, March 4, 2021

On the Web: http://bit.ly/TV-110

LIVE FACULTY: Kristin Epland, MSN, FNP-C; Niraj C. Patel, MD, MS

MODERATOR: Mark Ballow, MD

Take advantage of our LIVE Q&A segment during this webcast!

During the webcast **type a question in the box under the presentation**

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This continuing education activity is provided by

CME Outfitters

INFORMATION FOR PARTICIPANTS

Statement of Need

Primary immunodeficiency disorders (PIDs) are a group of genetic disorders that affect development and function in the immune system. PIDs can leave patients vulnerable to frequent, severe, and unusual infections, and are associated with significant morbidity; patients with PIDs are twice as likely to be hospitalized and have significantly longer hospital stays.

Timely diagnosis and early treatment interventions are critically important to mitigate the disease and economic burden of PID, but the management of PID can be challenging for clinicians due to its complexity. Human immune globulin (Ig) therapy has significantly improved life expectancy and quality of life (QoL) for patients with PID; however, selecting a therapeutic product can be a complex choice, as it requires a thorough understanding of the classification of PIDs and the appropriate matching of a personalized treatment based on patient-specific factors.

This CME Outfitters Live and On Demand webcast is a case-based activity featuring expert faculty addressing the diagnosis and management of PIDs, as well as strategies for raising PID awareness and education.

Learning Objectives

At the end of this CE activity, participants should be able to:

- Identify signs and symptoms of primary immunodeficiency to decrease diagnostic delays.
- Implement evidence-based treatment strategies to manage PID.
- Educate and inform patients about PID to reduce morbidity and improve (QoL).

The following learning objectives pertain only to those requesting CNE or CPE credit:

- Identify signs and symptoms of primary immunodeficiency to decrease diagnostic delays.
- Discuss evidence-based treatment strategies to manage PID.
- Describe ways to educate and inform patients about PID to reduce morbidity and improve (QoL).

Target Audience

Primary care physicians, pediatricians, specialists, infusion nurses, pharmacists, nurse practitioners, and physician assistants.

Financial Support

Supported by educational grants from CSL Behring LLC, Grifols, and Pfizer Inc.

CREDIT INFORMATION

CME Credit (Physicians)

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Universal Activity Number:
Live: 0376-0000-20-006-L01-P; 0376-0000-20-006-H01-P
Type: knowledge-based

ABIM/MOC Credit:

Successful completion of this CME activity, which includes participation in the evaluation component, enables the participant to earn up to 1.5 MOC points in the American Board of Internal Medicine's (ABIM) Maintenance of Certification (MOC) program. Participants will earn MOC points equivalent to the amount of CME credits claimed for the activity. It is the CME activity provider's responsibility to submit participant completion information to ACCME for the purpose of granting ABIM MOC credit.

Learning Formats:

Live activity
Enduring Material

ABP MOC Credit:

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Royal College MOC:

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MIPS Improvement Activity:

This activity counts towards MIPS Improvement Activity requirements under the Medicare Access and CHIP Reauthorization Act of 2015 (MACRA). Clinicians should submit their improvement activities by attestation via the CMS Quality Payment Program website.

CREDIT REQUIREMENTS

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There is no fee for participation in this activity. The estimated time for completion is 90 minutes. Questions? Please call **877.CME.PROS**.

FACULTY BIOS & DISCLOSURES

Mark Ballow, MD (Moderator)

Dr. Ballow is currently a Professor in the pediatric department at the University of South Florida, and the Morsani College of Medicine at John Hopkins All Children's Hospital in St Petersburg, Florida in the Division of Allergy/Immunology. Dr. Ballow received his medical degree from the University of Chicago School of Medicine, Chicago, Illinois. He then completed an internship and residency in pediatrics at Yale-New Haven Hospital in New Haven, Connecticut, followed by a fellowship at the University of Minnesota Hospital in Minneapolis, Minnesota in clinical immunology under the mentorship of Dr. Robert Good. After finishing his time in the Army at Walter Reed Hospital, Dr. Ballow joined the department of pediatrics at the UConn School of Medicine in Farmington, CT. From 1988 to 2012 Dr. Ballow was Chief of the Division of Allergy, Immunology and Pediatric Rheumatology at the Women and Children's Hospital of Buffalo, an affiliate hospital of SUNY Buffalo, School of Medicine and Biomedical Sciences as well as Training Program Director of the Allergy/Immunology fellowship program, and Director of the Immunobiology Laboratory. Dr. Ballow is board certified in pediatrics, allergy and immunology, and clinical laboratory immunology. Dr. Ballow was the President (2010-2011) of the American Academy of Allergy, Asthma. and Immunology (AAAAI). Dr. Ballow is also a member of the American College of Allergy, Asthma and Immunology, and the Clinical Immunology Society. He was a member of the Blood Product Advisory Committee of CBER/FDA. He currently serves on the medical advisory committee for the Immune Deficiency Foundation (IDF) and is consulting medical director for the IDF. Dr. Ballow is on the editorial board for *Journal of Allergy and Clinical Immunology: In Practice*, was co-editor for *Current Opinion in Allergy and Clinical Immunology* between 2001-2018. Dr. Ballow is author or coauthor of more than 180 peer-reviewed papers, 50 books/book chapters or monographs, and more than 100 abstracts. He serves on the data safety monitoring boards for four pharmaceutical phase III trials. His areas of research interest are primary immune deficiency disorders and immunoglobulin (IVIG) replacement therapy and its mechanisms of action.

Kristin Epland, MSN, FNP-C

Ms. Epland is a Family Nurse Practitioner specializing in the care and diagnosis of primary immunodeficiencies and autoimmune diseases at the Midwest Immunology Clinic in Minnesota. Ms. Epland is a 1998 graduate of the University of Minnesota School of Nursing Family Nurse Practitioner program. She has worked with children and adults with immunodeficiency diseases for over 20 years through home infusion nursing and currently as a part of Midwest Immunology Clinic and Infusion Center in Plymouth, Minnesota. She is presently a member, and past chairperson, of the Nurses Advisory Committee of the Immune Deficiency Foundation.

Niraj C. Patel, MD, MS

Dr. Patel is an associate professor of Pediatrics and Chief of the Division of Infectious Disease and Immunology at Levine Children's Hospital in Charlotte, NC. Dr. Patel received his medical degree from the University of Louisville. After completing his residency in pediatrics, also at the University of Louisville, he went on to complete fellowship training in both allergy and immunology and infectious diseases at Texas Children's Hospital, Baylor College of Medicine in Houston, TX. During this time, Dr. Patel also received a master's degree in clinical investigation.

Dr. Patel is a member of the Clinical Immunology Society, the American Academy of Allergy, Asthma, & Immunology, the American College of Allergy, Asthma and Immunology, and the Infectious Disease Society of America. Dr. Patel has been an author/coauthor of articles published in several peer-reviewed journals, including the *New England Journal of Medicine*, the *Journal of Allergy and Clinical Immunology*, and the *Journal of Clinical Immunology*.

His academic and clinical interests include primary immunodeficiency diseases and infections in immunocompromised hosts. Dr. Patel enjoys spending time with his wife, gardening, chauffeuring his 3 children around town, and is a national Gold Medal Champion in Tae Kwon Do.

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It is the policy of CME Outfitters, LLC, to ensure independence, balance, objectivity, and scientific rigor and integrity in all of their CE activities. Faculty must disclose to the participants any relationships with commercial companies whose products or devices may be mentioned in faculty presentations, or with the commercial supporter of this CE activity. CME Outfitters, LLC, has evaluated, identified, and attempted to resolve any potential conflicts of interest through a rigorous content validation procedure, use of evidence-based data/research, and a multidisciplinary peer review process. The following information is for participant information only. It is not assumed that these relationships will have a negative impact on the presentations.

Dr. Ballow reports that he is on the advisory committee for CSL Behring; and Takeda Pharmaceuticals U.S.A., Inc. He is a consultant for Grifols. He is on the speakers bureau for CSL Behring and Takeda Pharmaceuticals U.S.A., Inc.

Ms. Epland reports that she serves on the advisory committee for IDF Nurse Advisory Committee; Pharming Group NV and Takeda Pharmaceuticals, Inc. U.S.A. (Immunoglobulin Nurse Advisor). She serves as a consultant for the Takeda Pharmaceuticals, Inc. U.S.A speaker program.

Dr. Patel reports that he received research support from CSL Behring and Takeda. He is on the advisory committee for Baxalta Inc. and Horizon Therapeutics. He is on the speakers bureau for CSL Behring; Horizon Therapeutics and Takeda Pharmaceuticals U.S.A., Inc.

Howard Bliwise, MD (peer reviewer) has no disclosures to report.

Mae Ochoa, RPh (peer reviewer) has no disclosures to report.

Poshala Tish Aluwihare, PhD (planning committee) has no disclosures to report.

Evan Luberger (planning committee) has no disclosures to report.

Jan Perez (planning committee) has no disclosures to report.

Sharon Tordoff (planning committee) has no disclosures to report.

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CME Outfitters, LLC, gratefully acknowledges educational grants from CSL Behring LLC, Grifols, and Pfizer Inc. in support of this CME/CE activity.

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CONTINUING MEDICAL EDUCATION

Mark Ballow, MD

Professor, Department of Pediatrics
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Mark Ballow, MD
Disclosures

- ***Speakers Bureau:*** CSL Behring; Takeda Pharmaceuticals USA Inc.
- ***Consultant:*** Grifols
- ***Advisory Board:*** CSL Behring; Takeda Pharmaceuticals USA Inc.

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CONTINUING MEDICAL EDUCATION

Niraj C. Patel, MD, MS

Chief, Pediatric Infectious Disease and Immunology
Director, Charlotte Immunodeficiency Center
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Disclosures

- ***Research/Grants:*** CSL Behring; Takeda Pharmaceuticals USA Inc.
- ***Speakers Bureau:*** CSL Behring; Horizon Therapeutics; Takeda Pharmaceuticals USA Inc.
- ***Advisory Board:*** Baxalta Inc. and Horizon Therapeutics

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Family Nurse Practitioner
Midwest Immunology Clinic
Plymouth, MN

Kristin Epland, MSN, FNP-C

Disclosures

- ***Consultant:*** Takeda Pharmaceuticals, Inc. U.S.A speaker program
- ***Advisory Board:*** IDF Nurse Advisory Committee; Pharming Group NV; Takeda Pharmaceuticals, Inc. U.S.A (Immunoglobulin Nurse Advisor)

CME Outfitters
CONTINUING MEDICAL EDUCATION

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March 4, 2020

Primary Immunodeficiency Disorders

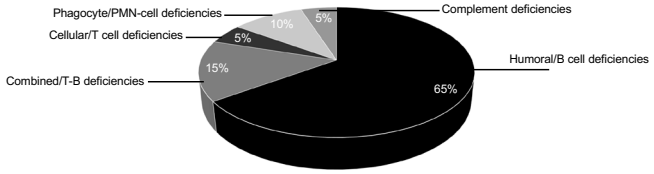
CME Outfitters
CONTINUING MEDICAL EDUCATION

Learning Objective 1

Identify signs and symptoms of primary immunodeficiency to decrease diagnostic delays.

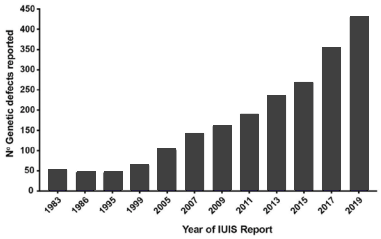
Primary Immunodeficiencies (PI)

- Prevalence:
  - IgA Deficiency - 1/600
  - CVID - 1/25000
  - SCID - 1/65,000



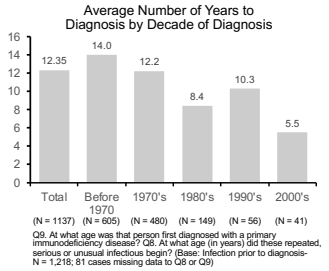
CVID = common variable immune deficiencies; Ig = immunoglobulin; PI = primary immunodeficiencies; SCID = severe combined immune deficiencies.
Bellanti, JA. *J Clin Immunol.* 2012;32(3):647; Kobrynski L, et al. *J Clin Immunol.* 2014;34(8):954-961; Ludvigsson JF, et al. *J Clin Immunol.* 2014;34(4):444-451; Tam JS, Routes JM. *Am J Rhinol Allergy.* 2013;27(4):260-265. Dorsey MJ, Puck JM. Immunol Allergy Clin North Am. 2019;39(1):1-11.

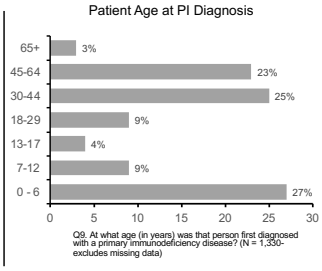
Inborn Errors of PI



IEI: = Inborn errors of immunity; IUIS = International Union of Immunological Societies.
SG Tangye, et al. *J Clin Immunol*. 2020 Jan 17. [Epub ahead of print].

Diagnostic Delays





Immune Deficiency Foundation. Primary immune deficiency diseases in America: 2007 the third national survey of patients. 2007. https://primaryimmune.org/wp-content/uploads/2011/04/Primary-Immunodeficiency-Diseases-in-America-2007The-Third-National-Survey-of-Patients.pdf.

Debra -Patient History

- 43-year-old woman
- Asthma since age 10
  - Hospitalized several times, recent hospitalization 1 year ago
    - Chronic bronchitis
      - Persistent cough, productive sputum especially in AM
    - Combination inhaled steroid, LABA
    - Allergy skin testing 7 years ago -negative
- Recurrent sinusitis
  - Saline washes, topical nasal steroids
  - Symptoms of post-nasal drainage
- ITP treated with IVIG 10 years prior

ITP = immune thrombocytopenic purpura; IVIG = intravenous immunoglobulin; LABA = long-acting beta-agonist.

The 10 Warning Signs of PI

1. Four or more new ear infections within 1 year
2. Two or more serious infections within 1 year
3. Two or more months on antibiotics with little effect
4. Two or more pneumonias within 1 year
5. Failure of an infant to gain weight or grow normally
6. Recurrent, deep skin or organ abscesses
7. Persistent thrush in mouth or fungal infection on skin
8. Need for intravenous antibiotics to clear infections
9. Two or more deep-seated infections including septicemia
10. A family history of PI

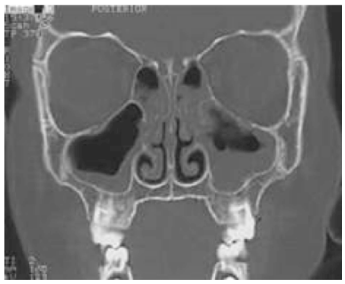
Jeffrey Modell Foundation Medical Advisory Board. 2016. http://downloads.info4pi.org/pdfs/10-Warning-Signs---Generic-Text--2-.pdf.

Debra - Physical Examination

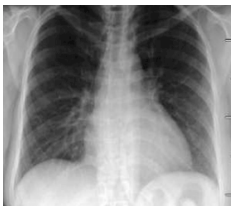
- HEENT –
  - Slightly inflamed nasal mucosa
    - Yellow secretions
  - Cobblestoning in posterior pharynx
    - White secretions
- Chest –
  - Scattered rhonchi and wheezes
- Abdomen –
  - Liver palpable 4 cm below the right costal margin
  - Spleen tip felt
- Other –
  - Anterior cervical lymph nodes present
  - A few small right axillary nodes

HEENT = head, ears, eyes, nose, throat.

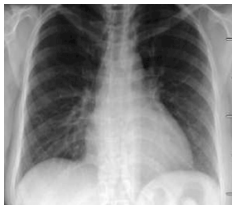
Debra - Imaging Studies

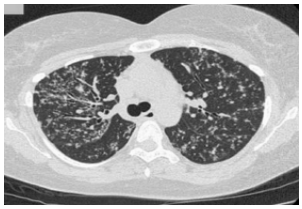


Debra - Imaging Studies



Debra - Imaging Studies





Humoral Immune Evaluation

- **Adaptive Immune System**
  - 1st stage
    - CBC with differential
    - Immunoglobulin production
    - IgG subclasses
  - 2nd stage
    - Isohemagglutinins
    - **Vaccine response**
    - Vaccine-specific antibody responses

CBC = complete blood count.
Bonilla FA. *J Allergy Clin Immunol.* 2018;141(2):474–481; Marsh RA, Orange JS. *Ann Allergy Asthma Immunol.* 2019;123(5):444–453.

Vaccines: The "Gold" Standard for Assessing Humoral Immune Function and Antibody Deficiency

Common vaccines:

- Tetanus (≥ 0.15 IU/mL), diphtheria (≥ 0.1 IU/ml) toxoid vaccines
- *Haemophilus influenzae* type B (HIB) conjugate vaccine (≥ 1.0 µg/mL)
- Pneumococcal polysaccharide vaccines (≥ 1.3 µg/mL)
- Influenza A/B (> 40 HI titer)

Orange JS, et al. *J Allergy Clin Immunol.* 2012;130(3 Suppl):S1-24.

Immune Evaluation

- Immune phenotyping/cell counts
  - 3rd Stage
    - Lymphocyte subset counts
  - 4th stage
    - B cell panel
    - T cell panel
    - Lymphocyte proliferative responses to mitogens/antigens

Cabral-Marques O, et al. *Front Immunol.* 2019;10:2742; McCusker C, et al. *Allergy Asthma Clin Immunol.* 2018;14(Suppl 2):61.

Debra - Laboratory Testing

- CMP - normal
- CBC with differential:
  - WBC - 14,250; normal differential
  - Hb and HCT – normal
  - Platelet count – normal
- Serum quantitative immunoglobulins:
  - IgG – 240 mg/dl
    - Normal range = 620 - 1400 mg/dL
  - IgA – 23 mg/dl
    - Normal range = 80 - 350 mg/dL
  - IgM - 40 mg/dL
    - Normal range = 45 - 250 mg/dL
  - IgE – 1 IU/ml
- Specific antibodies –
  - Tetanus – 0.1 IU/ml
  - Diphtheria – <0.04
  - Isohemagglutinins:
    - type 0 – anti-A -1:2 and anti-B – 1:4
  - Pneumococcal polysaccharides:
    - 2 of 23 serotypes in the protective range
- No recent TDAP or pneumococcal vaccines

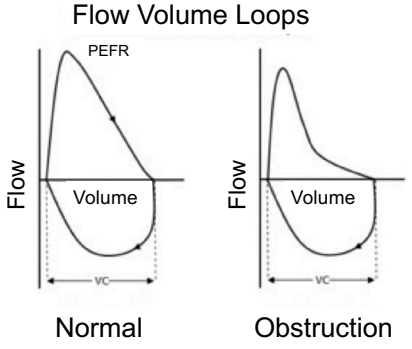
CMP = complete metabolic panel; Hb = hemoglobin; HCT = hematocrit; TDAP = tetanus, diphtheria, and pertussis; WBC = white blood cell.

Debra - Differential Diagnosis

- Primary immune deficiency
  - Exclude secondary causes
  - Common variable immunodeficiency
- Chronic lung disease:
  - Asthma
  - Bronchiectasis
  - Granulomatous interstitial lymphocytic lung disease
- Lymphoproliferative disease:
  - Hepatosplenomegaly
  - Nodular lymphoid hyperplasia
- Past history of ITP

ITP = immune thrombocytopenic purpura.

Debra's Spirometry



Diagnostic Criteria for CVID –ESID 2014

- At least one of the following
  - Increased susceptibility to infection
  - Autoimmune disease
  - Granulomatous disease
  - Unexplained polyclonal lymphoproliferation
  - Affected family member with antibody deficiency
- *AND* marked decrease in serum IgG and decrease IgA with or without a low IgM
- *AND* at least one of the following
  - Poor antibody response to vaccines (and/or absent isohemagglutinins)
  - Low switched memory B-cells
- *AND* secondary causes of hypogammaglobulinemia have been excluded
- *AND* diagnosis after age 4
- *AND* no evidence of profound T-cell deficiency

Seidel MG, et al. *J Allergy Clin Immunol Pract.* 2019;7(6):1763-1770.

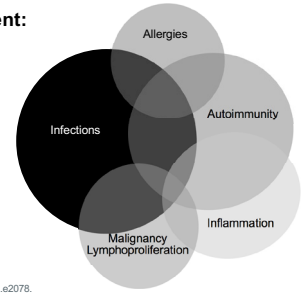
Debra's Diagnosis

- Primary antibody deficiency disease – CVID
- Lung disease
- Lymphoproliferative issues

Clinical Spectrum of PI in Adults

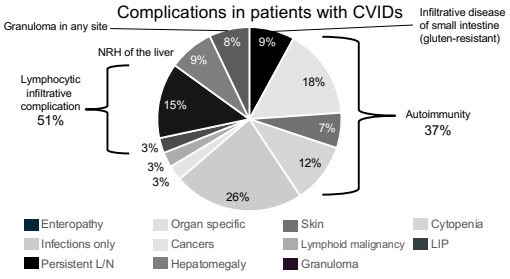
Multidisciplinary management:

- Primary care
- Allergy/Immunology
- Pulmonology
- Hematology
- Gastroenterology
- Infectious disease
- ENT
- Nutritionists/dieticians

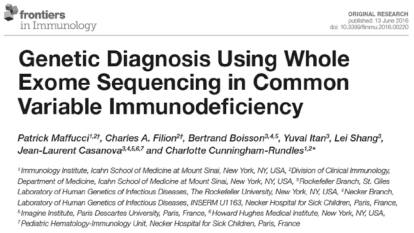


Bonilla FA, et al. *J Allergy Clin Immunol*. 2015;136(5):1186–205.e2078.

Medical Complications in Patients With CVID



L/N = lymphadenopathy; LIP = lymphoid interstitial pneumonitis; NRH = nodular regenerative hyperplasia.
Chapel H, Cunningham-Rundles C. *Br J Haematol*. 2009;145(6):709-727.


frontiers in Immunology

ORIGINAL RESEARCH

Genetic Diagnosis Using Whole Exome Sequencing in Common Variable Immunodeficiency

Patrick Maffucci[1,2†], Charles A. Filion[2†], Bertrand Boisson[3,4,5], Yuval Itan[3], Lei Shang[3], Jean-Laurent Casanova[3,4,5,6,7] and Charlotte Cunningham-Rundles[1,2]*

[1] Immunology Institute, Icahn School of Medicine at Mount Sinai, New York, NY, USA, [2] Division of Clinical Immunology, Department of Medicine, Icahn School of Medicine at Mount Sinai, New York, NY, USA, [3] Rockefeller Branch, St. Giles Laboratory of Human Genetics of Infectious Diseases, The Rockefeller University, New York, NY, USA, [4] Necker Branch, Laboratory of Human Genetics of Infectious Diseases, INSERM U1163, Necker Hospital for Sick Children, Paris, France, [5] Imagine Institute, Paris Descartes University, Paris, France, [6] Howard Hughes Medical Institute, New York, NY, USA, [7] Pediatric Hematology-Immunology Unit, Necker Hospital for Sick Children, Paris, France

- 50 CVID patients (average age 36 yr) with one or more of the following:
  - early onset
  - autoimmune/inflammatory manifestations
  - low B lymphocytes
  - and/or familial history of hypogammaglobulinemia
- Targeted gene screening (269 genes): 40% of patients with genetic diagnosis

Maffucci P, et al. *Front Immunol*. 2016;7:220.

CME Outfitters
CONTINUING MEDICAL EDUCATION

Learning Objective 2

Implement evidence-based treatment strategies to manage PI.

Debra – Management

- Antibody deficiency disease – CVID
  - Start Ig replacement therapy
- Sinus disease
  - Nasal irrigation
- Lung disease
  - Diffusion capacity
  - Prophylactic antibiotics
  - Possible lung biopsy
- Lymphoproliferative issues
  - Consider lymph node biopsy to r/o lymphoma

r/o = rule out.

IG Products

Route	Product	Dosage form
IV	Asceniv	10% liquid
	Bivigam	10% liquid
	Flebogamma DIF 5%	5% liquid
	Flebogamma DIF 10%	10% liquid
	Gammagard 5% S/D	Lyophilized
	Gammaplex	5% liquid
	Gammaplex	10% liquid
	Octagam 5%	5% liquid
	Octagam 10%	10% liquid
	Panzyga 10%	10% liquid
	Privigen	10% liquid
IV or SC	Gammagard liquid	10% liquid
	Gammaked	10% liquid
	Gamunex-C	10% liquid
SC	Cutaquig	16.5% solution
	Cuvitru	20% solution
	Hizentra	20% liquid
	Hyqvia	10% liquid + hyaluronidase
	Xembify	20% liquid

IV = intravenous; SC = subcutaneous.

Considerations for Selecting an Ig Product

	Intravenous Immunoglobulin (IVIG)	Subcutaneous Immunoglobulin (SCIG)	Hyaluronidase Facilitated Immunoglobulin (fSCIG)
Who?	Indicated for adult and pediatric patients with PI.	Indicated for adult and pediatric patients with PI.	Indicated for adult patients with PI.
How?	Usually administered by a nurse.	Self-administered.	Either self-administered or given by a nurse.
Where does it go?	Infused directly into the bloodstream through vein.	Infused/ injected under the skin into the subcutaneous tissues of arms, belly, outer buttock or thighs.	Infused under skin into subcutaneous tissues of belly, outer buttock or thighs.
When?	Usually given every 3-4 weeks.	Can be given on a flexible schedule from daily to every 2 weeks.	Can be given every 3-4 weeks.
How long?	Can take 2-6 hours to infuse.	Can take 5 minutes to 2 hours to infuse or inject.	Can take 1-2 hours to infuse.
Where is it given?	Home, hospital or outpatient infusion center.	Usually home setting after patient training	Home or outpatient infusion center
Side effects?	Often related to rate of infusion. Treat/prevent with other medications	Skin can be red and irritated at injection site. Often improves with each injection.	Skin can be red and irritated at injection site. Often improves with each injection. Volume per injection is larger than standard SC injection, so volume is more visible under skin, and may take 48-72 hours to absorb.

Immune Deficiency Foundation, USA. IDF guide to Ig therapy. 2018. https://primaryimmune.org/sites/default/files/publications/IDF%20Guide%20to%20Ig%20Therapy.pdf.

Facilitating Effective IVIG Delivery: Infusion Issues

- Dosing
- Infusion interval
- Infusion rates
- Monitoring

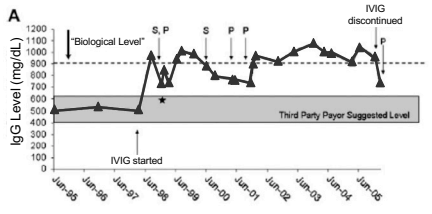
FA Bonilla, et al. *J Allergy Clin Immunol.* 2015;136(5):1186-1205.e1-78; EE Perez, et al. *J Allergy Clin Immunol.* 2017;139(3S): S1-S46; Sriaroon P, Ballow M. *Immunol Allergy Clin North Am.* 2015;35(4):713–730; Wasserman RL. *Immunol Allergy Clin North Am.* 2019;39(1):95-111.

Facilitating Effective SCIG Delivery: Infusion Issues

- Flexible dosing and schedule
  - Infusion interval
  - Volume per site
    - No. of sites
  - Infusion rate
- Supplies
  - Choice of needles
  - Choice of pumps

Skoda-Smith S, et al. *Ther Clin Risk Manag.* 2010;6:1-10; Thepot S, et al. *J Clin Immunol.* 2010;30(4):602-606.; Misbah S, et al. *Clin Exp Immunol.* 2009;158 Suppl 1:51-59; EE Perez, et al. *J Allergy Clin Immunol.* 2017;139(3S):S1-S46.; Jolles S, et al. *Clin Exp Immunol.* 2015;179(2):146–160. Wasserman RL. *Immunol Allergy Clin North Am.* 2019;39(1):95-111; Subcutaneous immunoglobulin (SCIg) Clinical Practice Guidance Principles. 2017. https://www2.health.vic.gov.au/-/media/health/files/collections/forms-and-templates/scig-clinical-practice-guidance-principles.pdf?la=en&hash=85378A7F2557DCDA79F9F83F0F4BC8033D0A2E8F.

Biological Trough Levels

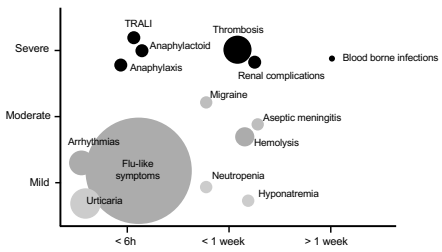


Titrate IgG trough level to clinical efficacy for an individual patient

S = acute sinusitis; P = pneumonia.

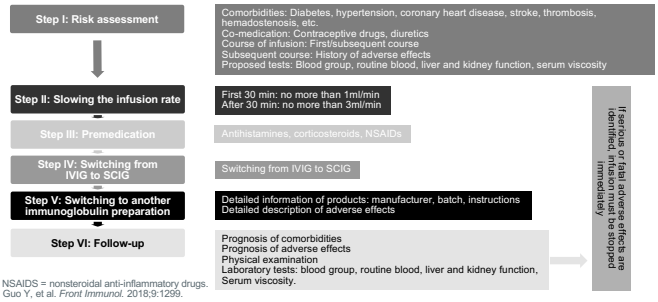
Bonagura VR, et al. *J Allergy Clin Immunol.* 2008;122(1):210-212.

IVIG Adverse Events



AE = adverse effects; TRALI = transfusion-related acute lung injury.
Vitiello G, et al. *Intern Emerg Med.* 2019;14(7):1041-1049.

Minimizing Ig-Associated AEs



NSAIDS = nonsteroidal anti-inflammatory drugs.
Guo Y, et al. *Front Immunol.* 2018;9:1299.

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Learning Objective 3

Educate and inform patients about PI to reduce morbidity and improve quality of life (QoL).

Shared Decision Making in PI

- Support patients to achieve informed preferences
  - Share best available evidence for Ig replacement therapy
- Discuss benefits/challenges of therapy options on an ongoing basis
- Select product/route of administration to minimize burden of care
  - Include patient/parent in SDM when choosing mode of IG administration, wherever possible

SDM = shared decision making.
Elwyn G, et al. *BMJ.* 2018;357:j1744; Lamb CC. *LymphoSign J.* 2018;5(3):100-114.

Considerations for Selecting an Ig Product

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Immune Deficiency Foundation, USA. IDF guide to Ig therapy. 2018. https://primaryimmune.org/sites/default/files/publications/IDF%20Guide%20to%20Ig%20Therapy.pdf.

hrQOL in CVID Patients Under Different Schedules of Ig Administration

- IgRT schedules do not impact the hrQoL in CVID if the treatment is established after an extensive educational period focused on individualizing the best therapeutic regimen

hrQOL = health-related quality of life; IgRT = immunoglobulin replacement therapy.
Pulvirenti F, et al. *J Clin Immunol.* 2019;39(2):159-170.

SMART Goals

Specific, **M**easurable, **A**ttainable, **R**elevant, **T**imely

- Conduct a careful history, physical exam, and screening evaluation that includes quantitative and qualitative tests to identify patients with PID.
- Individualize Ig dose and delivery to prevent infection, improve adherence, and QoL.
- Discuss therapeutic considerations and challenges, whenever possible, with patients on an ongoing basis.

Additional Resources

Visit the

Infectious Disease Hub

Where you will find free **PI resources**, including **FAQs** and a **Whiteboard Animation** designed to answer basic questions and empower patients to be armed with questions or concerns related to their PI care.

Whiteboard Animation

www.cmeoutfitters.com/infectious-disease-hub/

Questions for Faculty?

Type a question in the box under the presentation

OR

E-mail:
questions@cmeoutfitters.com

Coming Up...

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AFTER THE SHOW

Questions & Answers

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1. Actively participate in the meeting by **responding to questions** and/or **asking the faculty questions**
   (It's ok if you miss answering a question or get them wrong, you can still claim MOC)
2. Complete your post-test and evaluation at the conclusion of the webcast
3. Be sure to fill in your **ABIM ID number** and **DOB** (MM/DD) on the evaluation, so we can submit your credit to ABIM.

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- Actively participate by responding to ARS and/or asking the faculty questions
- Complete activity posttest and evaluation at the link provided
- Over the next 90 days, actively work to incorporate improvements in your clinical practice from this presentation.
- Complete the follow-up survey from CME Outfitters in approximately 3 months

CME Outfitters will send you confirmation of your participation to submit to CMS attesting to your completion of a CME for MIPS Improvement Activity.

CME for MIPS IMPROVEMENT ACTIVITY

CME Outfitters CONTINUING MEDICAL EDUCATION

A Multidisciplinary Approach to the Diagnosis and Optimal Management of Primary Immunodeficiency: The Latest Evidence and Best Practices

March 4, 2020

This continuing education activity is provided by CME Outfitters

TV-110-030420-90

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Please complete and FAX to **614.929.3600**

Activity Title and Faculty:

A Multidisciplinary Approach to the Diagnosis and Optimal Management of Primary Immunodeficiency: The Latest Evidence and Best Practices

with Mark Ballow, MD (Moderator); Kristin Epland, MSN, FNP-C; Niraj C. Patel, MD, MS

Site/Institution Name: ____

Practice Setting: ❑ Office-based ❑ Hospital ❑ Clinic ❑ Managed Care ❑ Small Group Practice (less than 5) ❑ Large Group Practice (more than 5) ❑ Other: ____________________

Address: ____

City: ______________________ State: ________ ZIP: ____________

Site Coordinator: ______________________ Phone: ______________________

Fax: ______________________ Email: ______________________

Completion Date: ____________ We participated in: ______________________

Attendee Name (please print)	Please Circle Discipline						
____________________	MD	DO	PA	NP	RN	Pharm	Other: __________
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____________________	MD	DO	PA	NP	RN	Pharm	Other: __________
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Questions? Call 877.CME.PROS. Thank you for participating in this continuing education activity!