

Immunology Update

Winter 2018

FROM THE JOHNS HOPKINS ADULT PRIMARY IMMUNODEFICIENCY CENTER OF EXCELLENCE

MESSAGE FROM THE DIRECTOR



Welcome to our first installment of Immunology Update! With this publication, we hope to keep you up-to-date with the work of the Johns Hopkins Adult Primary Immunodeficiency Center, as well as the latest in immunology medical advances. I welcome your feedback as we seek to provide a connection point and resource for people living with the challenges of weakened immune systems.

Over the course of my career, I have felt the pain, the anxiety, and in some cases desperation, of people who have spent years coping with illness, bouncing from doctor to doctor with no diagnosis or cure, and feeling unheard and unseen. We launched the Adult Primary Immunodeficiency Center to create a home for these patients, a home where they are welcomed and cared for in a comprehensive way by a multidisciplinary team who understands their unique and complex needs. Patients come to us from all over the world because of the level of precision and personal care they receive at the Center.

At the same time that we are drawing international patients into our Center, we are taking the Center's discoveries to a national and international audience. Not enough physicians are trained to diagnose and treat patients with immune deficiencies, but as you will see from this newsletter, we are being invited to teach and educate colleagues around the globe.

The impact of our Center is growing, and we are grateful to you for being an important part of our community and helping us to change the lives of so many people.

Warm regards,

Antoine Azar

ANTOINE AZAR, M.D.
 Director, Johns Hopkins Adult Primary Immunodeficiency Center

DAWN BORST
 Nurse, Johns Hopkins Adult Primary Immunodeficiency Center

OUR MISSION

To diagnose, treat, and cure Primary Immunodeficiency in adults.

<https://www.hopkinsmedicine.org/allergy/adult-primary-immunodeficiency-center.html>

RECENT MILESTONES

AWARDS AND HONORS

Dr. Antoine Azar is the 2018 recipient of the **Recognizing Excellence in Diagnosis Award** from the Vasculitis Foundation

AROUND THE GLOBE

On September 9, 2018, **Dr. Azar travelled to Asahi General Hospital in Japan to provide training** to third and fourth year medical residents to help increase their understanding of adult primary immunodeficiency, its impact, its diagnosis and possible treatments. Dr. Azar's global education calendar can be found below.

September 2018	Japan	<i>Asahi General Hospital</i>
November 2018	Seattle, WA	<i>American College of Allergy and Asthma</i>
December 2018	Tampa, FL	<i>University of South Florida</i>

IN THE NEWS

On October 8, 2018, Dr. Azar was officially named the **Tara Sakraida Parker and Richard Parker Center for Innovative Medicine (CIM) Scholar** at the Center for Innovative Medicine Annual Meeting.

Dr. Azar and one of his patients were recently **featured in a New York Times article** about complex illness.

Read the article → <https://nyti.ms/2CVirRo>

BreakOut

Latest Research: New Study Findings



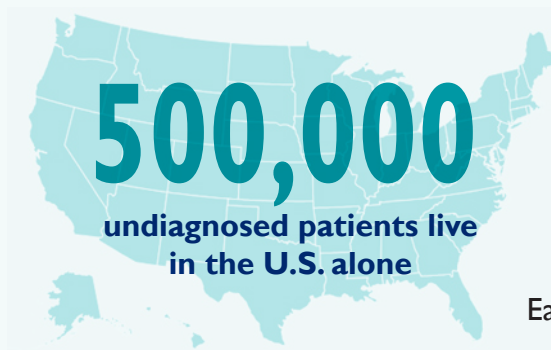
Dr. Azar and his research fellow recently completed a study whose findings will be presented at the national ACAA conference in November 2018. **Currently, there is no specific therapy for patients hospitalized with acute severe respiratory viral infections (RSV).** The treatment is supportive only and mortality rates are high (estimated at 41% in some published studies). The study evaluates the effectiveness of intravenous immunoglobulin (IVIG) therapy for hospitalized patients with acute respiratory infections and seeks to evaluate further potential treatment options. In a total of 80 patients and 99 hospital admissions, RSV was identified in 41.4% of the cases, rhinovirus in 23.2%, parainfluenza in 18.2%, and metapneumovirus in 17.2%. 94% of the patients had a secondary immunodeficiency most commonly due to solid organ transplants, bone marrow transplants, or chemotherapy. 4% had a primary immunodeficiency, and 2% were not immunodeficient. The average dose of IVIG received was 514 mg/kg and the average length of stay was 11.7 days. Clinical improvement was noted in 82.8% of patients. 9.1% developed superinfections that needed additional antibiotic coverage, 3.0% were readmitted for additional doses of IVIG, 4.1% died from complications and only 1.0% developed toxicities such as increased LFTs.

This is the largest case series exploring the use and outcomes of IVIG therapy for severe viral respiratory infections. This data will serve as a cornerstone for future evaluation of the effectiveness of IVIG in respiratory infections, both in retrospective and prospective controlled studies.

FAST FACTS



70% - 90%
 of persons living with
 primary immunodeficiency
 remain undiagnosed



Early diagnosis and treatment could
 save patients

\$100,000
 each year by reducing infections,
 hospitalization, antibiotic use,
 COPD exacerbations and time off
 from work