

*Department of Medicine
Lewis Katz School of Medicine at Temple University*



*2016 Annual Fellows, Residents and Medical Students
Research Symposium*

*Sol Sherry Awards for
Excellence in Research*

Wednesday, June 1, 2016

*Medical Education and Research Building
Luo Commons*

The Fellows and Residents Research Forum was initiated over 35 years ago to provide the Fellows and the Residents in the Department of Medicine with an opportunity to present their research effort. The Forum is a reflection of the ongoing research activities in the Department, and a year-end summation of the projects carried out by the Fellows and Residents.

Dedication

Dr. Sol Sherry 1916-1993



Sol Sherry, M.D., joined Temple University School of Medicine as professor and chairman of the Department of Medicine in 1968. In 1970, Dr. Sherry founded and served as director of the University's Specialized Center for Thrombosis Research, the largest of its kind in the United States, which was later named in his honor. He served as dean of the School of Medicine from 1984-86. He was a recipient of an honorary doctor of science degree, the University's first Distinguished Professor and was honored with the establishment of the Sol Sherry Chair in Medicine.

For his contributions to medical research, teaching and patient care, Dr. Sherry was the recipient of other numerous awards and honors. He was Master of the American College of Physicians and The John Phillips Memorial Medalist of the American College of Physicians; a Fellow of the Royal College of Physicians (London), and recipient of the Robert P. Grant Medal of the International Society on Thrombosis and Hemostasis--a society which he founded in 1977. Dr. Sherry also received awards from the American Heart Association, the Philadelphia County Medical Society, the Texas Heart Institute and the Swedish Society of Cardiology.

Distinguished Scientist Award and Lecture – 2016

“Know Thyself -- Understanding Tolerance and Autoimmunity”

**Philip L. Cohen, M.D.
Chief, Section of Rheumatology
Professor of Medicine
Lewis Katz School of Medicine at Temple University**



Dr. Cohen is a native of the Bronx, New York, where he attended public schools and The Bronx High School of Science. He graduated from The City College of New York in 1968 and went on to the Yale University School of Medicine, where he performed thesis research on regulatory T cells under Dr. Richard Gershon's direction.

After medical internship at Columbia-Presbyterian Medical Center, he became a Commissioned Officer in the U.S. Public Health Service in the Laboratory of Immunology, NIAID. He contributed to the understanding of early T-cell development and B-cell activation in Donald Mosier's laboratory. After completing his residency in New York, he became a Rheumatology fellow at the University of Texas Southwestern Medical Center in Dallas, where he investigated B-cell activation in autoimmune disease with the mentorship of Drs. Morris Ziff and Ellen Vitetta.

In 1979, Dr. Cohen established his laboratory at the University of North Carolina at Chapel Hill. His laboratory investigated the role of T cells and immunogen in anti-nuclear antibody responses, defined their immunoglobulin gene usage, demonstrated their MHC specificity, investigated the role of the microbiome, and defined a role for stochastic interactions in autoantibody production. He showed that rheumatoid arthritis synovium produces interleukin-1 and its inhibitor. He defined a role for apoptosis both in maintaining tolerance – through CD95 and its ligand – and in providing immunogenic nuclear debris for autoantibody production. Many of these studies were performed in collaboration with long-term colleague Dr. Robert Eisenberg.

Dr. Cohen relocated to the University of Pennsylvania in 1999. Here the major focus of his laboratory was in clearance of apoptotic cells. He discovered a key role for the mer tyrosine kinase in regulating immune responses as well as in phagocytosing apoptotic debris. He developed an interest in Sjögren's syndrome and directed a clinical trial of anti-CD20 (rituximab) in this illness.

At Temple University, where he became Chief of Rheumatology in 2008, he continued studies of the mer tyrosine kinase, defining its role in human macrophage differentiation and showing abnormalities in its ligands and in its soluble form in systemic lupus. Most recently, Dr. Cohen and colleague Wen-Hai Shao have discovered that many SLE patients have prion-like aggregation of the MAVS signaling protein that is key in the RIG-I pathway of type I interferon production, providing a possible explanation of the excessive type I interferon thought important in lupus pathogenesis.

Dr. Cohen has published 188 papers in peer-reviewed journals, along with many reviews and textbook chapters. He has served on many study sections and review groups, and was a member of the Board of Scientific Counselors of NIAMS. He served as chairman of the Arthritis Foundation's Medical and Scientific Council, and was designated a "Hero" of the Arthritis Foundation. He was named Master of the American College of Rheumatology, is a Kirkland Scholar in Lupus Research, and received the Lady Colyton Award for Excellence in Autoimmunity Research. He received the Central Carolina Bank Teaching Award and is a member of the American Society of Clinical Investigation and the College of Physicians of Philadelphia.

Acknowledgement of Funding

The Lewis Katz School of Medicine at Temple University Department of Medicine gratefully acknowledges and thanks the following corporate exhibitors for their generous support of our program this year:

Platinum Level:



Gold Level:



Fellows, Residents and Medical Students Research Symposium
Wednesday, June 1, 2016
Medical Education and Research Building

12:00 – 1:45 PM Luncheon, Poster Viewing and Poster Discussions (Luo Commons)

2:00 – 5:30 PM Oral Presentations
Fellows – Room 317
Residents – Room 319

5:45 PM Distinguished Scientist Award and Lecture – 2016 (Room 317)

“Know Thyself -- Understanding Tolerance and Autoimmunity”
Philip L. Cohen, MD
Chief, Section of Rheumatology
Professor of Medicine
Lewis Katz School of Medicine at Temple University

6:15 PM Presentation of Awards to Fellows and Residents (Room 317)

Reception (Luo Commons)

Poster Presentations – Fellows

Chair: Marissa Blum, MD

Judges: Drs. Marissa Blum, Iris Lee, Daniel Rubin

Shuchie Jain, DO (Endocrinology) <i>A Rare Cause of Diabetes Mellitus: Mitochondrial Diabetes</i>	Poster Board #1 Abstract #20
Jonathan Galli MD (Pulmonary) <i>Pirfenidone and Nintedanib for Pulmonary Fibrosis in a Large Clinical Practice: Tolerability and Adverse Drug Reactions</i>	Poster Board #2 Abstract #13
King Soon Goh MB BCh BAO (Rheumatology) <i>Eosinophilic Granulomatous Polyangiitis Presenting as Sick Sinus Syndrome</i>	Poster Board #3 Abstract #16
Swapnil Khare, MD (Endocrinology) <i>Unusual Case of Neurosarcooidosis with Hypercalcemia and Anterior Pituitary Dysfunction</i>	Poster Board #4 Abstract #25
Matthew Gordon, MD (Pulmonary) <i>Ultrasound Assessment of Healthy Diaphragm Function: Effects of Position and Inspiratory Resistance</i>	Poster Board #5 Abstract #18
Grace Shin, MD (Gastroenterology) <i>Avoiding Allergens Associated with Food-Related Type 4 Hypersensitivity Reactions Improves Symptoms of Irritable Bowel Syndrome</i>	Poster Board #6 Abstract #53
Sumangala Vasudevan, MD (Endocrinology) <i>FDG PET Positive Pituitary Macroadenoma in a Patient with GIST</i>	Poster Board #7 Abstract #56
Shikha Rathi, MD (Rheumatology) <i>Impact of Depression on SLE Flare</i>	Poster Board #8 Abstract #45
Lavanya Viswanathan, MD (Endocrinology) <i>Submandibular Mass: To be or not to be Ectopic Thyroid Tissue</i>	Poster Board #9 Abstract #57

Poster Presentations – Residents

Chair: Marissa Blum, MD

*Judges: Drs. Wissam Chatila, Adam Ehrlich, Avrum Gillespie,
Won Han, Sharon Herring, Anuradha Paranjape*

Arslan Mirza M.D (Cardiology)

*43 Year Old Female with Spontaneous Coronary Artery Dissection;
Challenges with Revascularization and Role of Optic Coherence
Tomography in Identification of Left Main Involvement*

**Poster Board #10
Abstract #35**

Arslan Mirza MD & Jaspreet Suri, MD (Cardiology)

*ST-Elevation Myocardial Infarction and Acute Limb Ischemia in
a Young Female: An Arterial Conundrum*

**Poster Board #11
Abstract #37**

Zachariah Dorey-Stein, MD (Gastroenterology/Hepatology)

*The utility of FibroSure (Fibrotest) Biomarkers for the Prediction of
Advanced Fibrosis and Cirrhosis in Patients with Chronic Hepatitis C*

**Poster Board #12
Abstract #9**

Tanya Reznick, MD (Endocrinology)

*Lipodystrophic Diabetes – A Report of Two Cases and Insight
Into Management*

**Poster Board #13
Abstract #48**

Robert Marron, MD (Pulmonary)

*Analyzing the Distribution of Cardiac Comorbidities Across GOLD
Grades and their Effect on Readmission Outcomes*

**Poster Board #14
Abstract #33**

Andrew Meillier, MD (Gastroenterology)

*Difference of Achalasia Subtypes based on Clinical Symptoms,
Radiographic Findings and Stasis Scores*

**Poster Board #15
Abstract #34**

Scott Norberg DO & Ankur Shah MD (General Internal Medicine)

*How Can Primary Care Physicians Improve the Referral System to
Medical Subspecialists?*

**Poster Board #16
Abstract #40**

Julia Judd, DO (Hematology/Oncology)

*Immune Related Adverse Events as a Biomarker in Non-Melanoma
Patients Treated with Programmed Death 1 Inhibitors (PD-1Is)*

**Poster Board #17
Abstract #22**

Zachary Wilmer Reichenbach MD, PhD (Gastroenterology)

*Dronabinol Increases Functional Chest Pain Threshold Without Altering
Anxiety, Depression, or Metabolic Parameters: A 28 Day Study*

**Poster Board #18
Abstract #47**

Catherine Dillane, MD (Cardiology)

Inverted Takotsubo Cardiomyopathy in the Context of Beta Agonist Use

**Poster Board #19
Abstract #6**

Catherine Dillane, MD (Cardiology)

Relationship Between Post Operative Brain Natriuretic Peptide Levels and Health-Related Quality of Life Scores in Patients Undergoing Trans-Catheter Aortic Valve Replacement

**Poster Board #20
Abstract #7**

Catherine Dillane, MD (Cardiology)

Relationship Between Post Operative Cardiac Troponin Elevation and Health-Related Quality of Life Scores in Patients Undergoing Trans-Catheter Aortic Valve Replacement

**Poster Board #21
Abstract #8**

Estefania Oliveros, MD (General Internal Medicine)

Case Report of Meningococemia without Meningitis

**Poster Board #22
Abstract #41**

Swati Rushi, MD (Pulmonary)

Fever of Unclear Origin as the Presenting Manifestation of Pulmonary Sarcoidosis – A Case Report

**Poster Board#23
Abstract #49**

Lauren Freid, MD (Rheumatology)

A Case of Rheumatoid Arthritis Developing after Treatment with Ipilimumab and Nivolumab for Non-Small Cell Lung Cancer

**Poster Board#24
Abstract #12**

Evan C. Klein, MD (Cardiology)

Cardiac Sarcoidosis – A Case Based Review of the Current Literature on Diagnosis, Complications, and Management

**Poster Board #25
Abstract #26**

Poster Presentations – Medical Students

Chair: Marissa Blum, MD

Judges: Drs. Ji Hoon Baang, Serban Constantinescu, Eman Hamad

Alisha Kabadi, BS (Gastroenterology)

Taste and Smell Disturbances in Patients with Gastroparesis and Gastroesophageal Reflux Disease

Poster Board #26

Abstract #23

Brandon Swed, BS (Hematology/Oncology)

Painful Limb, Dusky Toes, Intact Pulses: A Case of Venous Limb Gangrene

Poster Board #27

Abstract #54

Krishma Kumar, BS (Gastroenterology)

Effect of Bifidobacterium infantis 35624 (Align) on the Lactulose Hydrogen Breath Test for Small Intestinal Bacterial Overgrowth

Poster Board #28

Abstract #27

Marie Baumeister BS (Rheumatology)

Evaluation of Patient Reported Respiratory Symptoms and 6-Minute Walk Test as Clinical Screening Tools for Rheumatoid Arthritis Associated Lung Disease

Poster Board #29

Abstract #2

Taha Mur, BS (Gastroenterology/Hepatology)

Safety and Efficacy of Hepatitis C Treatment in Patients with HCV/HBV Coinfection and in Patients with HCV/HIV/HBV Coinfections using Interferon Free Regimens

Poster Board #30

Abstract #39

Oral Presentations – Fellows

Chair: A. Koneti Rao, MD

*Judges: Drs. Peter Axelrod, Roberto Caricchio,
Dan Edmundowicz, David Essex, Kevin Williams*

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|----------------|--|---------------------|
| 2:00 PM | King Soon Goh, MB BCh BAO (Rheumatology)
<i>An Analysis of Disease Progression in Patients with SLE Seen at the Temple Lupus Clinic</i> | Abstract #17 |
| 2:15 PM | Mohammed Saadi, MD (Gastroenterology)
<i>Intrapyloric Injection of Botulinum Toxin for Gastroparesis: Reassessing Efficacy with Utilization of EndoFLIP Pyloric Measurements</i> | Abstract #50 |
| 2:30 PM | Devin Weber, MD
(Infectious Diseases/Hematology/Oncology)
<i>Burkholderia cepacia Bacteremias in a Bone Marrow Transplant Unit Associated with Contaminated Water Baths Used for Stem Cell Thawing</i> | Abstract #58 |
| 2:45 PM | Shikha Rathi, MD (Rheumatology)
<i>Rheumatology Narcotic Contract</i> | Abstract #46 |
| 3:00 PM | Zubair Malik MD (Gastroenterology)
<i>Bravo pH Monitoring After Per Oral Endoscopic Myotomy (POEM) Overestimates True Acid Reflux</i> | Abstract #32 |
| 3:15 PM | Patrick Mulhall, MD (Pulmonary)
<i>Response of Outpatient Chronic Obstructive Pulmonary Disease Patients Respiratory Symptoms to Administration of Prednisone Therapy</i> | Abstract #38 |
| 3:30 PM | Ian G. Sheffer, MD, MBe (Infectious Diseases)
<i>Effectiveness of Brief Health Literacy Education Interventions for Medical Residents</i> | Abstract #52 |
| 3:45 PM | Kamel Hatahet, MD (Nephrology)
<i>Successful Treatment of Hepatitis C Virus Infection in Kidney Transplant Recipients is Associated With Proteinuria Reduction</i> | Abstract #19 |
| 4:00 PM | Michael J. Lavery, MD (Rheumatology/Dermatology)
<i>Clinical Aspects of Chronic Pruritus and Impact on Quality of Life in Patients with Scleroderma</i> | Abstract #30 |

- 4:15 PM** **Gregory Bernstein, MD (Gastroenterology)** **Abstract #3**
Prescriber Practice Patterns in Non-compliant Patients with Inflammatory Bowel Disease
- 4:30 PM** **Casey Godshall, MD (Infectious Diseases)** **Abstract #15**
High Rate of Undiagnosed Hepatitis C Viremia in an Inner City Hospital
- 4:45 PM** **Shuchie Jain, DO (Endocrinology)** **Abstract #21**
Insulin Stimulates a Burst in Intracellular Hydrogen Peroxide that is Short-lived, Highly Localized within the Cell, and Essential for Normal Balanced Insulin Action
- 5:00 PM** **Carlyn Tan, MD (Hematology/Oncology)** **Abstract #55**
Comparing Cyclophosphamide with G-CSF and Plerixafor with G-CSF as Stem Cell Mobilization (SCM) Regimens for Multiple Myeloma (MM) Patients: An Evaluation of Mobilization Efficacy and Toxicity
- 5:15 PM** **Antarpreet Kaur, MBBS (Pulmonary)** **Abstract #24**
Peripheral Blood Eosinophilia as a Marker of Exacerbations in Hospitalized Chronic Obstructive Pulmonary Disease Patients

Oral Presentations – Residents

Chair: Henry Parkman, MD

*Judges: Drs. Michael Bromberg, Crystal Gadegbeku,
James Mamary, Ajay Rao, Ellen Tedaldi*

2:00 PM	Andrew Peters, MD (Cardiology) <i>Identifying Prognostic Factors in Peripartium Cardiomyopathy: A 22 Year Retrospective Analysis of an Urban Population in the US</i>	Abstract #43
2:15 PM	Lauren Freid, MD (Rheumatology) <i>Implementation of Combined Rheumatology and Nephrology Clinics: An Evaluation of Changes in Outcomes in Patients with Lupus Nephritis</i>	Abstract #11
2:30 PM	Aloknath Pandya MD (Pulmonary) <i>Adverse Effects and Drug Discontinuation Rates of Antifibrotic Drug Therapy in Patients with FVC <50 %</i>	Abstract #42
2:45 PM	Osama El Shamy, MD (Nephrology) <i>Characteristics Associated with Consent for Genetic Research in Kidney Disease: Observations from the Michigan Kidney Translational Core Center</i>	Abstract #10
3:00 PM	Steven Back, MD, PhD (Cardiology) <i>Acute Heart Failure Secondary to Cardiac Amyloidosis: A Case Series</i>	Abstract #1
3:15 PM	Sui Kwong Li, MD (Infectious Diseases) <i>Ceftaroline in the Treatment of Methicillin-Resistant Staphylococcus Aureus Bloodstream Infections</i>	Abstract #31
3:30 PM	Amanda J. Podolski, MD (Hematology/Oncology) <i>Telomere Length as a Predictor of Overall Survival in Colorectal Cancer</i>	Abstract #44
3:45 PM	James Langworthy MD (Gastroenterology) <i>Abnormal Esophageal Manometry and pH Impedance is Independent of Lung Disease Etiology in Pre-lung Transplant Patients</i>	Abstract #29
4:00 PM	Ryan Schmidt, MD (General Internal Medicine) <i>Opiate Antagonist Therapy in High Risk Patients on Chronic Narcotics</i>	Abstract #51

4:15 PM	Arslan Mirza M.D (Cardiology) <i>Single Center Experience with Successful Left Ventricular Assist Device Explantation Following Myocardial Recovery</i>	Abstract #36
4:30 PM	Michele Glodowski (Endocrinology) <i>Thyroid Storm Coupled with Severe Postpartum Cardiomyopathy: Successful Management & Insight into Pathogenesis</i>	Abstract #14
4:45 PM	Alexandra Lane, MD (General Internal Medicine) <i>Preventing Readmission and Emergency Department Encounters with a Clinic Team-based Approach</i>	Abstract #28
5:00 PM	Rajiv Bhuta, MD (Gastroenterology) <i>Impedance Bolus Transit Analysis of Viscous Swallows with Apple Sauce to Enhance the Detection of Abnormalities Using High Resolution Esophageal Manometry with Impedance (HREMI)</i>	Abstract #4
5:15 PM	Catherine Dillane, MD (Cardiology) <i>A Shock Team Improves Survival in Cardiogenic Shock by Decreasing Time to Intervention</i>	Abstract #5

Acute Heart Failure Secondary to Cardiac Amyloidosis: A Case Series

Back S, Raza F, Alvarez R

Cardiac amyloidosis is a disease characterized by the abnormal deposition of protein in the heart, altering the architecture of myocardium and resulting in cardiomyopathy. Patients commonly present with signs of right-sided heart failure. The most common types of cardiac amyloid deposition are immunoglobulin light chain (AL) as well as wild-type (wt) and mutant transthyretin (ATTR). Cardiac involvement occurs in about 50% of patient with AL amyloidosis. Some reports indicate that 4% of African Americans possess a mutant variant of ATTR. The purpose of this study is to analyze three cases of acute heart failure due to amyloidosis. Cases of cardiac amyloidosis were identified in our heart-failure clinic and analyzed as a part of a case-series. The patients with cardiac amyloidosis included one case of AL, wt-ATTR and Val122Ile mutant ATTR. Each patient presented with symptoms of shortness of breath or volume overload. The diagnosis was made by endomyocardial biopsy and immunohistochemistry. Each patient was treated symptomatically with heart rate and afterload reduction as well as a diuretic. Definitive treatment strategy varied with respect to the patient's subtype of cardiac amyloidosis. The management of acute heart failure due to cardiac amyloidosis requires symptomatic treatment of heart failure as well as elimination of abnormal protein production. Acute heart failure management is similar regardless of the amyloid subtype, however, long term treatment and potential cure is dependent upon the source of amyloid production. Patients with AL amyloid require chemotherapy and stem cell transplantation while patients with ATTR amyloid are treated with liver transplantation.

Evaluation of Patient Reported Respiratory Symptoms and 6-Minute Walk Test as Clinical Screening Tools for Rheumatoid Arthritis Associated Lung Disease

Baumeister M, Patel N, Blum M

Background/Purpose: Rheumatoid arthritis (RA) is a systemic inflammatory disease that may involve the pulmonary system in 10-20% of patients. There is a lack of guidance related to screening of RA patients for interstitial lung disease (ILD). We evaluated the use of patient reported outcomes and 6 minute walk testing in the decision making process related to screening RA patients for ILD.

Methods: Patients with a diagnosis of RA and patients with RA and known ILD were recruited. Patients completed the following: Dyspnea-12 (D-12) questionnaire, cough/sputum scale, and Borg scale before and after a 6-minute walk test (6MWT). RA activity at time of visit was graded mild (1), moderate (2), or severe (3). Data about sex, age, ethnicity, and smoking status were also recorded.

Results: Among 264 patients meeting 2012 ACR diagnostic criteria for RA, 50 patients were recruited. Of the 50 patients, 4 had known RA-ILD. Females comprised 89% of the RA patients, whereas 50% of the RA-ILD patients were female. The mean age of the RA patients was 60 and the mean age of the RA-ILD patients was 69. 41% of the RA patients were smokers whereas 75% of the RA-ILD patients were smokers. RA-ILD patients had greater 6MWT and greater pre and post Borg dyspnea and fatigue scores than the RA controls.

Conclusions: These findings suggest a role for the 6MWT in the initial screening of RA patients for RA-ILD but do need to be evaluated in a larger cohort of patients.

**Prescriber Practice Patterns in Non-compliant Patients with
Inflammatory Bowel Disease**

Bernstein G, FriedenberG F, Ehrlich A

Background: Medication non-compliance in inflammatory bowel disease (IBD) is common and associated with suboptimal care. Since many IBD therapies have inherent risks following provider recommendations are important. It is not known how physicians alter treatment strategies in non-compliant IBD patients.

Methods: An email was distributed to American College of Gastroenterology members with an anonymous survey including demographic information, views on medication adherence, and questions pertaining to how a hypothetical non-compliant IBD patient might be managed.

Results: The 207 respondents were primarily male (74%) and in private practice (56%). Most have practiced independently for over 20 years (34%), followed by 1-5 years of independent practice (30%). Many prescribers (44%) had a patient population with only 1-10% IBD patients, while others (33%) had up to 25% IBD patients or more (23%).

Respondents considered medication adherence important, however the majority (75%) never use standardized adherence assessments. Common concerns physicians have in non-compliant patients include disease progression (94.7%) and development of antibodies to biologics (77.5%). In non-compliant IBD patients, most physicians continue the current therapy regardless of therapy type. Still, 76% of prescribers have altered treatment plans for non-compliant IBD patients including the use of IV infusions over home self-injections (84.6%), avoiding medications that require monitoring (65.1%) and avoiding polypharmacy (56.2%). Prescribing patterns were similar despite duration of physician practice or percentage of IBD patients.

Conclusion: Non-compliant patients frequently have altered treatment plans. Often, physicians choose regimens that deviate from the standard of care and may lead to suboptimal outcomes. Further study is warranted.

Impedance Bolus Transit Analysis of Viscous Swallows with Apple Sauce to Enhance the Detection of Abnormalities Using High Resolution Esophageal Manometry with Impedance (HREMI)

Saadi M, Bhuta RV, Malik ZA, Schey R, Parkman HP

Many patients with dysphagia have normal high resolution esophageal manometry (HREM). Addition of impedance and solid food may detect additional abnormalities. Multiple rapid swallows (MRS) has also been implemented to detect abnormalities in esophageal motility. The use of viscous swallows and impedance to analyze bolus transit during MRS have not been evaluated.

Aims: 1) To assess the value of viscous boluses (apple sauce) in detecting motor and/or transit abnormalities. 2) Compare MRS findings using apple sauce to saline.

Methods: Patients with esophageal symptoms underwent HREMI: 10 supine 5 ml saline swallows, 5 upright saline swallows followed by MRS (5 rapid swallows of saline) and 5 upright swallows of 5 mL apple sauce followed by MRS (5 rapid swallows of apple sauce). Bolus clearance for the 5 swallows was assessed with impedance.

Results: 16 patients were evaluated. Using conventional 10 supine wet swallows, 4/16 (25%) patients had hypercontractile esophagus, 6/16 (37%) weak peristalsis with small defects and 6/16 (37%) had normal esophageal motility. 11/16 (68%) of patients had incomplete clearance when apple sauce was used compared to only 4/16 (25%) clearance with saline swallows.

Conclusions: Viscous swallows using apple sauce showed 24% more impedance transit abnormalities with single swallows and 43% more abnormalities during MRS compared to saline in patients with esophageal symptoms. Thus, the addition of a viscous substance such as apple sauce for HREMI increases detection of esophageal function abnormalities.

**A Shock Team Improves Survival in Cardiogenic Shock
By Decreasing Time to Intervention**

Dillane C, Bove A, Cohen H, Bashir R, O'Murchu B, O'Neill B, Gomez-Abraham J, Shiose A, Schwartz D, Dries D, Punnoose L, Toyoda Y, Alvarez R, Hamad E

Introduction: Cardiogenic shock (CS) is a state of end-organ hypo-perfusion due to cardiac failure. A Shock Team was formed at our institution to rapidly assess patients in CS and intervene if appropriate. The team consists of a cardiothoracic surgeon, interventional cardiologist and heart failure physician. When CS is identified, a Shock Code is activated. The team assemble at bedside.

Method: We performed a retrospective analysis on 14 Shock Codes between 2014-2015.

Results: CS was confirmed in 13 patients. The average cardiac index was 1.67 L/min/m². CS occurred due to ST elevation myocardial infarctions (n=7), acute decompensated heart failure (ADHF) (n=5) and LAD dissection (n=1). Six shock codes occurred after cardiac arrest. Seven patients had a right heart catheterization (RHC) within 90 minutes of the shock code, median 24 minutes. All patients had a RHC.

8 patients received mechanical circulatory support in a median of 67 minutes including veno-arterial extracorporeal membrane oxygenation (n=3), intra aortic balloon pump (n=3) and tandem heart (n=2). Patients who received a RHC within 90 minutes and subsequent interventions including temporary mechanical circulatory support and/or inotropes had a 71% 30-day survival versus 16 % for those with RHC at > 90 minutes. (p = 0.064).

46% of all patients survived and were alive at 8 months. Eight patients were evaluated for advanced therapies; heart transplantation and ventricular assist devices (VAD). One patient received a heart transplant.

Conclusion: A Shock Team can potentially improve short and long term survival in cardiogenic shock patients by decreasing time to intervention and initiating evaluation for advanced therapies.

Inverted Takotsubo Cardiomyopathy in the Context of Beta Agonist Use

Dillane C, Peters A, Raza F, Schwartz D

Introduction: Takotsubo cardiomyopathy (TTC) is classically a transient dysfunction of the left ventricular (LV) apical and/or mid-ventricular segments. TTC is now sub-classified based on LV involvement. Inverted takotsubo describes basal and mid-ventricular segment hypo-kinesis with preservation of the apical segment. Chronic obstructive pulmonary disease (COPD) and acute respiratory failure are associated with TTC.

Clinical Vignette: A 64-year-old man with GOLD Stage IV COPD was transferred to the coronary care unit (CCU) with cardiogenic shock in the context of acute respiratory failure. Prior to presentation, he noted worsening dyspnea requiring albuterol nebulizer. On initial presentation, oxygen saturation was 80% on 4L nasal cannula and labs included a venous blood gas; pH 7.24, pCO₂ 65mmHg, cardiac troponin 0.93ng/mL and brain natriuretic peptide 1896pg/m. An electrocardiogram showed ST-segment elevation in leads II, III, and aVF. Transthoracic echocardiogram showed an ejection fraction of 5-10%, global LV hypo-kinesis with apical sparing. Coronary angiogram showed non-obstructive coronary artery disease. Cardiac index was 2.17 L/min/m² (while receiving inotropes). An intra-aortic balloon pump was placed. He was intubated for respiratory distress. A CT pulmonary angiogram demonstrated pneumonia but no embolus. He was treated for a COPD exacerbation. A repeat echocardiogram in 3 days showed improved LV function. He failed to wean from mechanical ventilation due to underlying pulmonary disease and elected to withdraw care.

Discussion: Typical TTC with β -agonist use has been reported. Studies show a greater density of B₂ adrenergic receptors at the LV apex. This case is unusual as the patient developed inverted TTC, with global LV hypo-kinesis except for the apex.

Relationship between Post Operative Brain Natriuretic Peptide Levels and Health-Related Quality of Life Scores in Patients Undergoing Trans-Catheter Aortic Valve Replacement

Dillane C, Lakhter V, Barlow J, Back S, O'Murchu B, Cohen H, Wheatley G, Patil P, Keane M, Boova R, Toyoda Y, O'Neill B

Background: When measured by the Kansas City Cardiomyopathy Questionnaire (KCCQ), trans-catheter aortic valve replacement (TAVR) improves health-related quality of life compared to standard therapy in patients with inoperable severe aortic stenosis. The release of brain natriuretic peptide (BNP) post TAVR is associated with increased mortality. The relationship between BNP release and KCCQ scores in TAVR is unclear.

Methods: We performed a retrospective analysis on 44 patients who underwent TAVR in our institution between December 2013 and July 2015. The KCCQ was completed at baseline and at 1-month post-TAVR to assess health-related quality of life (range, 0–100; higher better). BNP was assessed at baseline and at 1-month.

Results: BNP levels were available in 31 patients and KCCQ scores were available in 39. Pre-TAVR BNP levels were elevated and KCCQ scores were depressed (mean BNP 784; mean KCCQ 34). At one month post-TAVR, the mean BNP significantly decreased by 289 (mean BNP 495, $p = 0.0085$) and the mean KCCQ scores significantly increased by 17 (mean KCCQ 50.4, $p = < 0.00001$). Most patients had an improvement in BNP levels (68%) and an increase in KCCQ scores (87%). There was no significant correlation between a reduction in BNP levels and an increase in KCCQ scores (Figure 1).

Conclusion: Our retrospective analysis demonstrates a significant reduction in mean BNP levels and a significant increase in mean KCCQ scores in patients assessed 30-days post TAVR. However, an increase in health-related quality of life does not clearly correlate with a reduction in BNP levels.

Relationship between Post-Operative Cardiac Troponin Elevation and Health-Related Quality of Life Scores in Patients Undergoing Trans-Catheter Aortic Valve Replacement

Dillane C, Lakhter V, Back S, Barlow J, O'Murchu B, Cohen H, Wheatley G, Patil P, Keane M, Boova R, Toyoda Y, O'Neill B

Background: When measured by the Kansas City Cardiomyopathy Questionnaire (KCCQ), trans-catheter aortic valve replacement (TAVR) improves quality of life compared to standard therapy in patients with inoperable severe aortic stenosis. Elevated cardiac biomarkers post TAVR is associated with increased mortality. The relationship between cardiac troponin I (CTNI) release and KCCQ scores in TAVR is unclear.

Method: We performed a retrospective analysis on 44 patients who underwent TAVR between December 2013 and July 2015. The KCCQ was completed at baseline and at 1-month post-TAVR to assess health-related quality of life (range, 0–100; higher better). Peak CTNI within 24-hours of TAVR was recorded.

Results: Post-TAVR CTNI was available in 41 patients and KCCQ scores in 39. No post-procedural electrocardiogram indicated myocardial ischemia and 64% had known coronary artery disease. All 41 patients demonstrated elevated CTNI post TAVR. Twenty-three patients had a peak CTNI of ≤ 1.0 (Group 1; mean CTNI 0.64). Eighteen patients had a peak CTNI of >1 (Group 2; mean CTNI 2.3). At one month post-TAVR, mean KCCQ scores significantly increased by 17 (mean KCCQ 50.4, $p = < 0.00001$). Group 1 had a mean increase in KCCQ score of 14 points (correlation coefficient -0.19, $P=0.39$) while Group 2 had a mean increase of 21 points (correlation coefficient 0.2; $p= 0.49$). The correlation between peak CTNI and KCCQ did not reach statistical significance ($p=0.066$).

Conclusions: There may be a correlation between post procedural CTNI and quality of life at 30-days post TAVR. However, the current data suggest that a correlation, if it exists, is inverse.

The Utility of Fibrosure (Fibrotest) Biomarkers for the Prediction of Advanced Fibrosis and Cirrhosis in Patients with Chronic Hepatitis C

Mur T, Aziz S, Dorey-Stein Z, Alsabbagh E

Introduction: Accurate determination of liver fibrosis stage is essential for predicting prognosis and appropriate follow up for patients with Chronic HCV even after the eradication of the underlying HCV. Liver biopsies (LB) are still considered the gold standard for staging of fibrosis. Less invasive measurements proposed include serum biomarkers. We aim to test the utility of Fibrosure to stage fibrosis in HCV patients and compare it to LB.

Methods: We utilized data from LB done over one year period for patients with chronic HCV who also had fibrosure checked within a calendar year. All biopsies were percutaneous, using a 16G BioPince core biopsy needle. When a range of fibrosis stage was given on the biopsy, the higher stage was used.

Results: A total of 75 LB were included. Median biopsy length was 2.3 cm. Only 5 biopsies (6.6%) were ≤ 1.5 cm. 39 patients had high fibrosure results consistent with F4, only 5 of them had evidence of cirrhosis based on the LB translated into a PPV of 12.8% (certainty of presence of cirrhosis) and the NPV was 97.2%. When dividing patients into patients with advanced fibrosis (F3, F4) vs early stage fibrosis (F0, F1, F2), 37% had advanced fibrosis on LB and the PPV became 40% while NPV was 69%. No significant correlation was found between Fibrosure classifications and findings of advanced fibrosis on LB ($p=0.5$).

Conclusion: In patients with chronic HCV, Fibrosure wasn't a reliable marker to diagnose cirrhosis. Fibrosure can be used to rule out advanced fibrosis and decrease the need for biopsies.

**Characteristics Associated with Consent for Genetic Research in Kidney Disease:
Observations from the Michigan Kidney Translational Core Center**

Troost JP, Hawkins J, Jenkins DR, Gipson DS, Kretzler M, El Shamy O, Bellovich K, Perumal K, Bhat Z, Massengill S, Steigerwalt S, Brosius F, Gadegbeku CA

Background and objectives: The goal of this study was to determine whether demographic, clinical or socio-economic factors are associated with willingness to consent to future genetic research among participants in a prospective kidney disease cohort and biobank network.

Design, setting, participants, & measurements: The Clinical Phenotyping Resource and Biobank Core (C-PROBE) of the O'Brien Michigan Kidney Translational Core Center enrolled 1,232 patients with CKD from 2009 to 2015 across seven sites in the United States to facilitate nephrology research. Patients were asked at annual study visits for consent to provide DNA samples for future genetic studies. We reported consent rates at the initial study visit, and longitudinally. We compared characteristics of participants who consented to and refused participation in genetic studies. Multinomial logistic regression analyses were used to identify predictors of consent.

Results: Ninety-four percent of C-PROBE participants agreed to consent to genetic studies at their initial study visit. Among patients with at least two study visits, 89% consented at all visits, 2% consistently declined, and 9% changed consent status over time. African American participants were 3-fold more likely to initially decline consent ($p < 0.001$), but this difference by race disappeared when we assessed likelihood to consent across multiple study visits.

Conclusions: Overall, genetic consent rates for kidney research within the C-PROBE cohort are high. The high enthusiasm for genetic research among kidney disease patients in the precision medicine era is encouraging.

Implementation of Combined Rheumatology and Nephrology Clinics: An Evaluation of Changes in Outcomes in Patients with Lupus Nephritis

Zavitsanos A, Freid L, Goh KS, Rathi S, Lee I, Caricchio R

Introduction: Systemic lupus erythematosus is a multi-organ disease often requiring co-management of several subspecialties, particularly nephrology. Clinically evident renal disease is commonly found in up to 50% of lupus patients. Multi-disciplinary approach to the management of lupus nephritis patients can expedite renal biopsies and treatment leading to less end organ damage.

Objective: To evaluate outcomes in lupus nephritis patients 1 year before and 1 year after initiation of joint nephrology and rheumatology lupus clinic.

Methods: Using Temple University Hospital electronic health records, we extracted various data including SLEDAI, SLICC, anti-dsDNA levels, C3, C4, urine protein/creatinine, creatinine levels, vitals, number of office visits, and number of renal biopsies from lupus nephritis patients following in the combined nephrology and rheumatology clinic. We then compared these data 1 year prior to initiation of the clinic and 1 year afterward.

Preliminary Results: Longitudinal data from 12 lupus nephritis patients following in the combined clinic was evaluated. Average systolic blood pressures were 128.7mmHg and 136.13mmHg one year before and after start of the clinic. Average urine protein/creatinine ratios from 8/12 patients were 595.5mg/g and 1394.51mg/g one year before and after respectively. Average SLEDAI scores decreased from 5.93 to 3.61 one year before and after initiation of the clinic.

Preliminary Conclusion: Preliminary results demonstrate that although average systolic blood pressure and urine protein/creatinine appeared to increase 1 year after clinic was started, average SLEDAI scores did improve. More patients and other longitudinal clinical and laboratory variables are currently under investigation.

A Case of Rheumatoid Arthritis Developing after Treatment with Ipilimumab and Nivolumab for Non-Small Cell Lung Cancer

Zavitsanos A, Freid L, Caricchio R

Introduction: The general concept behind cancer immunotherapy involves development of antibodies that target lymphocyte receptors or their ligands in order to enhance endogenous anti-tumor activity. Both CTLA-4 and PD-1 are important molecules in the inhibitory check point pathway of T-lymphocytes. However, it is thought that blocking these inhibitory pathways with agents such as ipilimumab and nivolumab, induce a tolerance break against the tumor but also predisposes the patient to immune-related adverse events (irAEs) (2). We present a case of rheumatoid arthritis developing shortly after treatment with ipilimumab and nivolumab.

Case: Our patient is a 75 year old African American female with a past medical history of hypertension, chronic kidney disease, and stage IV non-small cell lung cancer diagnosed in July 2014. In September 2014, she was started on ipilimumab and nivolumab. She was referred to rheumatology clinic in October 2015 for progressive bilateral wrist and hand pain associated with 2-3 hours of morning stiffness. Physical exam was consistent with synovitis of her bilateral wrists and metacarpal phalangeal (MCP) joints bilaterally. Lab work showed positive ANA 1:1280 nucleolar, negative RF and CCP, and elevated ESR and CRP. Radiographs of the hands and wrists were pertinent for marginal erosions of the wrists and MCPs consistent with rheumatoid arthritis. She was treated with methotrexate 10mg weekly, folic acid 1mg daily, and prednisone 5mg daily with improvement of her symptoms.

Discussion: Immune-related adverse events have been reported in up to 72% of patients receiving ipilimumab. There have been a variety of irAEs reported. However, to our knowledge this is the first reported case of rheumatoid arthritis developing after treatment with ipilimumab and nivolumab documented in the literature.

**Pirfenidone and Nintedanib for Pulmonary Fibrosis in a Large Clinical Practice:
Tolerability and Adverse Drug Reactions**

Galli J, Pandya A, Vega-Olivio M, Dass C, Zhao H, Criner G

Rationale: In clinical practice many patients who receive nintedanib or pirfenidone for treatment of pulmonary fibrosis have significant medical comorbidities or baseline characteristics that would have led to exclusion from clinical trial participation. Little is known about the drug safety and tolerability of nintedanib and pirfenidone in non-clinical trial patients from a large clinical practice.

Methods: We conducted a retrospective, single-center, chart review study on subjects prescribed nintedanib or pirfenidone for treatment of pulmonary fibrosis (any etiology) from September 2014 to February 2016. 186 subjects were included in the study: 129 received pirfenidone and 57 were prescribed nintedanib. The primary outcome for the study was drug discontinuation due to an adverse drug event.

Results: Subjects at baseline had a significant degree of respiratory impairment with a large proportion requiring home oxygen therapy (117/186 or 63%) and a mean diffusion capacity of carbon monoxide (DLCO) % predicted of 36 ± 14 . Drug discontinuation due to an adverse event occurred in 20.9% of subjects on pirfenidone and 26.3% on nintedanib. Adverse events occurring with the highest frequency on pirfenidone were nausea (26.4%), rash/photosensitivity (14.7%), and dyspepsia/gastroesophageal reflux disease (12.4%). On nintedanib therapy diarrhea (52.6%) and nausea (29.8%) were reported most often.

Conclusion: Patients with pulmonary fibrosis treated with nintedanib or pirfenidone in a large clinical practice had drug tolerability and adverse event profiles comparable to subjects enrolled in clinical trials despite having a greater degree of respiratory impairment and a multitude of medical comorbidities.

Thyroid Storm Coupled with Severe Postpartum Cardiomyopathy: Successful Management & Insight Into Pathogenesis

Glodowski M and Vaz CD

We describe the management of postpartum thyroid storm with cardiomyopathy.

25-year-old female with Graves' disease presented to the ED with palpitations. She was 3 weeks postpartum after pregnancy complicated by severe hyperthyroidism due to medication non-adherence. TSI 241 (<140). In the ED she was given Metoprolol and subsequently had a cardiac arrest (PEA) followed by ROSC. Laboratory data revealed TSH 0.04 [0.29 - 5.10 MIU/ML], free T4 5.7 [0.6 - 1.6 NG/DL], and free T3 1,320 [250.0 - 390.0 PG/DL]. Wartofsky score was 50. Echo showed LVEF 30%. Swan-Ganz catheter revealed CI of 1.84. She was treated with PTU (200 mg every 6 hours), hydrocortisone (100 mg IV every 8 hours), Lugol's iodine (10 drops every 8 hours) and Esmolol drip, which was discontinued on the same day due to worsening hypotension and decreased CI. Hemodynamics improved. She was restarted on beta-blocker and transitioned back to Methimazole. On discharge TSH 0.02, free T4 0.5, free T3 210.

The association between hyperthyroidism and high output heart failure is well known, but low output heart failure is rarely seen. High output heart failure itself may lead to low output heart failure. Low output heart failure may also result from the tachycardia associated with hyperthyroidism. Our patient had elements of both postpartum cardiomyopathy and tachycardia-induced cardiomyopathy, compounded by the onset of thyroid storm. While most studies report exacerbation or relapse of Graves' occurs between 2-8 months postpartum, our patient presented at just 3 weeks.

Postpartum patients with Graves' disease should be monitored for thyroid storm, which can exacerbate postpartum cardiomyopathy. Beta-blockers should be used judiciously in thyrotoxicosis associated with low output heart failure.

High Rate of Undiagnosed Hepatitis C Viremia in an Inner City Hospital

Godshall C and Bettiker R

One percent of the US population is infected with HCV; most are unaware of it. We measured the point prevalence of HCV viremia among inpatients to determine the rate of undiagnosed HCV.

We collected data from medical records on 366 inpatients on one weekday in 2014. Plasma for HCV PCR was available for 341 patients. Patients with available sera (296) were prescreened with the lower-cost HCV antibody—those found antibody-negative were not tested by PCR. All data were de-identified prior to testing.

We found 30 patients to be HCV PCR positive (9% of all tested patients). Sixteen of these had a documented history of HCV, suggesting almost half were unaware of their infection. We found no differences between the 14 undiagnosed patients and HCV test-negative patients. Of the 14 with undiagnosed viremia, 12 had prior interactions with the health care system, 9 were over 50 yrs old, and 4 had a h/o illicit drug use. Compared with nonviremic patients, viremics with h/o HCV were statistically more likely to have histories of HIV or drug use, or to be > 50 years old. Of 30 antibody-positive samples paired with plasma, 7 were PCR-negative (spontaneous clearance rate of 23%). There was no record of anyone receiving HCV therapy.

HCV viremia among inpatients in our hospital was 9 times higher than the US population average, with almost half unaware of their diagnosis. Nearly all patients had interactions with the health care system prior to the current hospitalization, which represent missed testing opportunities.

Eosinophilic Granulomatous Polyangiitis Presenting as Sick Sinus Syndrome

Goh KS, Caricchio R

Introduction: The presentation of Eosinophilic Granulomatous Polyangiitis (EGPA) varies among patients, making the diagnosis challenging. We present a case of EGPA presenting as Sick Sinus Syndrome

Case Report: An 88 year old male presented with a 2 week history of general weakness, associated with worsening of dyspnea on exertion. He had long standing uncontrolled asthma and sinusitis for 50 years with a history of persistent eosinophilia. There was a recent deterioration of cardiac function within a year and newly diagnosed right foot drop 2 weeks prior to this admission. Physical examination revealed stigmata of heart failure, right foot drop without active synovitis. There were elevated eosinophil count and inflammatory markers; ANCA antibodies were undetectable. His echocardiogram confirmed reduction of cardiac function within 9 months (EF decreased to 40% from 60%). A myocardial biopsy was performed along with cardiac ablation. The biopsy showed eosinophilic infiltration with focal activity and patchy fibrosis compatible with eosinophilic myocarditis. He received induction therapy with steroid and Cyclophosphamide, followed by maintenance therapy with Azathioprine. He responded to therapy with undetectable eosinophil counts, resolution of the foot droop, normalization of inflammatory markers and partial recover of respiratory and cardiac function.

Discussion: We report one of the oldest patient diagnosed with EGPA despite negative ANCA. Cardiac biopsy was performed without imposing more risks to the patient, as he required cardiac ablation. ANCA is present in more than a third of patient with CSS, however heart disease and fever are associated with negative ANCA, which predict poor prognosis. Early diagnosis and treatment of EGPA may improve the outcome. This patient was treated with a “tailored” induction therapy given the advanced age.

Conclusion: Cardiac involvement in EGPA carries an unfavorable prognosis, especially in an elderly patient and warrant extra consideration in the goals of care and treatment outcome.

**An Analysis of Disease Progression in Patients with SLE
Seen at the Temple Lupus Clinic**

Goh KS, Zavitsanos A, Rathi S, Freid L, Caricchio R

Background: Disease activity and the organ damage are major prognostic factors in SLE. Therefore their prevention should be a priority in SLE.

Objective: The goal here was to trend disease progression and identify the factors that predict organ damage in patients with SLE seen at the Temple Lupus Clinic.

Methods: The activity (SLEDAI) and damage (SLICC) Indexes were “embedded” within the EMR for the Temple Lupus Clinic and collected at each visit from 01/2014 till 12/2015. The indexes were then investigated as a surrogate for disease progression.

Results: 115 patients were identified. Lupus patients demonstrated disease-related damage progression. Overall, the majority of patients was African American (65.2%), and females (93.9%), and with an average age of 44.3 years, moreover 75.6% of patients had government health insurance. 16% of the patients with no initial end organ damage, increased their score within the first year of observation; while 18.3% patients who presented with a damage index of 1 or more progressed toward worse score. The average time to transition to a worse damage score was 8.9 months. Surprisingly the SLEDAI varied greatly and preliminary results indicate that the SLICC/DI did not correlate with the SLEDAI.

Conclusion: The lupus population investigated within the Temple Lupus Clinic appears to have a faster progression toward the organ damage when compared to established cohorts available in the literature. This appears to be particularly true for patients that present initially without organ damage. Factors that influence this pattern are currently investigated, including the impact of low socio-economic status.

**Ultrasound Assessment of Healthy Diaphragm Function:
Effects of Position and Inspiratory Resistance**

Gordon M, Dass C, Liberator C, Criner G

Background: Ultrasonic evaluation of diaphragm function is an emerging field in predicting liberation from mechanical ventilation. Normal values and standardized techniques are lacking. We provide a standardized baseline in healthy individuals and assess the physiologic effects of positional changes and increasing inspiratory resistance on diaphragm function.

Methods: Healthy volunteers were assessed in the supine, 45°, and 90° position at tidal breathing and total lung capacity (TLC). Inspiratory resistance was applied via inspiratory pressure threshold device set at 0, 15, 21, 27, and 33cmH₂O. Diaphragmatic measurements were taken in the right 8th–11th intercostal spaces, mid-clavicular to anterior-axillary line. Thickness was assessed at the zone of apposition, excursion was measured from the posterior 1/3 of the diaphragm. Diaphragmatic thickening was assessed at the zone of apposition as fractional thickening (End-inspiratory thickness – Tidal-thickness)/(Tidal-Thickness).

Results: 7 healthy subjects completed diaphragm measurements. The average age was 32 years, BMI was 25.9kg/m². Image acquisition was 100%.

Diaphragm thickness during tidal breathing was 2.1mm±0.5mm. At TLC, thickness increases to 4.2mm±1.5mm. During TLC, diaphragm thickness and fractional thickening are not statistically changed by alterations in position or resistance.

Diaphragm excursion increases significantly as the patient is reclined from 90° (54mm±8mm) to 45° (50mm±10mm) to supine (43mm±9mm). There is no statistical change in excursion with increasing respiratory resistance.

Conclusions: Diaphragm excursion at TLC measured via ultrasound was dependent on position but not resistive load. In contrast, diaphragm thickness at total lung capacity is independent of position and resistive load.

Successful Treatment of Hepatitis C Virus Infection in Kidney Transplant Recipients is Associated with Proteinuria Reduction

Hatahet K, Rao S, Gillespie A, Ghanta M, Lee I

Kidney transplant recipients (KTRs) with Hepatitis C virus infection (HCV+ve) have inferior patient and allograft survival compared to HCV negative KTRs. There are limited data on the use of novel direct acting antiviral agents (DAA) in KTRs.

We studied virologic response, graft function, proteinuria, and acute rejection in 11 HCV+ve KTRs who completed DAA therapy (Rx); 7M:4F, mean age-59yrs, all deceased donor KT, median time from KT to initiation of DAA-13 months (range 6-124 months). DAA regimens utilized were Ledipasvir /Sofosbuvir (73%), Sofosbuvir / Simeprevir (18%) and Ribavirin/Sofosbuvir (9%). DAA Rxs were well tolerated with the exception of dose modification of ribavirin due to anemia. Majority, 91% (10/11) had sustained virologic response at 12 weeks post DAA Rx. One patient who did not achieve remission was excluded from the analysis.

Serum creatinine (SCr) and spot urine protein /creatinine ratio (Up/c) at 3 and 6 months pre- and post- DAA Rx were analyzed. The mean SCr pre and post DAA Rx was similar (1.4mg/dl). There was a significant reduction in proteinuria, with median U p/c ratio pre-DAA Rx of 0.38mg/g (range 0.05-1.32mg/g) and median U p/c ratio post-DAA Rx of 0.18 mg/g (range 0.05-0.48 mg/g) (p=0.02). There were no episodes of acute rejection associated with DAA Rx.

In conclusion, DAA Rxs were effective, safe, and well tolerated in HCV+ve KTRs. The graft function remained stable during DAA Rx without any episodes of acute rejection. The decrease in proteinuria could represent the amelioration of HCV-related kidney effects and potentially impact graft survival.

A Rare Cause of Diabetes Mellitus: Mitochondrial Diabetes

Jain S, Anolik J

Objective: A rare form of diabetes mellitus (DM) is maternally inherited diabetes and deafness (MIDD) caused by mutations in mitochondrial DNA. The most commonly associated gene mutation is an A to G transversion in 3243 in the mitochondrial DNA-encoded tRNA gene. It is prevalent in 1.5% of the diabetic population worldwide. It is characterized by a defect in insulin secretion and sensorineural hearing.

Results or Case Presentation: A 25-year-old female with medical history of mitochondrial DNA defect presented after an elevated glucose and a subsequent high HgA1C of 11.1%. There was no family history of diabetes mellitus. Her mother, with the same defect, was recently diagnosed with mitochondrial myopathy. Physical exam did not reveal any pertinent positives. She was started on a basal bolus insulin regimen with excellent glycemic control. C-peptide level was 2.3 ng/mL (0.80 - 3.10 ng/mL) and GAD antibody level was <1. Her Mitochondrial DNA deficiency was seen in A3243G. She did not have overt hearing loss with planned formal audiometry testing. She remains on basal bolus insulin with excellent diabetes control.

Conclusion: MIDD can have a presentation similar to Type 1 or Type 2 Diabetes Mellitus based on the level of insulinopenia. Most commonly high frequency hearing loss predates the diabetes. Hearing loss, negative GAD antibody, elevated lactate level with a strong maternal family history of diabetes should raise the suspicion of mitochondrial diabetes. This case is a rare presentation given her mutation was known prior to onset of DM no hearing loss thus far.

Insulin Stimulates a Burst in Intracellular Hydrogen Peroxide that is Short-Lived, Highly Localized within the Cell, and Essential for Normal Balanced Insulin Action

Jain S, Williams KJ, Wu X

Chronic overnutrition causes a cluster of seemingly unrelated health problems called the 'metabolic' or 'atherometabolic' syndrome. Each component of the atherometabolic syndrome is worsened by, pathway-selective insulin resistance and responsiveness (SEIRR). It leads to fatty liver, atherogenic dyslipoproteinemia, deteriorating glucose control, and substantially increased risks for type 2 diabetes mellitus (T2DM) and cardiovascular events.

Normal insulin signaling includes two canonical limbs – namely, the MAP kinase (ERK) limb and the metabolic (PI3K-AKT) limb. The Williams laboratory recently discovered a noncanonical third limb of insulin signaling, in which insulin activates an oxide transport chain that functions as the master regulator of the balance amongst the other limbs of insulin signaling.

The oxide transport chain (NSAPP) begins when insulin stimulates NAD(P)H oxidase-4 (NOX4) to generate the superoxide anion ($O_2^{\bullet-}$). After several molecular hand-offs, the chain ends when aquaporins (AQPs) channel H_2O_2 to inactivate the phosphatase and tensin homolog deleted on chromosome 10 (PTEN) and protein-tyrosine phosphatases (PTPases), which are intracellular enzymes that otherwise block insulin-stimulated glucose control. Importantly, the NSAPP oxide transport chain fails to function during chronic overnutrition, resulting in this harmful pattern of SEIRR.

Insulin had long been known to provoke a transient intracellular burst of H_2O_2 . The NSAPP oxide transport chain appears to be the molecular explanation. In new, unpublished data, we found that the insulin-stimulated burst of H_2O_2 is highly localized to a single perinuclear area within each hepatocyte. The organelles and molecular participants in the precise subcellular localization of the insulin-stimulated H_2O_2 burst are currently under investigation.

Immune Related Adverse Events as a Biomarker in Non-Melanoma Patients Treated with Programmed Death 1 Inhibitors (PD-1Is)

Judd J, Zibelman M, Handorf E, O'Neil J, Bentota S, Doyle J, Uzzo RG, Bauman J, Borghaei H, Plimack, Mehra R, Geynisman DM

Background: The PD-1Is pembrolizumab (P) and nivolumab (N) have demonstrated clinical activity in multiple tumor types. They can lead to unique immune related adverse events (irAEs). Studies have shown conflicting results on whether the development of irAEs correlates with clinical response in melanoma patients (pts). Correlation of irAEs with better response in other cancer subtypes is unknown.

Methods: Retrospective study of non-melanoma pts who received PD-1Is. Primary objectives were to correlate the development of any irAE, or any irAE requiring steroid use, with clinical response. Response was recorded at first restaging scan, and endpoints included overall response rate (ORR) per RECIST, overall survival (OS), and time to next therapy or death (TTNTD). Fisher's exact tests and Kaplan-Meier curves with log-rank tests were used.

Results: 135 patients treated with N or P; average age 64 years, 65% were male. Cancer subtype: NSCLC (44%), RCC (23%), head and neck cancer (18%), urothelial carcinoma (13%), other (2%). IrAEs were noted in 45.9% with steroids required in 30.4%. Endocrinopathies, dermatitis and colitis occurred in 25.9%, 7.4% and 6.7% of patients, respectively. Of the 112 pts for whom complete response data was available, we found no significant correlation between ORR and either incidence of any irAE ($p=0.21$) or the use of steroids ($p=0.27$). There was also no significant association of irAE incidence or steroid use with OS ($p=0.981$, $p=0.954$ respectively) or TTNTD ($p=0.277$, $p=0.377$ respectively).

Conclusion: This retrospective review showed no association between the development of irAEs and outcomes in non-melanoma pts treated with PD-1Is.

Taste and Smell Disturbances in Patients with Gastroparesis and Gastroesophageal Reflux Disease

Kabadi A, Saadi M, Schey R, Parkman HP

Patients with gastroparesis and gastroesophageal reflux disease (GERD) often report decreased enjoyment when eating, presumably from upper GI symptoms. From experience, many patients have also reported changes in taste and smell. Disturbances to tastes and smells are notable among elderly and smokers, but have not been studied in patients with gastrointestinal disorders.

Aims: (1) determine if taste/smell disturbances are present in patients with gastroparesis and/or GERD; (2) establish if taste/smell changes relate to gastrointestinal symptom severity.

Methods: Patients with gastroparesis and/or GERD completed questionnaires: Taste and Smell Survey, PGI-SYM and demographic information. PGI-SYM was used to calculate gastroparesis cardinal symptom index and heartburn/regurgitation score.

Results: 76 subjects were enrolled: healthy controls (HC) (n=13), gastroparesis (n=30), gastroparesis/GERD (n=23) and GERD (n=10). Compared to HC, taste and smell disturbances were significantly higher in patients with gastroparesis, GERD and gastroparesis/GERD. The mean taste score was significantly higher in the gastroparesis/GERD group than the gastroparesis group alone (5.13±3.06 vs 3.13±2.66, p=0.017). Using mean+2SD of HC as the upper limit of normal, the gastroparesis/GERD patients had the highest prevalence of taste and smell abnormalities (70% and 52%, respectively) when compared to gastroparesis patients (40%, 40%), GERD patients (60%, 40%) and HC (8%, 8%). Taste score strongly correlated with HB/RG score (r=0.637, p<0.001) and GSCI (r=0.536, p<0.001). Smell score correlated to HB/RG score (r=0.513, p<0.001) and GSCI (r=0.495, p<0.001).

Conclusions: Taste and smell abnormalities are prominent in gastroparesis, GERD and gastroparesis/GERD patients. Both gastroparesis and GERD symptom severity are significantly correlated with taste and smell changes.

Peripheral Blood Eosinophilia as a Marker of Exacerbations in Hospitalized Chronic Obstructive Pulmonary Disease Patients

Kaur A, Dominguez-Castillo E, Duffy S, Marron R, Zhao H, Criner GJ

Rationale: Airway inflammation is associated with the disease progression and exacerbation episodes in COPD (AECOPD). Eosinophilia has been reported in sputum, BAL and airway wall in patients with asthma and chronic bronchitis. Previous studies with peripheral blood eosinophilia (PBE) and AECOPD have shown that almost one-fourth of the patients have eosinophilia during an exacerbation. There is some evidence that eosinophilia might be associated with steroid responsiveness in patients with exacerbations. However, there is paucity of data defining relationship of PBE and outcomes in patients hospitalized with AECOPD. We hypothesized that PBE are increased in AECOPD, especially severe exacerbations that require hospitalization. We tried to discern the relationship between PBE and severity and duration of AECOPD.

Methods: We conducted a retrospective chart review of COPD patients from our clinic who got admitted to Temple University Hospital for AECOPD in past 3 years. We analyzed their demographics, comorbidities, medication history, spirometry and laboratory evaluation during an exacerbation, after stratifying them into low (<0.25K/mm³) and high (>0.25K/mm³) eosinophil groups. We further looked at their treatment patterns during hospitalization in terms of use of steroids dose and duration, length of stay (LOS), and readmission rates, as markers of steroid responsiveness.

Results: 90 patients who had a hospitalization in past 2 years were stratified into low eosinophil (n=13) and high eosinophil (n=77) groups per the admission hematology data. The groups had no difference in the hospitalizations in the past year for AECOPD, LOS, steroid dose or duration and readmission rates, though there was a trend towards higher LOS and corticosteroid therapy duration in high eosinophil group.

Conclusion: In this cohort of patients hospitalized with AECOPD, patients with higher eosinophil counts had a trend towards having longer length of stay and duration of steroid therapy, though there was no difference in exacerbation or readmission rates.

Unusual Case of Neurosarcoidosis with Hypercalcemia and Anterior Pituitary Dysfunction

Khare S and Anolik J

Introduction: Neurosarcoidosis (NS) is a rare disorder. When present patients usually present as diabetes insipidus (DI) or disturbances in thirst, sleep, appetite or temperature. Pituitary lesions may also lead to partial or pan-hypopituitarism. We present an unusual case of NS with pan-hypopituitarism in absence of DI.

Case Presentation: 62 year old male with distant history of sarcoidosis who developed axonal neuropathy. It improved after short course of prednisone. A few weeks later he was hospitalized for surgery and was found to have hypercalcemia. Labs revealed calcium 13.8, Mg 1.3, Phosphorus 2.7, Cr 2.08, Na 142, 25 (OH) Vit D 22, 1, 25 (OH) Vit D <8, PTH 2, ACE 106. He denied thirst or polyuria. Subsequent work up revealed prolactin 12, LH <0.2, FSH <0.7, free testosterone <1, serum osmolality 290, urine osmolality 441, TSH 0.21, free T4 0.6, cortisol 3.7, ACTH 45. MRI showed a homogeneously enhancing hypothalamic mass measuring 1.3 cm with normal sella. He was started on levothyroxine, testosterone gel and prednisone 5 mg daily. His calcium normalized. There is no evidence of active sarcoid elsewhere although his gait abnormality and numbness in his low extremities have progressed.

Discussion: Sarcoidosis is an inflammatory non-caseating granulomatous multisystem disease of unknown etiology. NS occurs in 5-15% of cases of sarcoidosis and anterior pituitary dysfunction in the absence of DI is very uncommon as is the presentation with hypercalcemia. We cannot explain the low 1, 25 Vit D although it was obtained when he was already on steroids.

NS is a sometimes life-threatening disorder generally occurring in patients with substantial systemic involvement. Our patient was in remission for approximately 25 years when he developed NS with no systemic features.

Onset of NS is most often in the fourth or fifth decade of life. Presentation of NS can be acute, subacute or chronic. Its course may be monophasic, relapsing-remitting or progressive. When the pituitary is involved, DI is the most common clinical feature. Our patient is unusual as his NS presented with hypercalcemia and he has anterior pituitary dysfunction without DI or systemic disease.

Spontaneous remission is rare and immunosuppressive therapy is generally required. MRI abnormalities can improve or disappear under such treatment, but pituitary dysfunction is generally irreversible.

Our patient is on replacement hormone therapy and is undergoing neurologic evaluation prior to initiating definitive therapy for the NS. Interestingly his calcium remains normal on 5 mg of prednisone daily.

**Cardiac Sarcoidosis – A Case Based Review of the Current Literature on
Diagnosis, Complications, and Management**

Klein EC, Al Maluli H, Pierce MJ, Cooper JA, Alvarez RJ

Cardiac sarcoidosis is an infiltrative disease of the myocardium with presentations that vary from asymptomatic involvement to sudden cardiac death. It is important to maintain cardiac sarcoidosis in the differential diagnosis for patients with suitable symptoms as the vast majority of diagnoses discovered post-mortem. In this case, patient EM presented with new-onset non-ischemic cardiomyopathy at the age of 53. While Sarcoidosis most commonly presents between 20 and 40 years old, there is a second peak between 50 and 65. The disease is also more common in women.

In Ms. EM's case, laboratory and echocardiographic findings were non-specific, but high-clinical suspicion for this disease led EM to undergo a cardiac MRI which showed late gadolinium enhancement in a non-coronary distribution. Subsequent PET scan revealed diffuse areas of FDG hypermetabolism throughout the left ventricle with hypermetabolic uptake in multiple lymph nodes, and lymph node biopsy positive for non-necrotizing, non-caseating granulomas confirmed the diagnosis.

EM was started on a regimen of prednisone 80mg (approximately 0.8mg/kg) for three weeks followed by 60mg through to the time of her follow-up PET scan done 2-months later. This repeat scan demonstrated a dramatic decrease in the size and intensity of EM's multiple sites of previously increased metabolic activity, with a near complete resolution of hyperactive lymphadenopathy. Additionally, repeat TTE done 3-months after diagnosis demonstrated improvement of left ventricular ejection fraction from 10 to 25%, with an improvement in NYHA functional classification from class III to class II.

Effect of Bifidobacterium infantis 35624 (Align) on the Lactulose Hydrogen Breath Test for Small Intestinal Bacterial Overgrowth

Kumar K, Saadi M, Ramsey FV, Schey R, Parkman HP

Background: Small intestinal bacterial overgrowth (SIBO) may be a cause of symptoms in patients with abdominal bloating, distension, and gas. SIBO is commonly assessed using a Lactulose Hydrogen Breath Test (LHBT). Many SIBO patients take probiotic supplements, such as Align (Bifidobacterium infantis 35624), to restore beneficial bacteria to the GI tract. The aim of this study is to determine the effect of B. infantis 35624 on hydrogen and methane production during LHBT.

Methods: Healthy subjects underwent LHBT before and after 2 weeks of daily Align administration. Hydrogen and methane concentrations were measured for each breath sample (Quintron Instruments, Inc). Results are expressed as mean \pm SE and analyzed using repeated measures ANCOVA. A breath test was considered positive if hydrogen and/or methane increased >20 ppm above baseline by 90 minutes of the test or if a dual hydrogen peak was present.

Results: 19 healthy subjects were studied. Hydrogen levels were similar pre- and post-Probiotic across the three-hour study ($p=0.768$). In contrast, methane levels were significantly affected with Probiotic administration ($p=0.012$). A rise in methane >20 ppm was seen in 3 subjects pre-Probiotic and 6 post-Probiotic.

Conclusions: This study found that 2 weeks of B. infantis 35624 (Align) supplementation affects LHBT assessment for SIBO by significantly increasing methane, but not hydrogen, production after lactulose administration. Methane levels reached values that would be considered positive for SIBO patients. This study suggests that patients undergoing breath testing should discontinue probiotic use prior to the test as these supplements may alter the test results.

**Preventing Readmission and Emergency Department Encounters with a
Clinic Team-based Approach**

Alexandra Lane, MD

As a safety-net hospital, Temple University Hospital physicians encounter a large number of super-utilizers—patients with complex medical needs and socioeconomic barriers that result in frequent hospitalizations and a high percentage of healthcare utilization. Much national attention has focused on interventions to reduce readmissions for these patients, but these interventions have largely fixated on creating new resources or programs requiring monetary and personnel expansion. Given that this type of expansion is a limiting factor for many safety-net hospitals, we propose to use existing resources, namely volunteer medical students, at an urban academic setting to enact similar improvements.

With focused attention and increased availability, the aim is to provide team-based multidisciplinary care via students to super-utilizers to decrease readmissions and Emergency Department visits. A secondary aim is to improve quality of life for the patients and improve their trust in the healthcare system. Finally, we hope to provide meaningful patient interaction for students during a period of their academic life when patient access is limited and their experience working within a multidisciplinary team is only theoretical.

Although the project is still ongoing, preliminary data suggests some success, evidenced by a decrease number of ED visits, admissions, and costs overall for those enrolled. Insights and barriers into how such a program can be run and managed have also been gained and are hypothesis-generating for future research in the field.

Abnormal Esophageal Manometry and pH Impedance is Independent of Lung Disease Etiology in Pre-lung Transplant Patients

Langworthy J, Schey R, Ehrlich A, Midani D, Gaeckle N

Background: Esophageal dysmotility and gastroesophageal reflux (GER) are increasingly being investigated as potential contributors to abnormal allograft function following lung transplantation. However, there is limited evidence in the literature utilizing HREM (High Resolution Esophageal Manometry) and esophageal impedance testing comparing fibrotic lung disease, COPD, and other types of advanced lung disease.

Methods: We conducted a retrospective chart review of patients presenting to our institution over a five-year period for pre-lung transplant evaluation. The study population included patients who underwent HREM and esophageal impedance testing, as well as completed a pre-procedure symptom questionnaire. Manometry abnormalities were defined as any of the following: presence of a high-pressure segment >1cm, Integrated Relaxation Pressure (IRP) > 15 mmHg, Distal Contractile Integral (DCI) < 450 or > 5000, bolus clearance < 80%, or presence of hiatal hernia > 2cm. Impedance abnormalities were defined as percent time pH under 4 > 4.5% and DeMeester GER composite pH score <15.

Results: One hundred fifty one patients met the study criteria. Our study population included 98/151 (65%) patients with fibrotic lung disease, 31/151 (20%) with COPD, 20/151 (15%) with other pulmonary diagnoses. 96/151 (64%) were male. Mean age was 61.7. We found 79% had abnormal esophageal manometry findings, 59% reported upper gastrointestinal (UGI) symptoms, and 22% had abnormal pH impedance testing. The prevalence of abnormal esophageal motility, UGI symptoms, abnormal acid exposure was not statistically different between fibrotic, COPD, and other advanced lung disease groups.

Conclusions: Manometry abnormalities, UGI symptoms, and abnormal acid exposure are highly prevalent in patients with advanced lung disease. However, this appears to be independent of the etiology of lung disease.

Clinical Aspects of Chronic Pruritus and Impact on Quality of Life in Patients with Scleroderma

**Lavery MJ, Stull C, Lee H, Valdes-Rodriguez R,
Nattkemper LA, Blum M, Yosipovitch G**

Introduction: Scleroderma has been reported to have a high prevalence of chronic itch (42.6%). However, clinical characteristics of pruritus and its impact on quality of life have not been previously assessed.

Objectives: We assessed the prevalence, characteristics, and effect of chronic itch on quality of life in patients with scleroderma.

Methods: 36 subjects diagnosed with scleroderma were recruited to participate in the study from February 2014 to November 2015. All participants were asked if they suffer from chronic itch, and the patients that had itch were asked to fill out a validated questionnaire for the assessment of pruritus, as well as a quality of life questionnaire (ItchyQoL).

Results: The mean age was 58 ± 13 (range 42-86), 33 (92%) of the participants were female. Of all the patients, 12 of the 36 subjects (33%) reported suffering from chronic itch. The mean NRS itch intensity was significantly high (8.7 ± 2.2 , scale 0-10). Eight subjects (80%) reported itch that interfered with their sleep, and 7 subjects (70%) reported that itch restricted their life. The most common locations for pruritus were the scalp and arms. All twelve patients (100%) reported exacerbation of their itch by dry skin, and itch was reported to be worst during the winter. Those that suffered from itch had a total QoL score of 67 ± 19.7 (scale 21-105), which correlated significantly with NRS itch intensity ($r=0.7$, $p 0.01$).

Conclusions: Chronic itch in scleroderma patients is prevalent, highly associated with dry skin, and adversely affects quality of life.

**Ceftaroline in the Treatment of Methicillin-Resistant Staphylococcus Aureus
Bloodstream Infections**

Schultz S, Li SK and Gallagher J

Methicillin-resistant Staphylococcus aureus (MRSA) bloodstream infections (BSIs) are complicated and have limited treatment options. Ceftaroline, a novel cephalosporin with anti-MRSA activity, has limited published data for MRSA BSIs, thus we sought to describe our experience with this indication. This was a retrospective case series of patients who received ceftaroline for treatment of MRSA BSI at Temple University Hospital (TUH) in Philadelphia, PA from July 2013 to September 2014. The primary outcome was clinical success, defined as a composite of 30-day survival and clearance of MRSA from blood cultures on antimicrobial therapy. The secondary outcome was microbiological cure defined as clearance of MRSA from blood cultures on ceftaroline. 34 patients who received greater than 24 hours of ceftaroline administration were identified. 19 patients were excluded for indications other than MRSA BSI. Patients had a median age of 52 and 46% were male. Of 15 patients included, 12 (80%) achieved clinical success. All 15 patients achieved microbiological cure. All patients had clearance of their BSI within 96 hours. 3 patients had proven endocarditis; 2 of these had clinical success. 2 patients had infected ventricular assist devices; all of these had clinical success. 3 patients (20%) experienced mortality within 30 days. 2 patients developed Clostridium difficile infection on ceftaroline; no other significant adverse drug reactions were reported. In conclusion, ceftaroline was an effective therapy for MRSA BSIs in our treatment-experienced population.

**Bravo pH Monitoring After Per Oral Endoscopic Myotomy (POEM)
Overestimates True Acid Reflux**

Malik Z, Saadi M, Modayil R, Schey R, Parkman H, Stavropoulos S

Per oral endoscopic myotomy (POEM) has been increasingly used as treatment for achalasia. Success rates have been good, but patients may be at increased risk for acid gastroesophageal reflux (GERD). Previous studies have shown stasis fermentation can result in pH drops below 4, but changing a pH cutoff to <3 can correct for this.

Aim: To analyze post-POEM Bravo studies using a pH cutoff of 3 and compare these to using a pH of 4 to help differentiate between stasis fermentation and true reflux.

Methods: Patients who underwent POEM underwent an endoscopically placed Bravo pH monitor after the POEM procedure to assess for GERD. All studies were analyzed traditionally for acid reflux at a pH<4, then were reanalyzed at a pH<3.

Results: Ninety-one patients underwent a POEM followed by a Bravo study. Fifty patients had a positive Bravo using standard analysis. Only 30 patients had a positive study when using a pH<3 ($p<0.01$). Twenty patients with a positive study at pH<4 became negative at a pH < 3. Endoscopically, 54% of patients who were positive at pH 4 had mucosal breaks, while 73% who were positive at pH 3 had mucosal breaks.

Conclusions: Reflux occurs post-POEM, however, conventional analysis overestimates the number of patients with GERD. Our analysis shows a reduction in the number of patients from 55% to 33% meeting criteria for GERD. Analysis of the Bravo measuring acid reflux at a pH<3 rather than a pH<4 may improve detection of true reflux and eliminate effect of stasis fermentation.

Analyzing the Distribution of Cardiac Comorbidities Across GOLD Grades and Their Effect on Readmission Outcomes

Marron R, Duffy S, Kaur A, Dominguez E, Gaeckle N, Criner G

Rationale: Multiple clinical tools have been developed to classify patients with Chronic Obstructive Pulmonary Disease (COPD). Traditionally, patients have been classified by their level of airflow obstruction. Recently the GOLD Classification changed to incorporate symptom burden and exacerbation rate. These classifications may overlook the importance of comorbid illness, in particular cardiac disease, in predicting the disease course of COPD patients.

Methods: We performed a retrospective chart review of 227 patients with COPD in the outpatient clinic records at Temple Lung Center. Comorbidities, demographics, smoking history, acute exacerbations of COPD (AECOPD), and hospitalization data were collected. Patients were placed into subgroups based on GOLD classification (A, B, C, D) as well as subgroups of patients with and without cardiac comorbidities. Cardiac comorbidities were defined as a history of myocardial infarction, coronary artery disease, or an ejection fraction of less than 50% on echocardiography.

Results: Of 227 patients, 73 (32.2%) had cardiac comorbidities. Patients with cardiac comorbidities had an increased rate of COPD hospitalizations per person year (1 vs 0.62,) and higher rates of COPD readmission within 30 days (0.46 vs 0.09) and 90 days (1.08 vs 0.45). They had similar rates of overall AECOPD per person-year in a 12-month period (1.45 for those with cardiac comorbidities, 1.42 for those without, p=NS) and in a 24-month period (1.55 vs 1.35). FEV1 was also near equivalent among the groups (52.2% predicted vs 57.6% predicted). Patients with cardiac comorbidities were more frequently GOLD D, and less frequently GOLD A or B than patients without cardiac disease. Cardiac comorbidities were distributed similarly in group C.

Conclusion: COPD patients with cardiac comorbidities had a higher rate of COPD hospitalizations and a significantly higher readmission rate at 30-and 90-days than patients without cardiac disease. They were more frequently classified as GOLD D and less frequently as GOLD A and B, implying dyspnea and/or exacerbation rate could be worse in these patients. Higher readmission rates in patients with cardiac comorbidities may reflect the difficulty in differentiating the symptoms of COPD from those of heart disease and properly treating the etiology of those symptoms.

**Difference of Achalasia Subtypes based on Clinical Symptoms,
Radiographic Findings and Stasis Scores**

Meillier A, Midani D, Caroline D, Saadi D, Schey R, Parkman H

Background: Three subtypes of achalasia are defined with high-resolution esophageal manometry (HREM): subtype I shows no pressurization with swallows, II has increased isobaric pan-esophageal pressure, and III has distal esophageal spastic non-isobaric contractions. Studies describing the subtypes based on radiological, clinical and stasis scores are limited.

Aim: To determine the differences in clinical symptoms, radiographic findings and stasis scores for the three subtypes of achalasia.

Methods: Patients undergoing HREM received a questionnaire for current symptoms and previous treatments. The questions included presence of symptoms and severity. Pre-HREM barium swallow tests were evaluated for maximum esophageal diameter. Stasis scores were calculated using the transit patterns on HREM.

Results: 108 patients with HREM diagnosis of achalasia (I n=8, II n=84, III n=16) from 1/2012-6/2015 were included. Gender distribution was similar among subtypes. Patient age was younger for subtype I ($38\pm 16Y$), compared to II ($55\pm 17Y$) and III ($63\pm 17Y$) ($p=0.03$). Esophageal symptoms did not differ between subtypes regarding severity of nausea, chest pain, coughing and heartburn except with type I having increased vomiting severity (2.8 ± 1.4 vs 1.4 ± 1.4 vs 1.2 ± 1.2 , $P<0.01$). A significant difference in esophageal dilation radiologically was seen between subtype II and III (35.1 ± 14.4 vs 24.0 ± 7.2 mm, $P=0.023$). Stasis scores did not significantly differ among the subtypes.

Conclusions: Achalasia subtypes had limited variation among clinical symptoms with vomiting severity being the exception. The maximum diameter of the esophagus in subtype II was significantly higher than subtype III. Esophageal stasis scores were similar. Thus, HREM remains fundamental in assessing the achalasia subtypes.

43 Year Old Female with Spontaneous Coronary Artery Dissection; Challenges with Revascularization and Role of Optic Coherence Tomography in Identification of Left Main Involvement

**Mirza A, Lakhter V, Hasani AZ, Toyoda Y, Cohen H,
O'Murchu B, O'Neill B, Bashir R**

A 43-years-old African American female cop who presented to an outside hospital with sub-sternal chest pain soon after shoveling snow. Initial Electrocardiogram (EKG) revealed normal sinus rhythm with T wave inversions in leads V4 - V6. Pt was admitted at the outside hospital and underwent a Persantine stress test which did not result in any reproduction of symptoms, EKG changes or perfusion abnormalities. Half an hour after her negative stress test, patient experienced sudden excruciating chest pain. Repeat EKG now showed new anterior ST segment elevations. She was referred to Temple University Hospital for emergent Percutaneous Intervention (PCI). During emergency catheterization, coronary angiogram revealed progressive tapering of the Left Anterior Descending (LAD) artery with total occlusion in the mid segment highly suggestive of Spontaneous Coronary Artery Dissection (SCAD). After flow in the LAD was restored with wiring and thrombectomy, patient was noted to have a spiraling dissection extending into Left Main (LM) coronary artery which was confirmed by Optical Coherence Tomography (OCT). The coronary guidewire was left inside the LAD. An Intra Aortic Balloon Pump (IABP) was placed and an emergency off-pump coronary artery bypass (CABG) was performed. Coronary guidewire was removed intraoperatively after successful grafting of LIMA anastomosis. Biopsy of the aorta was normal. The patient had an uneventful course following her surgery. Follow-up Echocardiogram revealed an EF of 40% at one month and 60% at six month.

**Single Center Experience with Successful Left Ventricular Assist Device
Explantation Following Myocardial Recovery**

Mirza A, Raza F, Shiose A, Schwartz D, Toyoda Y, Alvarez R, Hamad E

Purpose: Very few patients with Ventricular Assist Device (VAD) achieve functional myocardial recovery to the point of VAD explantation. Therefore, we aim to share our experience of 6 patients who received Heartmate II LVAD at our center and subsequently underwent successful explantation following myocardial recovery.

Results: Baseline characteristics of overall cohort (n=6) were: Age (years) 53.7 ± 11.7 , 67 % Males, 50 % with Non-Ischemic Cardiomyopathy, 50 % Caucasians and 33.3% African Americans, Mean Duration of VAD support was 8 months. Mean VAD free survival was 18.7 months. Comparison of echocardiographic and hemodynamic data (Pre-Implant Vs Pre-Explant) revealed significant improvement in LVEF (%) [14.1 ± 6.6 vs 53.3 ± 9.8], LVIDd (mm) [56.3 ± 4.8 vs 43.5 ± 5.7], LVIDs(mm) [49.0 ± 9.0 vs 34.3 ± 6.3], Base RVIDd (mm) [38.5 ± 6.1 vs 35.4 ± 5.2], RA pressure (mmHg)[13.2 ± 2.7 vs 5.2 ± 4.4], Mean PAP (mmHg) [38 ± 6.7 vs 15.7 ± 7.7], CO (L/min) [3.9 ± 1.2 vs 5.8 ± 1.6], CI (L/min/m²) was 1.9 ± 0.5 vs 2.7 ± 0.7 (p=0.024).

Conclusion: There was a significant improvement in LVEF, LVIDd, LVIDs, RA pressure, Mean PA pressure and Cardiac Index to near normal values in all patients who underwent successful LVAD explantation and who remained VAD free for more than a year post explant. Patients who have near normalization of these parameters are more likely to tolerate explant and remain VAD free.

**ST-Elevation Myocardial Infarction and Acute Limb Ischemia in a Young Female:
An Arterial Conundrum**

Mirza A, Suri J, Mangrolia H, Alvarez R, Hamad E

Oral contraceptives (OC) have already shown to increase the risk of venous thromboembolism in women over 35 years old who have a history of smoking. Data, through studies are now presenting ever increasing evidence of arterial events in these women who use OCs. However the role of prothrombotic mutations, such as G20210A (prothrombin) gene, in arterial thrombosis remains controversial. Here we present the case of 34 year old female with a history of smoking, on OCs, who was admitted for a massive myocardial infarction involving the left anterior descending artery, later found to have a significant occlusion at the aortic bifurcation. It was discovered on subsequent testing that she was also heterozygous for the prothrombin gene mutation. Our case report highlights that there may be a synergistic effect of Factor II prothrombin mutations and oral contraceptives in arterial thrombosis. Furthermore, concomitant use of OC and smoking in young women may increase risk of Myocardial infarction and should therefore be avoided.

Response of Respiratory Symptoms in Outpatient Chronic Obstructive Pulmonary Disease Patients to Administration of Prednisone Therapy

Mulhall P, Leung J, Criner G

Background: Prednisone is commonly prescribed to treat patients with an escalation of respiratory symptoms that herald an acute exacerbation of chronic obstructive pulmonary disease (COPD). The dose and duration of prednisone that is required to achieve improvement of patient symptoms is not clear. Some authorities recommend 5 days of therapy, others 10-21 days of standard doses or tapering therapy. We sought to determine the temporal response of the patient's daily symptoms of COPD exacerbation to a 10 day course of prednisone by using an electronic diary on a daily basis.

Methods: 40 consecutive patients that were enrolled into a COPD telemedicine based disease management program were administered 10 days of 30 mg of prednisone had retrospective review of their daily as well as peak expiratory flow rates. Dyspnea (modified Borg), sputum quantity, color and consistency, peak flow rates and upper airways symptoms of sneezing, sore throat, cough and wheeze were recorded.

Results: 40 patients with mean FEV1 36 % of predicted were enrolled. Patients had 90 days of monitoring. 31 patients had administration of steroids for increased symptoms from baseline. 5 patients had resolution of symptoms in 3 days, 6 patients in 7 days and 7 in 10 days. 11 patients had ongoing symptoms despite 10 days of prednisone therapy.

Conclusion: There is marked heterogeneity in the response to systemic steroids for the treatment of symptoms associated with COPD exacerbations. Development of better tools to determine the indication, likelihood of benefit and duration of treatment with corticosteroids for the onset of a COPD exacerbation are desperately needed.

**Safety and Efficacy of Hepatitis C Treatment in Patients with HCV/HBV
Coinfection and in Patients with HCV/HIV/HBV Coinfections
Using Interferon Free Regimens**

Mur T, Kutmah A, Alsabbagh E

Introduction: Patients with HCV/ HBV coinfection are at increased risk for accelerated fibrosis progression, decompensated cirrhosis and development of HCC. The new HCV treatment regimens with direct acting antivirals (DAAs) proved to be very safe and effective with high SVR rates, however, these medications don't have an activity against HBV and patients with known HBV coinfection were excluded from enrollment in the HCV treatment trials.

Methods: we identified patients with HBV/HCV coinfection including those who had HIV, we treated HCV based on genotype and according to treatment guidelines for HCV mono-infection. Outcomes of interest were safety (measured by discontinuation of therapy due to side effects, serious AEs) and efficacy determined by SVR.

Results: 7 patients with HCV infection and chronic inactive carrier state of HBV were treated. All except one had elevated liver enzymes. 1 patient was lost to follow up and didn't have SVR labs. 6 patients had GT 1a, all treated with sofosbuvir/ledipasvir and 1 patient had GT2, treated with sofosbuvir/ribavirin. 3 patients were on HBV treatment prior to initiating HCV therapy. At end of treatment all patients had normal enzymes. All 6 patients achieved SVR. 3 of 4 patients who were not on HBV treatment had a rise in their HBV DNA (75%). No serious SEs or discontinuation of treatment

Conclusion: in our experience, same medications used for HCV mono-infection were effective in treatment of patients with HCV/HBV and HIV coinfection with high SVR rate, treatment was safe but special attention is needed to monitor for possible HBV activation.

How Can Primary Care Physicians Improve the Referral System to Medical Subspecialists?

Norberg S, Shah A, Leung J, Shah K, Judd J, Yee JL

An important part of outpatient medicine is the ability of a primary care provider to refer a patient to a medical subspecialist when necessary. Previous quality improvement projects performed by internal medicine residents have shown that most of the patients sent for referrals do not actually see a subspecialist. There are multiple different reasons for the lack of success on getting patients to see the intended subspecialist, some of which is the suboptimal utilization of our electronic medical record system, EPIC. Since EPIC was identified as an important tool in optimizing efficient referrals to medical subspecialists, the goal of our quality improvement project was to determine what tools subspecialists feel is the most effective and efficient way to communicate with the referring primary care provider regarding the referred patient. This was accomplished by sending a short survey to attending physicians and fellows from different medical subspecialties. Based on the survey, 75% of medical subspecialists feel that EPIC is the tool that provides the most efficient and effective communication between the referring physician and subspecialist. Of those 75%, about half of subspecialists use the most recent progress note to best understand why the patient was referred to them and the other half use a personal EPIC message written by the referring provider explaining the reason for the consult (if this is provided by the referring provider). This survey confirms the importance of optimizing our electronic medical record in order to provide our patients with appropriate medical subspecialty care.

Case Report of Meningococemia without Meningitis

Oliveros E, Bohsali F, Taqui B

Meningococemia may present with or without obvious meningitis. The infrequency of this bacteremia without meningitis makes it challenging to diagnose. A 25 year-old female presents with fever and abdominal pain. Endorses rigors and occipital headaches described as non-radiating, pressure like, aggravated by light without alleviating factors. Her past medical history is significant for systemic lupus erythematosus, antiphospholipid antibody syndrome, and idiopathic thrombocytopenic purpura with subsequent splenectomy. Current medications include mycophenolate, hydroxychloroquine and prednisone. Upon physical examination, she is febrile, tachycardic, blood pressure 116/64 mmHg, respiratory rate 16/minute and pulse oximetry saturation of 99%. Lungs and cardiovascular exam are within normal limits. Abdomen reveals a left flank scar, which is soft, non-tender, and without distention. Neurological exam reveals photophobia, without nuchal rigidity. She has leukocytosis to 22,000 (9% bandemia). Urinalysis, chest X-ray, C3 and C4 complements are normal. Chlamydia and gonorrhea serologies are negative. Initially treated with intravenous fluids, vancomycin and piperacillin-tazobactam. Mycophenolate was held. Computed tomography of the abdomen and pelvis did not show any abnormalities. Lower extremity duplex was negative for deep vein thrombosis. On day 3, her blood cultures grew 2/4 bottles of *Neisseria meningitidis*. Her antibiotics were narrowed to ciprofloxacin, and her contacts were notified.

Meningococcus is infrequently associated with sepsis in the hospitalized population. However, asplenic patients and those with terminal complement deficiency are at especially high risk of infection due to their inability to capitalize on the antigenicity of the capsule. This patient was likely an asymptomatic *Neisseria* carrier with reduced immune function.

Adverse Effects and Drug Discontinuation Rates of Antifibrotic Drug Therapy In Patients With FVC <50 %

Pandya A, Galli J, Vega-Olivio M, Criner G

Both nintedanib and pirfenidone have been FDA approved in idiopathic pulmonary fibrosis (IPF). Both the ASCEND and CAPACITY trials for pirfenidone and the INPULSIS trial for nintedanib used an exclusion criteria of a forced vital capacity (FVC) of <50%. Studies have shown that a significant change in FVC (>10%) in IPF was associated with a higher mortality. Determining the tolerability of antifibrotic therapy in those with an FVC <50% may help slow the rate of decline in individuals with severely reduced lung function. The hypothesis was that the rates of adverse effects and drug discontinuation rates would be similar in patients with FVC <50% compared to those with FVC >50%. A single-center, retrospective chart review study on patient receiving treatment with pirfenidone or nintedanib with radiographic and clinical evidence or lung biopsy proven IPF from September 2014 to October 2015 was performed. 29 subjects with an FVC <50% were matched with 29 other subjects in the study population by age, gender, and home oxygen use. Multivariate analysis showed there were no significant differences between the two groups based upon adverse reaction (Nausea, P = 0.557; Diarrhea, P = 0.753; GERD/Dyspepsia, P = 0.717; Rash/Photosensitivity, P = 0.446) or rate of drug discontinuation (P = 0.119). Nintedanib and pirfenidone maybe used in patients with an FVC <50% without an increase in adverse effects or drug discontinuation rates. Future studies will show if patients with an FVC <50% on antifibrotic therapy have the same benefits of disease free progression and reduction in decline of FVC as the original study population.

Identifying Prognostic Factors in Peripartum Cardiomyopathy: A 22 Year Retrospective Analysis of an Urban Population in the US

Peters A, Caroline M, Zhao H, Tsai E

Background: Outcomes for peripartum cardiomyopathy (PPCM) vary widely. Many patients recover cardiac function and survive event-free, while others develop advanced heart failure and require cardiac transplantation. Whether patient characteristics or baseline cardiac parameters can predict early and late major adverse outcomes is unknown.

Methods and Results: We analyzed multiple clinical and echocardiographic characteristics in 53 PPCM patients with at least 6 months follow-up data, in a single urban university medical center. The primary composite outcome was persistent or progressive severe LV dysfunction (LVEF<30%), mechanical circulatory support device implantation, cardiac transplantation, or death. Median (IQR) follow-up period was 6.9 (2.9-9.1) years. Nine (16.6%) PPCM patients had persistent severe LV dysfunction at 3 years or beyond. Fifteen patients (28%) underwent cardiac transplantation, at a Median (IQR) 5.5 (2.4-9.8) months from PPCM diagnosis. Univariable logistic regression identified diabetes (OR 3.93, 95% CI 1.06-14.49), moderate to severe RV dysfunction (OR 3.85, 95% CI 1.22-12.1), LV internal diameter in diastole (LVIDd) >60mm (OR 11.1, 95% CI 1.2-99), and LVEF<30% (OR 12.57, 95% CI 2.5-63.24) as associated with persistent severe LV dysfunction. Initial moderate to severe RV dysfunction was associated with cardiac transplantation, LVAD or death within the first 12 months of follow-up (OR 31, 95% CI 3.6-268.7).

Conclusions: Right ventricular dysfunction and its severity can identify high-risk PPCM patients who may benefit from early referral and evaluation for advanced therapies for chronic heart failure, including mechanical circulatory support device implantation or cardiac transplantation.

Telomere Length as a Predictor of Overall Survival in Colorectal Cancer

Podolski A, Lewis B, Devarajan K, Cohen S, Dotan E

Background: Telomeres have important functions in regulating cell replication and maintaining genome integrity. Telomere length (TL) decreases with cell divisions and shortened TL often leads to genomic instability, resulting in loss of cell-cycle control, a hallmark of cancer. Telomere shortening has been found to be associated with increased risk of several cancers and various studies indicate that a short TL may be a sign of poor survival. The goal of the current study was to assess the role of TL as a prognostic indicator of overall survival in colorectal cancer.

Methods: DNA samples of 221 patients with colorectal cancer were identified through the tumor repository at Fox Chase Cancer Center. TL was determined analyzing 10 ng of genomic DNA via high throughput qPCR assay in SpectraCell Laboratories. Various statistical studies including the Cox PH model and log rank test were then used to analyze TL and its percentile in comparison to survival.

Results: No statistical difference was seen with TL and overall survival (OS) both when looked at with univariate analysis ($p=0.68$) and via multivariate analysis that accounted for age and stage ($p=0.13$; HR 0.94). There was a statistical difference noted between OS and age ($p=0.05$; HR 0.7) and stage ($p<0.0001$; HR 3.09), as well as between TL and age ($p= 0.0013$)

Conclusions: Similar to the general population, telomere shortening is seen with increasing age among patients with CRC. Increasing age and higher disease stage were also associated with decreased OS. However, no correlation was seen between TL and OS.

Impact of Depression on SLE Flare

Rathi S, Zavitsanos A, Goh KS, Caricchio R

Background: Neuropsychiatric systemic lupus erythematosus (NPSLE) involves a wide range of peripheral and central neuropsychiatric manifestations. Depression is one of those and is very common in patients with SLE.

Objective: The aim of this study is to identify the prevalence of depression in our patient population at Temple University Hospital, which is predominantly of African Americans, and also to determine its impact on SLE flare.

Method: Data were collected from patients with SLE at each clinic encounter since January 2014, included the demographics, medical, surgical and social history, medication list, laboratory results. Systemic Lupus Erythematosus Disease Activity Index (SLEDAI), Systemic Lupus International Collaborating Clinic (SLICC) Damage Index (SDI), were incorporated into our Epic Care EHR and routinely extracted to facilitate disease monitoring.

Result: Out of 220 patients 68 patients have diagnosis of depression mentioned in their medical history. On review of these patient charts, 58.8% patients are on antidepressants. Also approximately 58.8% patients had SLEDAI documented in their charts, which further helps us decide the frequency of lupus flare. 30% patients had flare and out of these patient 50% patients were not on any antidepressants.

Conclusion: Prevalence of depression is high in SLE patients, and our study clearly demonstrates that the risk of lupus flare is high in those untreated for depression. Hence, early prompt recognition of depression in lupus patients and its appropriate management is essential.

Rheumatology Narcotic Contract

Zavitsanos A, Goh KS, Rathi S, Livshits A, Tan I

Background: The use of opioids in treating chronic pain can be difficult and is a common problem across multiple specialties. Opioid contracts have become more common place to help standardize the way in which physicians prescribe narcotic medications. Currently, our office has no formal policy on prescribing controlled substances which has caused work flow issues among several office personnel.

Objective: To determine whether implementing a narcotic contract in the section of rheumatology will improve work flow by decreasing the number of narcotic-related EPIC phone messages.

Methods: TUH rheumatology practice implemented a formal written narcotic contract in August 2015. Using questionnaires, we polled our staff of various positions on their perception of the number of narcotic prescription-related patient phone interactions both 6 months before and after initiation of the contract. Furthermore, using the number of EPIC messages as our objective measurement, we compared the number of narcotic related phone encounters resulting in a narcotic prescription 6 months before and after the implementation of the contract.

Results: After we surveyed our ancillary staff, 4/4 perceived the number of narcotic-related phone calls as increasing or unchanged over the one year study period. Seventy three percent of providers agreed that the contract had positive effect on office work flow. Total number of epic messages and total number of office visits resulting in narcotic prescription decreased by 20% and 16% respectively, six months after implementation of the contract.

Conclusion: After implementation of the narcotic contract there was an overall objective decrease in number of phone calls and office visits resulting in narcotic prescriptions.

Dronabinol Increases Functional Chest Pain Threshold Without Altering Anxiety, Depression, or Metabolic Parameters: A 28 Day Study

Reichenbach Z, Malik Z, Sloan J, Bayman L, Valestin J, Rizvi-Toner A, Hasmi S, Catalano J, van den Berg P, Parkman H, Schey R

200,000 U.S. patients a year receive a diagnosis of Functional Chest Pain (FCP). Patients with FCP suffer from a poor quality of life and frequent cardiovascular workups impose great financial burdens on the healthcare system. Present treatments remain ineffective.

We designed a double blind, placebo controlled study to determine if treatment with the CB1 agonist, Dronabinol, could reduce esophageal hypersensitivity and relieve the symptoms of FCP. We hypothesized that since the CB1R plays an integral role in refinement of neuronal signaling that our administration of Dronabinol would reduce FCP symptoms. Our secondary end points examined changes in anxiety, depression, weight, and numerous metabolic parameters.

We found that Dronabinol significantly decreased pain intensity, odynophagia frequency, and tended to reduce the frequency of chest pain. Dronabinol treated patients showed significantly higher pain threshold during Esophageal Balloon Distention Testing (EBDT). No changes were found in measures of the Beck Depression or Anxiety indexes among the groups. There was no change in weight among the groups. The Dronabinol group tended to show a reduction in triglycerides, calculated LDL, and non-HDL cholesterol, although not significant. Other metabolic parameters remained unchanged with treatment. No significant side effects were reported by patients in the study.

Lipodystrophic Diabetes – A Report of Two Cases and Insight Into Management

Reznick T, Jain S, Vaz C

BACKGROUND: Lipodystrophy syndromes involve complete or partial loss of adipose tissue with sparing or accumulation of fat in other regions. Metabolic derangements include severe insulin resistance (IR), hypertriglyceridemia and hepatosteatosis. Clinical presentation may lead to misdiagnosis or excessive testing for Cushing's syndrome. Mediators of IR in lipodystrophy include increased TNF, free fatty acids, leptin and/or adiponectin deficiency.

CASE SERIES: 36-year-old female with uncontrolled diabetes (diagnosed at age 20) on 180 units of insulin daily. She had wasted extremities with increased abdominal and facial adipose tissue. BMI was 30 kg/m². Appearance changes began at age 8. Multiple tests for Cushing's syndrome were negative. HbA1C was 10.2%, triglycerides 4382, and she had hepatosteatosis.

46-year-old male with uncontrolled diabetes on 160 units of insulin daily. HbA1C was 9.9% and triglycerides were 1007. He had increased facial and abdominal adipose tissue relative to the extremities with acromegaloid features. BMI was 38 kg/m². Appearance changes started 5 years prior.

They had no striae or bruising, but had obesity and diabetes, which can lead to misdiagnosis. Both patients are being treated for lipodystrophy with double-concentrated basal insulin, prandial insulin, liraglutide and metformin with improvement. For hypertriglyceridemia, fibrate, high dose statin and low carbohydrate diet were started.

CONCLUSIONS: These cases highlight the importance of having a high index of suspicion for lipodystrophy. Lipodystrophic diabetes should be considered with severe IR, hypertriglyceridemia and lipodystrophy with fat accumulation in other regions. We highlight the successful use of a combination regimen of newer anti-diabetes agents for lipodystrophic diabetes with severe IR.

Fever of Unclear Origin as the Presenting Manifestation of Pulmonary Sarcoidosis – A Case Report

Swati Rushi, M.D

Sarcoidosis is a chronic systemic inflammatory disease characterized by granulomatosis usually involving the lungs. Symptoms are nonspecific and vary, although respiratory complaints are the most common, and in many cases diagnosis is delayed because sarcoidosis mimics other diseases in presentation. This is the report of a 25 year old female who presented with fever and cough, and was initially diagnosed with multifocal bronchopneumonia, however continued to have fever and cough despite antibiotic treatment. Abdominal imaging was performed to identify a source of infection. The patient was discovered to have multiple pulmonary nodules, and given her initial diagnosis of sepsis, out of concern for septic emboli an extensive workup was initiated. Transthoracic echocardiogram was negative for endocarditis and blood cultures grew no bacteria. She was found to have non-necrotizing granulomatous inflammation on transbronchial biopsy, consistent with sarcoidosis. After one year on prednisone, she no longer has cough and her sarcoidosis is in remission. A compilation of clinical, radiologic and histopathological data is required to establish a diagnosis of pulmonary sarcoidosis.

**Intrapyloric Injection of Botulinum Toxin for Gastroparesis:
Reassessing Efficacy with Utilization of EndoFLIP Pyloric Measurements****Saadi M, Malik Z, Schey R, Yu D, Parkman H**

Botulinum toxin A (BoNT/A) injection into the pylorus has been used to treat gastroparesis. Pyloric sphincter abnormalities have been detected in gastroparesis using Endoscopic Functional Luminal Imaging Probe (EndoFLIP). Use of EndoFLIP may help identify patients that might respond to BoNT/A. **Aims:** Measure pyloric sphincter characteristics using EndoFLIP in patients with gastroparesis. Determine symptom responses to BoNT/A treatment for gastroparesis. Correlate pyloric sphincter characteristics using EndoFLIP to the response to pyloric sphincter BoNT/A injections. **Methods:** EndoFLIP was performed on patients undergoing EGD for treatment of gastroparesis with BoNT/A 200 Units. EndoFLIP pyloric measurements (diameter, cross sectional area [CSA], pressure, and distensibility) were taken. PGI-SYM questionnaire was obtained and at follow up at 2, 4, 8 and 12 weeks. **Results:** 25 patients were enrolled (16 idiopathic gastroparesis (IG), 9 diabetic gastroparesis (DG)) with EndoFLIP assessment in 24 patients. 18 patients (75%) have had follow up at 2 weeks, 12 (50%) 4 week follow up, and 9 (38%) 8 week follow up. In IG, gastric retention (GR) at 4 hours had a negative correlation with EndoFLIP 40 cc balloon ($r = -0.60$; $P = 0.06$) and compliance ($r = -0.82$, $P = 0.04$). DG had no significant correlation in GR with distensibility and compliance ($r = -0.40$; $P = 0.50$, $r = 0.40$; $P = 0.60$). In DG, CSA-40 correlated with baseline retching ($r = -0.45$; $p = 0.08$) and vomiting ($r = -0.36$; $p = 0.19$) while compliance-40 correlated with gastric retention at 2 hours ($r = 0.76$; $p = 0.03$). Mean GCSI scores were reduced at 2, 4, and 8 weeks (1.96 ± 0.20 , $P < 0.001$, 2.15 ± 0.26 , $P = 0.11$, and 2.87 ± 0.16 , $P = 0.83$) respectively compared to baseline (3.23 ± 0.22). In both IG and DG, distensibility at 40 cc balloon distension correlated with significant improvement in their loss of appetite at 2 weeks ($r = 0.48$; $P = 0.047$) and 4 weeks ($r = 0.53$; $P = 0.09$). **Conclusions:** EndoFLIP compliance and distensibility correlate with GR. BoNT/A improved symptoms of nausea and retching. Improvement in loss of appetite correlated with pyloric distensibility in patients with GP.

Opiate Antagonist Therapy in High Risk Patients on Chronic Narcotics

Schmidt R, Szczesniak R, Chen X, Schmitz K, Yackoski J

Due to the recent increase in prescription opioid overdoses along with the increase in global consumption of these medications, it is paramount to identify patients at high risk for overdose and provide them with the means necessary to prevent such negative outcomes. The purpose of this study was to investigate the effect of education and opiate antagonist prescription on negative outcomes.

High risk patients were identified from the Jone's Hall and Medicine Group Practice by medical record query based on daily opioid dose equivalents. Once identified patients were invited to one of multiple resident run educational sessions regarding opiate overdose, prevention and treatment. Education materials were distributed to participants along with interactive instruction of opiate antagonist administration. At the end of the session participants were asked to complete a post class assessment as well as given a prescription for an emergent opiate antagonist. Participants were asked to disclose which pharmacy they would fill the medication. Fill rates of the opiate antagonist were collected by telephoning individual pharmacies and tallied for all participants.

Primary outcomes include percentage of prescriptions filled as well as assessment scores. Currently still in the data collection process of the project as there are still educational sessions remaining.

**Effectiveness of Brief Health Literacy Education Interventions
for Medical Residents**

Sheffer IG, Lucki M, Hyde J

Background: Low health literacy (LHL) has been linked to poor outcomes for many types of patients. Some research exists on methods to educate medical residents about health literacy, but described methods are resource intensive and may be difficult to implement.

Materials & Methods: Our study was part of a larger project examining health literacy in Temple's IM residency physicians and patients. We sought to test the effectiveness of two small educational initiatives on resident understanding of key concepts, and comfort in communicating with LHL patients. Residents were divided into two groups. Half received a self-study guide on health literacy and protected time to complete it. The others received an in-person lecture. Residents were asked to complete a survey rating their confidence communicating with LHL patients and testing key health literacy concepts immediately before and after the educational interventions.

Results: The two groups showed no difference in comfort communicating with LHL patients. Impact on understanding key concepts was variable for both interventions. Most residents identified their patient population as having low health literacy, and health literacy as important to outcomes. Residents in the self-study group had a lower rate of intervention completion based on number of surveys returned.

Conclusions: Though most residents recognize the impact low health literacy can have on patients, a brief educational intervention does not appear to be sufficient to improve comfort caring for this population. More work is needed to explore effective and efficient ways to increase resident skill in communication with patients of low health literacy.

Avoiding Allergens Associated with Food-Related Type 4 Hypersensitivity Reactions Improves Symptoms of Irritable Bowel Syndrome

Shin G, Luther S, Hong I, Ehrlich AC, Stierstorfer M, Smith MS

Background: Irritable Bowel Syndrome (IBS) is a functional gastrointestinal disorder characterized by abdominal pain coinciding with altered stool form or frequency. Recent developments in the pathophysiology of IBS point to various causes including food triggers. Food related type 4 hypersensitivity reactions may contribute to IBS symptoms.

Objective: To evaluate if identified type 4 food allergens, when eliminated from the diet, alleviate symptoms of IBS, suggesting that the etiology of symptoms in some cases is a newly described disease, allergic contact enteritis (ACE).

Methods: Patients with IBS were eligible for study participation. Skin patch testing was initiated using an extensive panel of type 4 food allergens identified in the literature. Patch test application was on day 1. On day 3, patches were removed and initial results read. Final patch test was read on day 4 or 5, to allow for any delayed reactions. A questionnaire was distributed at this time, to be completed after one month of avoiding the identified allergens.

Results: Thirty-nine patients were included in this study. Initial score was 6.36 ± 2.5 out of 10 for abdominal pain/discomfort. We found improvements in the severity of abdominal pain by 4.22 ± 2.7 points ($p < 0.001$) and IBS symptoms by 5.44 ± 3.3 points ($p < 0.001$), all after one month of food avoidance. Overall, 85% of patients had some or total improvement in their symptoms.

Conclusion: 85% of patients with IBS had some or complete improvement with avoidance of known type 4 food allergens. This raises questions about a possible overlap between IBS and allergic contact enteritis.

Painful Limb, Dusky Toes, Intact Pulses: a Case of Venous Limb Gangrene

Swed B, Godwin JL, Rao AK

Gangrene of the digits or skin is typically associated with arterial occlusions. However, gangrene and skin necrosis may occur with intact arterial pulses due to thrombotic pathogenesis within the venous circulation; this clinical entity – venous limb gangrene – is not well recognized and can have major consequences if not promptly treated.

A 24-year old woman presented to the hospital with pain and swelling of her right lower extremity (RLE), a rash on her right thigh and cyanotic right toes. Three weeks prior to admission the patient was diagnosed with an unprovoked deep vein thrombosis (DVT) of her RLE and pulmonary emboli, treated with enoxaparin, and then bridged to warfarin. Ten days prior to admission, she returned to the ED with worsening bilateral lower extremity pain and swelling, was found to have worsening of her RLE DVT and a new left lower extremity DVT. Her International Normalized Ratio (INR) was subtherapeutic at 1.7. Due to extensive clot burden, she underwent catheter-directed thrombolysis of the RLE DVT and received perioperative heparin. She was bridged to warfarin and discharged. One day later she returned to the hospital with excruciating RLE pain. Her exam revealed a new 4 x 5 cm irregular, erythematous lesion on the right anterior thigh, and her right foot toes were cyanotic. Bilateral dorsalis pedis pulses were located via Doppler. Laboratory studies revealed an INR of 6.8, prothrombin time of 89 sec (normal: 10.3 – 13.5 sec), PTT of 55.6 sec (normal: 26.7 – 36.2 sec), platelet count of 121 K/mm³ (two weeks earlier: 466 K/mm³), plasma D-Dimer of 67,196 ng/mL (normal: <230 ng/mL), Factor VII of 0.01 U/mL (normal: 0.50 – 1.50 U/mL), and protein C of 0.23 U/mL (normal: 0.78 – 1.32 U/mL). She was administered vitamin K and fresh frozen plasma, and started on argatroban for suspected heparin-induced thrombocytopenia (HIT). She was admitted to the Burn Unit and received pain control with hydromorphone. The HIT antibody test was strongly positive. Within two days, the cyanosis of her toes resolved; however, her right anterior thigh lesion continued to evolve, becoming multiple large hemorrhagic bullae. She underwent anterior thigh skin debridement with an autologous skin graft. Pathology from the thigh revealed fibrinoid thrombi in the dermal and subcutaneous venules, and the patient was diagnosed with venous limb gangrene associated with warfarin in the context of HIT.

This case demonstrates venous limb gangrene, a rare clinical entity associated with warfarin-induced imbalance between procoagulant and anticoagulant mechanisms in the context of heparin-induced thrombocytopenia. Prompt recognition and treatment, including cessation and reversal of warfarin, as well as systemic anticoagulation with a non-heparin agent, can save both life and limb.

Comparing Cyclophosphamide with G-CSF and Plerixafor with G-CSF as Stem Cell Mobilization (SCM) Regimens for Multiple Myeloma (MM) Patients: An Evaluation of Mobilization Efficacy and Toxicity

Tan CR, Pancari PA, Ulicny J, Martin ME, Barta SK, Kropf PL, Fung HC

Background: The ideal regimen for SCM in MM remains controversial. Chemomobilization with cyclophosphamide (CY) and G-CSF (G) remains the most commonly used regimen. However, studies suggest that plerixafor (P) with G may be superior or equally effective as CY/G. We compared the efficacy and toxicities associated with CY/G vs P/G in MM pts at our institution.

Methods: From 1/2012 – 5/2015, 80 pts received CY/G or P/G for SCM. Demographics, D1 CD34+ cell yield, total CD34+ yield, toxicities, and time to engraftment were analyzed.

Results: Of the 80 pts, 35 had CY/G and 45 received P/G. The median D1 CD34+ cell yield and total yield were not different between the 2 groups. There was 1 poor mobilizer ($< 2 \times 10^6$ cells/kg in ≤ 3 aphereses) in the CY/G arm and none in the P/G arm. The number of good mobilizers and rapid mobilizers ($\geq 4 \times 10^6$ cells/kg on D1) in both arms were comparable. Most pts completed SCM in 1 session. CY/G was associated with more severe neutropenia (86% vs 0%, $P=0.01$), febrile neutropenia (6% vs 0%, $P = 0.31$), hospitalization (14% vs 0%, $P=0.08$), and delay in apheresis (14% vs 0%, $P=0.08$). Median time to neutrophil and platelet recovery were similar in both groups.

Conclusion: Our data indicate that CY/G and P/G result in robust SCM in most MM pts. Both regimens were comparable in efficacy and efficiency. However, P/G was better tolerated and had a more predictable course. Therefore, P/G may be considered as the preferred SCM regimen in MM.

FDG PET Positive Pituitary Macroadenoma in a Patient with GIST

Vasudevan S, Hall M, Veloski C

Case: A 60 y/o man with distant history of acoustic neuroma presented to ER with SOB for 1 month. CT chest showed pneumonia, 4 x 4 cm mass in left upper quadrant and multiple hypodensities in liver. CT guided liver biopsy diagnosed GIST. Molecular analysis of the GIST was positive for KIT mutation; hence he was treated with Imatinib. Staging FDG PET/CT revealed increased uptake in pituitary. MRI brain revealed a 4.1 x 2.5 x 3.4 cm sellar mass. Labs revealed low TSH, low free T3, low cortisol, normal prolactin, normal GH, normal IGF-1, low FSH, low LH and low total testosterone, consistent with hypopituitarism. He was treated with levothyroxine, steroids and testosterone. He subsequently underwent transsphenoidal resection of the pituitary mass.

Discussion: Pituitary incidentalomas are asymptomatic lesions. The normal pituitary does not accumulate FDG. Incidental pituitary uptake on FDG PET/CT is a rare finding with most cases diagnosed as clinically non-functioning PA. One study showed FDG uptake with SUVmax > 2.4 was 94.7 % sensitive and 100 % specific for PA. In this case, the finding of the PA in association with metastatic GIST positive for a KIT mutation may suggest a genetic association. PA has been reported in association with GIST usually in the setting of SDH germline mutations. Interestingly, KIT expression has been described in a subset of PA although no mutations were found in exons 9, 11, 13 and 17 of KIT gene. We have recommended mutational analysis of the PA and genetic testing.

Submandibular Mass: To be or not to be Ectopic Thyroid Tissue

Viswanathan L, Vaz CL, Bawa S, Figueiredo V

Introduction: Ectopic thyroid tissue usually results from faulty embryogenesis and migration of the thyroid gland. Cases of ectopic thyroid detected in the lateral cervical region were regarded as malignant (metastatic) lesions. Here we present a case of a lateral ectopic thyroid nodule.

Case presentation: A 83 year old female with past medical history of coronary artery disease, hypertension, type 2 diabetes mellitus and COPD had presented for evaluation of an incidentally palpated thyroid nodule. She denied dysphagia, palpitations or tremors. She denied family history of thyroid malignancy or prior radiation exposure. On US imaging, multinodular goiter with benign findings were noted. During follow up she reported an enlarging right submandibular mass over 4 months but clinically euthyroid. CT neck with contrast revealed a peripherally enhancing soft tissue mass - 2.3 x2.7cm abutting right thyrohyoid muscle. This was confirmed with US to be a 2.7x2.3x2.6 cm heterogenous mass with peripheral vascularity likely pedunculated thyroid nodule. FNAC was benign consistent with a dominant nodule in multinodular goiter.

Discussion: Lateral ectopic thyroid gland is formed when the cells of the lateral anlage do not join those of the median. Such ectopic tissue either present as a lateral, palpable, mobile, painless mass or become evident during adolescence or pregnancy. Our case is unusual due to the aggressive size increase suspicious for malignancy. Thyroid cancer metastases should always be excluded. Complete surgical resection is recommended based on size, local symptoms and potential of malignant transformation. In asymptomatic cases regular follow-up is adequate.

Burkholderia Cepacia Bacteremias in a Bone Marrow Transplant Unit Associated with Contaminated Water Baths Used for Stem Cell Thawing

Weber D, Want T, Fung H, Axelrod P

Two non-neutropenic autologous stem cell transplant recipients in a bone marrow transplant unit had *Burkholderia cepacia* bacteremia during a 2 day period in August 2015. Patient A had multiple myeloma; she developed fatigue and a temperature of 102.2F three days following her transplant. Three of 3 blood culture sets that day grew *B cepacia*, and 1/2 was positive 48 hours later. Stem cell cultures before transplant had no growth. Patient B also had myeloma and three days post-transplant she had a temperature of 100.9F. Similarly, 3/3 sets of blood cultures that day grew *B cepacia* and 3/3 did two days later. Catheters were removed from both patients; one of two catheter tips grew *B cepacia*. Both patients received antibiotics and recovered. De-ionized water from a water bath used to defrost stored, frozen stem cells grew *B. cepacia* as did 5 swab cultures from water bath surfaces. Cultures from containers and surfaces used in stem cell storage and transport had no growth or grew another organism. De-ionized water prepared in a dedicated sink had no growth, but surfaces of a plastic jug used to transport water from this sink to the water bath grew *B cepacia*. This jug was not routinely cleaned between uses. *B cepacia* strains were molecularly typed using pulse field gel electrophoresis. Patient and environmental samples had identical molecular fingerprints. Sterile water replaced de-ionized water and no further cases occurred. Sterility is crucial for all products in contact with IV infused materials, including stem cells.