## **Table of Contents**

7	ABSTRACTS A time and charge analysis of the management of incomplete abortion with a manual vacuum aspirator in the emergency department versus electric suction curettage in the operating room
7	Autonomic Dysfunction in Complex Regional Pain Syndrome: A possible relationship with the GI tract?
7	Bowel Wall Thickening on CT Scan Is a High-Yield Colonoscopic Indication
8	Discordant Results of Biochemical and Genotypic Identification of an Unusual Bacterial Isolate
9	Do Not "Forget" about Heavy Metals
9	"Double Positive" Anti-glomerular Basement Membrane (GBM) Antibody and Myeloperoxidase Anti Neutrophil Cytoplasmic Antibody (MPO-ANCA) in a Patient with Goodpasture's Disease
10	Evaluation of pleural and peritoneal effusions in 541 patients with advanced malignant tumors
10	Factors Associated with Increased Mortality in Patients Undergoing Extraction of Cardiovascular Implantable Electronic Devices Due to Infection
11	Follicular Lesion of Undetermined Significance (FLUS): A Four-Year Institutional Analysis
11	Functional Near-infrared Spectroscopy Effectively Distinguishes Optimal Levels of Anesthesia During Colonoscopy
11	Gender preference for endoscopists among US Muslims: a population and acculturation based survey
12	Here or St. Elsewhere: Does the cost of medical liability correlate with resident migration after graduation?
12	Improving outcomes in obstetrical hemorrhage by implementing a multidisciplinary team management and a delivery planning checklist
12	Increasing the Follow-up Rate after Hysteroscopic Sterilization
13	Maternal Plasma, as a Source for fetal DNA, is better than Fetal Cells for Noninvasive Prenatal Diagnosis of Single Gene Disorders
13	Multidisciplinary Rounds on Labor & Delivery Improves Team Communication and Solidarity which Could Enhance Patient Safety

- 13 Risk Stratification of Lead Extraction in Octogenarians Using the Charlson Risk index
- Single cell cytomorphologic pattern of metastatic breast carcinoma in pleural and peritoneal effusions, a diagnostic pitfall
- 14 Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy to improve compliance with national safety & Department of the Team based strategy in the Te
- 14 The LGL registry: A review of 79 patients with large granular lymphocytic leukemia and Rheumatoid arthritis

- The use of a domestic violence (DV) screening card to improve screening and referral rates for DV in a resident continuity clinic. A quality improvement project
- Treatment Challenges in the Management of Cryptococcal neoformans Meningitis in HIV Patients in a University Hospital in Philadelphia
- Where there is sodium there may be sepsis
- Why do Patients Bounce Back? Findings of Reasons for Readmission among Patients Age 65 and Older in a Community Hospital

## CASE REPORTS AND REVIEWS

- 18 A Rare Case of Cytomegalovirus Colitis Mimicking Ischemic Colitis in an Immunocompetent Patient
- 18 A Rare case of Myeloid Sarcoma Presenting as an Anorectal Ulcer in a Pregnant Woman
- 19 A Rare Case of Pharyngitis: Lemierre's Disease
- A rare condition causing a grave problem: Idiopathic hypertrophic pachymeningitis (IHPM)
- 20 A Unique Case of Refractory Iron Deficiency Anemia associated with H. pylori
- 21 Acute Pancreatitis after Starting Methimazole for Grave's Disease A Distinct Clinical Entity
- 21 ADEM or Aseptic Meningitis: an Enigmatous Clinical Presentation
- 22 An Interesting Case of Norwegian Scabies
- 23 Aneurysmal Bone Cyst of the Spine
- 23 Angiosarcoma of the Gastrointestinal Tract: A Rare Case
- Antithrombin III (ATIII) Concentrate for Treatment of Hepatic Sinusoidal Occlusion Syndrome in an Infant with Acute Lymphoblastic Leukemia (ALL)
- 24 Are Hyperacute T waves in chest pain an indication for an emergency PCI?
- 25 Botulinum Toxin in Treating a Child with Hirschsprung's Disease
- 25 Candida utilis: A Rare Pathogen
- 26 Constrictive Pericarditis in a 13 year-old after Mediastinal Radiotherapy
- 26 Delayed Onset Epidural Hematoma
- 27 Elevated CA 19-9 in a female with multifocal biliary strictures
- 27 Epithelioid Hemangioendothelioma (EHE) in a patient with CREST syndrome
- 28 Esophageal Stricture Post Alkali Ingestion
- 28 Fatty metamorphosis or calcification of the infarcted myocardium on Cardiac MRI
- Fibromuscular dysplasia presenting as a renal artery infarction in a 35 year-old female with no predisposing factor

- 29 Heerfordt's Syndrome: Rare variant of Sarcoidosis
- 30 Herpes Esophagitis in an Immunocompetent Patient Causing Anemia and Exertional Angina
- 30 Hydroxyurea Treatment for Stuttering Priapism in a Prepubertal Male with Sickle Cell Disease
- 31 Iatrogenic duodenal perforation treated with endoscopic placement of metallic clips
- 31 Infected Urachal Cyst resulting in an Umbilical-Urachal Sinus Tract in an Adult
- 32 Inferior Vena Cava (IVC) Leiomyosarcoma: A Rare Entity
- 33 Large Latero-Cervical Mass in a Teenager Proves NOT to be Lymphoma
- 33 Lyme Disease Presenting with Facial Palsy and Myocarditis Mimicking Myocardial Infarction
- 34 Negative Pressure Pulmonary Edema after Robotic—Assisted Radical Prostatectomy
- 35 Painless Acute Aortic Dissection (AAD): A Question Without an Answer
- 35 Palmoplantar Pustulosis
- 36 Posterior Reversible Encephalopathy Syndrome (PRES): a catastrophic but reversible side-effect of bevacisumab
- **36** Postpericardiotomy syndrome in an infant
- 37 Pott's Puffy Tumor (PPT): a forgotten complication of chronic sinusitis
- 38 Proximal protection device with flow reversal system for safer carotid artery stenting (CAS)
- 38 Recurrent Gastrinoma and Diffuse Cutaneous Metastasis, a unique case
- **39** Recurrent Syncope Think Outside the Box
- 40 Stroke in Fabry's Disease: A Disease Without a Clear Prophylaxis
- 40 SUNCT A Typical Case Of Short-lasting, Unilateral, Neuralgiform Headache Attacks
- 41 Superior Vena Cava Echocardiography Detecting Lead Fibrosis prior to Device Extraction
- 41 The Role of Clinically Applied Medical Geography in Diagnosis
- Tubal Occlusion with Adiana Prior to In Vitro Fertilization in a Patient with Hydrosalpinx and Multiple Abdominal Surgeries
- 42 Uterine Rupture
- 43 Vocal Cord Dysfunction (VCD)
- Water Immersion Endoscopy and the Use of Underwater Argon Plasma Coagulation in the Control of Upper Gastrointestinal Bleeding
- When a Simple Earache is not that Simple

## **ORIGINAL ARTICLES**

- 46 A Rare Case of Metachronous Skip Metastasis of Pancreatic Cancer to the Colon
- Anemia in Women of Child-Bearing Age in a Resident Community Care Clinic: A Study to Help Identify and Reach the National Goals
- 51 CT-imaging to Evaluate Radiation-induced Pre-clinical Myelopathic Changes
- 53 Direct Bilirubin Levels are usually Unnecessary in the Newborn Nursery
- 55 Endovascular Repair of Aneurysmal Aberrant Artery in Adult Pulmonary Sequestration
- Fecal Incontinence: Prevalence, Severity, and Quality of Life Data from an Outpatient Gastroenterology Practice
- Metastatic diffuse gastric carcinoma presenting as an acute cholecystitis report of an unusual case with review of literature
- MR Imaging Findings of Occult Ipsilateral and Contralateral Breast Cancer in Women with Recently Diagnosed Breast Cancer in a Community Setting
- Rare Case of Stercoral Perforation Requiring Subtotal Colectomy

#### MEDICAL ESSAY

Quick Response (QR) codes for poster presentations

#### **POEM**

**70** My Answer

## **REVIEW ARTICLES**

- 71 A Case of Idiopathic Chronic Colonic Pseudo-Obstruction
- 74 Congenital Absence of Inferior Vena Cava as a Rare Cause of Recurrent Deep Vein Thrombosis
- Non-acute Compartment Syndrome of the Lower Extremity as a Presentation of Cutaneous T-Cell Lymphoma: A Review of Literature and Case Report
- 79 Subacute Stent Thrombosis A Case Report and Review of the Literature
- **82** Toxicology of Clonidine

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## Cover Art submitted by Stephanie Johnson, Drexel University College of Medicine, Class of 2013

## **Editorial Policy**

The *DrexelMed Journal (DMJ)* features the scholarly activities of our graduate medical education trainees. This journal was created to highlight the many interesting and diverse scholarly activities and research at Drexel University College of Medicine and its participating affiliates (listed above). Recognizing that scholarly activity takes many forms, the *Journal* aims to publish all such efforts, and welcomes original research, reviews, case reports, and technical reports alike.

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If residents from another institution would like to participate, they should request their designated institution official (DIO) to communicate directly with the Vice Dean of GME at DUCOM: Dr. Mark Woodland (215) 762-3500.

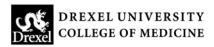
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### **EDITORIAL COMMENTS**

We are happy to present the sixth issue of the *DrexelMed Journal*, featuring the scholarly activities of the graduate medical education trainees of Drexel University College of Medicine, Hahnemann University Hospital, Abington Memorial Hospital, Allegheny General Hospital, Easton Hospital, Mercy Health System, Monmouth Medical Center, St. Christopher's Hospital for Children, St. Peter's University Hospital, and Virtua Health.

This edition of the *DMJ* includes again an impressive scope and range of types of projects, subject matter, and depth of research. Residents and fellows working with faculty in over fourteen specialties from eight institutions, not including collaborative physicians from outside of our GME scope, have taken the time to present their scholarly activity to us. This year's edition also includes a poignant medical essay related to use of technology and a touching medical essay.

We are very proud to bring you this edition of the *DrexelMed Journal*. We hope you take the time to peruse and appreciate the diverse richness of this year's edition and encourage you to submit your work for next year's edition!

David Berkson, MD

Editor-in-Chief Asst. Professor, Program Director, Family Medicine Mark B. Woodland, MS, MD

Program Director, OB-GYN Vice Dean, GME

Clinical Professor, OB-GYN

Jay M. Yanoff, EdD

Chief GME Officer, DIO Hahnemann University Hospital

## **DEAN'S RECOGNITION**

I offer my congratulations to Dr.'s Berkson, Woodland, and Yanoff for the 6<sup>th</sup> Edition of the DrexelMed Journal. Six years ago when Drs. Yanoff and Woodland initiated this effort, it was whole heartedly supported by the College of Medicine to emphasize the scholarly activities of our residents. At the time it was in line with the Strategic Plan of the College of Medicine and the research mission of the College of Medicine. Since then, the challenge has been to continue to expand beyond the halls of our primary GME affiliates and I am pleased to see the participation of our affiliates continues to expand in this edition.

Finally, my personal appreciation to the many residents represented in this journal and to those of you in training who have ongoing scholarly activities. At Drexel, academic inquiry through research and innovation is part of our basic mission. We hope throughout your training programs and your professional careers that you continue your endeavors to move medicine forward.

Daniel V. Schidlow, MD

Interim Dean, College of Medicine Drexel University College of Medicine

Abstract: A time and charge analysis of the management of incomplete abortion with a manual vacuum aspirator in the emergency department versus electric suction curettage in the operating room

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#### **OBJECTIVE**

To evaluate hospital time and charges associated with management of incomplete abortions using manual vacuum aspirator (MVA) in the emergency department (ED) versus electric suction curettage (ESC) in the operating room (OR). This study was conducted to determine if MVA in the ED is a more efficient and cost effective method of managing incomplete abortions.

## **METHODS**

After implementing a protocol for performing MVA in the ED for incomplete abortions we conducted a retrospective cohort study over a one-year period. The times of admission, start of procedure, discharge time and total hospital charges were recorded for each patient. Differences in times were analyzed with an unpaired t test and differences in charges were examined with a Mann-Whitney U test.

#### RESULTS

Twenty-five patient charts were reviewed. 15 patients underwent an ESC in the OR (group 1), 10 patients underwent a MVA in the ED (group 2). Average time to procedure was 7:46 (h:min) in group 1 and 7:03 in group 2 (p=0.683). Average time to discharge in group 1 was 10:51 and 10:46 in group 2 (p = 0.461). The average total charges were \$29,529.00 in group 1 and \$9,296.00 in group 2 (p <0.001).

## CONCLUSION

This pilot study demonstrates that surgical management of incomplete abortions using MVA in the ED is more cost effective. Our data failed to demonstrate a statistically significant difference in time to procedure or total hospital time. Of interest MVA has become the preferred method for surgical management of incomplete abortion at our institution.

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## Abstract: Autonomic Dysfunction in Complex Regional Pain Syndrome: A possible relationship with the GI tract?

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\*\*Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology

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#### **OBJECTIVE**

Complex Regional Pain Syndrome(CRPS) is a neuropathic disorder following minor trauma characterized by chronic pain, skin changes, and swelling. Literature supports that CRPS is a multi-organ disease with autonomic dysfunction but to our knowledge no study to date has examined its influence on the gastrointestinal tract.

#### **METHODS**

We conducted a retrospective chart review of CRPS patients seen at the DUCOM CRPS clinic from January 2001-September 2011. Each chart contained a questionnaire with a detailed review of systems containing nine GI symptoms which was completed by each patient.

#### RESULTS

Of 1053 CRPS patients that completed the questionnaire, 849(81%) had either thyroid disease or were on narcotics and were excluded. The remaining 204(19%) patients were included in the study. One hundred sixty nine(83%) were females and the mean age was 42.4. One hundred eighteen patients(58%) reported at least one gastrointestinal symptom. The most common symptoms were nausea 59(29%), constipation 48(24%), indigestion 42(21%), lack of appetite 35(17%) and dysphagia 35(17%). Nausea was the only symptom that was significantly more common in women than men(33% vs.12%,p=0.02). Location of injury did not statistically predict if CRPS patients developed upper vs. lower GI symptoms. CRPS duration of greater than three years was significantly associated with dysphagia(p=0.04) but no other GI symptoms. CRPS patients without thyroid disease that are not on narcotics still have a wide range of GI symptoms.

#### CONCLUSION

The connection between the autonomic dysfunction seen in CRPS patients and gastrointestinal motility is an area that needs further exploration given the plethora of GI symptoms seen in this study.

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## Abstract: Bowel Wall Thickening on CT Scan Is a High-Yield Colonoscopic Indication

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#### **OBJECTIVE**

Wall thickening in the lower GI tract (WTLGIT) is a common finding on computerized tomography(CT) and raises the concern for neoplasia.

#### **METHODS**

We performed a retrospective study of patients with WTLGIT

reported in CT reports between 2008-2010 to evaluate significance of these findings. Significant correlating colonoscopy/surgical findings were defined as: Polyps >1cm, masses, ulceration, edema, erythema, perforation, evidence of infectious colitis and diverticulitis. Subjects with prior lower GI diagnoses were excluded.

#### RESULTS

188/2505(7.5%) CT described WTLGIT. The mean age was  $(54.9\pm19.4)$  and 101(54%) were females. 105(56%) were African American and 63(34%) were whites. 70/188(37%) patients underwent colonoscopy and 14/188(7.4%) underwent surgery. 53/84(63%) had correlative thickening based on diagnostic intervention (colonoscopy 39/70(55.7%) and surgery 14/14(100%)). Patients with correlating intervention findings were significantly older (59.6  $\pm$  17.9y) than those without (48.2  $\pm$ 17.6y;p=0.006). African Americans 34/48(70%) were more likely to have correlated findings then whites 11/25(44%;p=0.04). Patients who did not undergo diagnostic intervention were significantly more likely to have a presumed or diagnosed infectious cause for WTLGIT [75/104(72%) vs. 40/84(47%);p<0.0009]. Patients with adenocarcinoma were more likely to have weight loss [5/7(71%)] than in those without cancer [5/77(6.5%)] who underwent diagnostic intervention and had a correlation with WTLGIT;(p=0.0002).

#### CONCLUSION

The majority of patients with WTLGIT had significant correlating findings. Older and African American patients had significantly higher rates of significant correlative pathological findings on diagnostic intervention. Hypoalbuminemia and symptoms, besides weight loss, did not correlate with WTLGIT. Weight loss was seen in the majority of those with cancer and may help stratify patients to undergo diagnostic intervention.

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## Abstract: Discordant Results of Biochemical and Genotypic Identification of an Unusual Bacterial Isolate

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## INTRODUCTION

A 68-year-old man developed left knee pain and fever after

admission for anemia. The joint was warm, swollen, and tender. Arthrocentesis was performed and blood cultures obtained. He was started on IV Vancomycin. Synovial fluid had 15,700 WBC/mm3 (85% neutrophils), monosodium urate crystals, but no bacteria. Both sets of blood cultures grew Gram-negative diplococci, identified as Neisseria mucosa by the RapID NH system. Antibiotics were changed to ceftriaxone. The patient improved clinically. Follow-up blood cultures remained negative and he was discharged on amoxicillin.

#### **OBJECTIVE**

To verify phenotypic identification of an unusual pathogen.

#### METHODS

16S rRNA gene amplification and sequencing of the isolate was performed in our laboratory. The sequence was compared to entries in GenBank using BLAST. The organism was also sent to the CDC for identification.

#### RESULTS

The portion of the 16S rRNA amplified in our laboratory most closely matched an uncultured unnamed bacterium in GenBank. In contrast, genotypic and phenotypic analysis performed by the CDC also using 16S rRNA amplification identified the organism as N. subflava, a species different from N. mucosa.

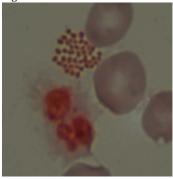
#### CONCLUSION

The RapID NH system, which is commonly used in clinical laboratories, has variable accuracy in identifying non-gonococcal, non-meningiococcal Neisseria spp. While 16S rRNA gene amplification is often considered the gold standard in identifying bacterial species, interpretation of sequencing results depends on the quality of the databases used. Public domains such as GeneBank may have erroneous information on unusual organisms. Identification of rare bacterial isolates should thus be verified in a reference laboratory.

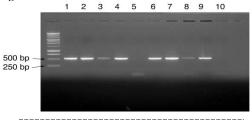
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Figure 1. Gram stain







Abstract: Do Not "Forget" about Heavy Metals

Srividhya Lakshmanan, MD, Mahesh Krishnamurthy, MD Easton Hospital: Department of Medicine

## INTRODUCTION

A 49 year Hispanic female presented with a 4 month history of widely fluctuating symptoms of forgetfulness, personality changes, memory loss, agitation, hallucinations and delusions. Her functional status was severely compromised. Examination revealed MMSE score of 11/30. The reminder of the physical examination was normal. Extensive work up including lumbar puncture, MRI of the brain and an EEG were negative.

A 24 hour urine collection for heavy metals revealed an arsenic level of 153 mcg/L (Normal < 100mcg/L). Retrospectively, the patient reported frequent consumption of seafood and use of alternative medications (homeopathic) from her home country of Colombia for digestive and skin problems. The patient was treated with DMSA at 500 mg twice per day for 2 weeks. She was told to stop taking the homeopathic medications and to avoid eating seafood. Her symptoms improved significantly.

#### CONCLUSION

Heavy metal poisonings are a rare, but preventable and reversible, cause of rapidly progressive neurological dysfunction. Arsenicals had been used medicinally to treat syphilis, skin conditions, trypanosomiasis and leukemias. I A number of homeopathic medications are known to contain arsenic. Peripheral neuropathy, encephalopathy, cognitive impairment, disorientation, hallucinations, agitations and memory problems are the main neurological effects of chronic arsenic toxicity.2,3,4 The most accurate test for arsenic exposure is a 24-hour urine arsenic level without shellfish for the prior 48 to 72 hours.

We emphasize the need for meticulous investigation for reversible causes of dementia with progressively declining cognitive functions even in the absence of clinical signs of heavy metal overdose.

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Abstract: "Double Positive" Anti-glomerular Basement Membrane (GBM) Antibody and Myeloperoxidase Anti Neutrophil Cytoplasmic Antibody (MPO-ANCA) in a Patient with Goodpasture's Disease

Julieta Gilson, MD, Robert Gayner, MD, Mahesh Krishnamurthy, MD, Nonihal Singh, MD, Dhavalkumar Sureja, MD

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#### INTRODUCTION

Goodpasture's syndrome is an uncommon condition causing acute renal failure due to rapidly progressive glomerulonephritis. Patients may present with isolated renal failure, lung hemorrhage or both. Approximately 10-38% of patients with anti-GBM antibody may have circulating ANCA, which is usually antimyeloperoxidase. Untreated disease is rapidly progressive and fatal. Cyclophosphamide, corticosteroids and plasmapheresis are the gold standard treatment. One-hundred-percent crescentic glomerular involvement on biopsy and early requirement of dialysis is generally associated with irreversible renal failure, even with aggressive therapy.

#### CASE

A 42-year-old female with a history of hypertension presented with vomiting, cough, and leg swelling of one week duration. She was found to have severe acute renal failure with a serum creatinine of 19 mg/dl. Urinalysis showed 4+ proteinuria and 5+ hematuria. Renal ultrasound showed a horseshoe kidney. Chest x-ray was normal. Emergent dialysis was performed. Further work-up revealed that she was positive for MPO-ANCA and anti-GBM antibody. Renal biopsy revealed a diffuse crescentic glomerulonephritis (Fig 1.) with segmental necrotizing features and 3+ intense linear anti-GBM staining with IgG (Fig 2). Additional findings were mesangioproliferative glomerulonephritis with predominately mesangial IgA deposits, tubular atrophy, and interstitial fibrosis. She received treatment with pulse solumedrol followed by oral prednisone, cytoxan and plasmapheresis without short-term response.

#### CONCULUSION

Goodpasture's disease typically presents as an acute renal failure accompanied by pulmonary hemorrhage. We present an uncommon case of "double positive" antibodies in a patient without pulmonary involvement. Early diagnosis is vital for improved outcome. Clinical outcome is poorly defined in such patients. Prognosis remains poor in dialysis dependent patients. 4,5

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Figure 1. Glomeruli containing circumferential crescent

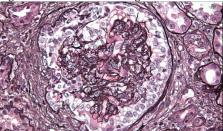
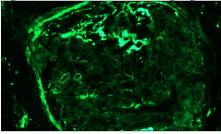


Figure 2. Immunofluorescence findings: IgG



Abstract: Evaluation of pleural and peritoneal effusions in 541 patients with advanced malignant tumors

Yuebo Gan, MD, Xiaoli Chen, MD, Cheryl Hanau, MD, Beth Mapow, DO

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## **OBJECTIVE**

In order to find cytomorphologic patterns and to reduce false positive and negative results, we analyzed retrospectively 541 cases of pleural (271) and peritoneal (270) effusions in the patients with advanced stage malignant tumors over the last 5 years.

#### RESULTS

Of the cases reviewed 45% were diagnosed as positive, 46% as negative and 9% as atypical cytology. In the peritoneal effusions ovarian carcinoma showed the highest rate of positive cytology (92%), followed by gastric (60%) and pancreatic cancer (49%), whereas hepatocellular carcinoma showed the lowest positive rate (3%). Pleural effusions had variable positive rates among breast (60%), lung (50%), ovarian (50%), pancreatic (43%), colonic (35%), and esophageal (31%) carcinomas. Immunohistochemistry was performed on 81 morphologically negative or equivocal cases and revealed malignant cells in 31 cases (38%). The malignancies with the high false negativity by morphology alone were breast carcinoma, gastric signet ring cell carcinoma, squamous carcinoma, well-differentiated neuroendocrine tumors, prostatic carcinoma, melanoma, and lymphoma. All of the cases that were reported as atypical cytology showed rare atypical cells that were too few to get an unequivocal immunohistochemical result. Seven cases with positive peritoneal or pleural biopsies repeatedly showed negative cytology. Among them, 6 showed marked desmoplastic changes.

## CONCLUSION

Some tumors show high false negative morphology and immunohistochemistry is diagnostically important for these tumors. Atypical diagnosis is mainly due to the rareness of atypical cells and a definitive diagnosis can often be obtained on repeat specimens. Tumor fibrosis may be implicated in cases with false negative cytology.

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Abstract: Factors Associated with Increased Mortality in Patients Undergoing Extraction of Cardiovascular Implantable Electronic Devices Due to Infection

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#### INTRODUCTION

With a rising number of cardiovascular implantable electronic devices (CIED) needed especially in the elderly population the burden of device-related infections is increasing. Antimicrobial therapy alone has an unacceptably high treatment failure rate. Thus device removal including lead extraction must be sought.

#### METHODS

We reviewed medical records of 400 patients who underwent device extraction for CIED-related infection at a tertiary referral center between 1991 and 2007.

#### RESULTS

The average patient age was 69 years. One-month and one-year mortality were 8 and 24%, respectively. Increased one-year mortality was associated with advanced age. Patients more likely to die within one year presented with fever (OR 2.0; 95% CI 1.2-3.1), hypotension (OR 3.7; 1.7-8.0), evidence of a vegetation on echocardiography (OR 2.7; 1.7-4.6), bacteremia (OR 2.8; 1.7-4.6), and Staphylococcus aureus infections (OR 2.3; 1.4-0.9). Other risk factors were referral from an outside institution (OR 1.9; 1.1-3.3), presence of chronic central venous catheters (OR 3.3; 1.5-7.4), hemodialysis (OR 3.8; 1.9-7.6), and secondary focus of infection (OR 2.8; 1.6-4.8). Contrastingly, local signs of swelling (OR 0.5; 0.3-0.9), erosion (OR 0.3; 0.1-0.5) and drainage (OR 0.5; 0.3-0.8) were associated with a survival benefit. After multivariate analysis, advanced age, chronic hemodialysis, and erosion remained statistically significant.

#### CONCLUSION

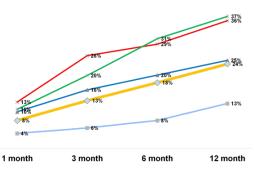
CIED are a known treatment modality for several cardiac conditions. When infections occur device removal is necessary. In our experience, infection and needed extraction/hospitalization were associated with complications. More emphasis must be placed on early detection of CIED infections and better prevention strategies should be sought for high-risk populations.

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Figure 1. Cumulative Mortality After Extraction

Pocket \*Pocket with bacteremia \*Occult bacteremia \*Endocarditits \*Total\*



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## Abstract: Follicular Lesion of Undetermined Significance (FLUS): A Four-Year Institutional Analysis

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#### **OBJECTIVE**

FLUS was defined as heterogeneous cytomorphologic findings neither convincingly benign nor sufficiently atypical for a diagnosis of "follicular neoplasm" or "suspicious for malignancy" on thyroid fine-needle aspiration (FNA). We investigated how frequently the FLUS term was used and the follow-up histopathologic findings.

#### **METHODS**

A computerized search of cytology records in our institution was performed from January 1st, 2008 to December 31st, 2011. All cases using the term FLUS were identified. Follow up corresponding surgical pathology reports were obtained for the FLUS cases. The frequency of FLUS cases and surgical resections of FLUS cases were evaluated.

#### RESULTS

A total of 839 thyroid nodule FNAs were examined with 80 (9.5%) of them classified as FLUS. A total of 76 patients, 60 females and 16 males from different racial background made up the cohort. There were 38 African Americans, 29 Caucasians and 9 others. Age range was from 25 to 95 years (mean 57 years). Surgical resections after FLUS were in 19 patients. Of those resected specimens, 3 (15.8%) had a malignant nodule (follicular variants of papillary carcinoma), 3 (15.8%) had a neoplastic nodule (follicular adenoma), and 13 (68.4%) had a multinodular goiter.

#### CONCLUSION

Our data indicated that the usage of the term FLUS is slightly more than the 7% recommended but is less than the average usage (12%) among other three institutions. Because the majority of the FLUS resection cases belong to non-neoplastic disease, clinical follow-up with a repeat ultrasound-guided FNA is appropriate as recommended. 1.3

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Abstract: Functional Near-infrared Spectroscopy Effectively Distinguishes Optimal Levels of Anesthesia During Colonoscopy

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#### INTRODUCTION

Safe colonoscopy is limited by the risk of over- and undersedation. Current technologies monitor complications of sedation. We developed a functional near-infrared spectroscopy (fNIR) technology that is a safe, non-invasive device which detects the hemodynamic response of the brain's frontal cortex to cognitive activation using specific wavelengths of light, introduced at the scalp, to detect changes in mean deoxygenated hemoglobin (deoxy-Hb) in the frontal cortex.

#### OBJECTIVE

We propose that fNIR can predict over-sedation and undersedation sooner than standard clinical signs in patients undergoing outpatient colonoscopy.

#### METHODS

We obtained consent from 30 patients undergoing colonoscopy. Synchronous recording of PAO2, expired CO2, BP, HR, and ECG tracing were recorded in addition to mean changes in deoxy-Hb by our fNIR device during sedation. Signs of wakefulness were recorded using the Ramsay Sedation Scale.

#### RESULTS

fNIR ratios during wakefulness were easily distinguished from deep sleep. Twenty one events of unintended wakefulness were identified by a Ramsay score to <5, movement, or speaking. For each event, there was a statistically significant difference in mean deoxy-Hb levels in the one minute prior to wakefulness when compared to 2, 3, 4, and 5 minute mean deoxy-Hb levels prior to wakefulness (p=0.01,0.01,0.02,0.01 respectively). fNIR also appeared to identify patients who became over-sedated leading to hypoxemia, but we did not have enough of these adverse events to report at this time.

## CONCLUSION

fNIR is an exciting new technology that may provide a more optimal level of anesthesia during endoscopic procedures in the future

Abstract: Gender preference for endoscopists among US Muslims: a population and acculturation based survey

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#### **OBJECTIVE**

Gender preference for the endoscopist has been cited as being as high as 40% in women and 22% in men in a diverse US population. The aim of this study was to determine the gender preference for endoscopist in US Muslims.

#### **METHODS**

Adult Muslims from three community centers completed a questionnaire including demographics, country of origin, length of stay in US, income level, education level and gender preferences toward endoscopist. The validated Vancouver Index of Acculturation (VIA) survey was administered to determine level of acculturation.

## RESULTS

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159 Muslims completed the questionnaire of which 91(57%) were men and 68 (43%) women. Gender preference for endoscopist in our Muslim population was 65.4%. Muslim women had a significantly higher gender preference for endoscopist as compared to Muslim men (82.3% vs. 52.7%; p value < 0.0001). One hundred percent of women and 94.4% of men with gender preference expressed preference for same gender endoscopist. Marital status, age, educational level, and socioeconomic status did not have a significant influence on gender preference for endoscopist. Muslims who have lived in US for >10 years had a higher gender preference for endoscopist as compared to those who have not (68.9% vs. 51%; p value=NS). Analysis of VIA score shows that 67.9% Muslims had a high heritage score; 18.8% had a high mainstream score. Women had high heritage score as compared to men (72% vs. 64.8%; p=NS). Muslims with a high heritage score tend to have a gender preference as compared to those with a high mainstream score (66% vs. 53%; p = NS).

#### CONCULUSION

Both Muslim men and women have a strikingly high gender preference for endoscopist. Muslim women who predominantly associate themselves with their heritage almost always prefer to have a woman perform their endoscopic procedures.

# Abstract: Here or St. Elsewhere: Does the cost of medical liability correlate with resident migration after graduation?

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#### **OBJECTIVE**

To establish resident retention rates in each state over a 7 year period and to determine if there is a correlation between resident retention and the cost of medical liability premium.

#### METHODS

We conducted a descriptive study of graduating fourth year residents from 2004-2010. The data was obtained from CREOG and the ACOG office of medical liability. Percentage retention was calculated for each state using PGY4 CREOG data. For each year, the top 5 and bottom five medical liability premiums states were identified using data obtained from ACOG. Regression analysis was used to calculate the difference in resident retention rate by high and low medical liability base rate premiums.

## RESULTS

The overall national resident retention rate from 2004-2011 was 48%. High premium states had a 15% lower retention rate when compared to the low premium states. This result was statistically significant (p<0.0001).

## CONCLUSION

The results of the study suggest that the cost of medical liability affects resident retention. However a direct causal relationship cannot be determined using the available data. In a time where the conversation about the cost of medical liability and its effect on physician migration is at the forefront of health care reform, it is important for further studies to be conducted regarding this topic.

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Abstract: Improving outcomes in obstetrical hemorrhage by implementing a multidisciplinary team management and a delivery planning checklist

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#### INTRODUCTION

Obstetrical hemorrhage provides multiple challenges to any Labor Unit with a high risk of morbidity and mortality including hysterectomy.

#### **METHODS**

In cases identified at greatest risk of hemorrhage (previa and/or accreta) prior to admission, a multidisciplinary team-based approach to managing/preventing hemorrhage was implemented to improve maternal outcome. The team included representation from anesthesia, nursing, obstetrics, gynecology oncology, interventional radiology, perfusion therapy, and blood bank. In addition, a pre-admission/intrapartum checklist was implemented to ensure communication and coordination of care among team members. Using hysterectomy as a marker for severe hemorrhage, we reviewed all cases of obstetrical hemorrhage resulting in cesarean hysterectomy from January 2009 to January 2012 before and after implementation of our team-based management protocol.

#### RESULTS

Fourteen obstetric cases of cesarean hysterectomy were identified from January 1, 2009 to January 1, 2012. In six of the cases, management involved implementation of the multi-disciplinary team-based approach with a preadmission/intrapartum checklist coordinating care. There was no difference between the team-based management group vs the group without the teambased management in regards to age, BMI, OR time, or length of stay. Those patients managed utilizing the team-based approach had significantly lower blood loss (2283 +/- 580 ml vs 7643 +/- 2022 ml; p=0.037); less units of blood transfused (4.7 +/- 9.0 units vs 29.4 +/- 24.2 units; p=0.038) and shorter length of stay in the ICU (0.3 +/- 0.5 days vs 1.4 +/- 0.8 days; p=0.014).

#### CONCLUSION

A multidisciplinary team approach to managing patients at high risk for obstetrical hemorrhage utilizing a preadmission/intrapartum checklist should be expected to improve pregnancy outcome.

## Abstract: Increasing the Follow-up Rate after Hysteroscopic Sterilization

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#### INTRODUCTION

Risk of pregnancy after Essure placement, with confirmation of tubal occlusion, has been estimated to be about 0.1%. Failure to follow-up in 3 months to evaluate tubal patency has been associated with the majority of unintended pregnancies after Essure. Steps have been made to promote compliance to reduce the failure rate associated with departure from the follow-up protocol.

#### **OBJECTIVE**

The objective of this study was to evaluate if a computer generated scheduling reminder increases the hysterosalpingogram (HSG) follow-up rate after Essure placement. Our goal was to compare follow-up rates before and after the reminder system was put into practice.

#### METHODS

We performed a retrospective chart review comparing HSG follow-up rates before and after implementation of a computer generated reminder system, in a University teaching hospital. Group 1 consisted of patients who underwent Essure sterilization before July 2009 and Group 2 consisted of patients from July 2009 to December 2011. Data were collected using our hospital and outpatient electronic medical records.

#### RESULTS

A total of 157 women underwent an Essure procedure between January 2005 and December 2011, in both the office setting and outpatient surgical unit. 98 were performed before July 2009 and 59 after July 2009. Sixty-four percent were compliant with their HSG follow-up in Group 1 compared to seventy-five percent in Group 2. Failure to recall the need to schedule a HSG in 3 months accounted for the majority of protocol noncompliance.

#### CONCLUSION

The findings of this study suggest that HSG follow-up rates increase after implementation of a computer reminder system.

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Abstract: Maternal Plasma, as a Source for fetal DNA, is better than Fetal Cells for Noninvasive Prenatal Diagnosis of Single Gene Disorders

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## OBJECTIVE

Noninvasive prenatal diagnosis using fetal material in maternal blood would provide a safe and accurate alternative to prenatal diagnosis by amniocentesis or CVS. Our aim was to determine the source of fetal material that was reported to achieve higher accuracy of NIPD of single gene disorder.

#### METHODS

Pubmed search of the articles describing prenatal diagnosis of single gene disorders from maternal blood published to date were reviewed. We tabulated the disorders and the source of the sample which was obtained from maternal blood. The accuracy of the diagnosis, combined sensitivity, specificity, false positive rate, and false negative rate were calculated for all studies.

#### RESULTS

We identified 43 articles describing 15 single gene disorders. The source of fetal material was maternal plasma and fetal cells in 28 and 15 of the studies, respectively. The noninvasive prenatal diagnosis from maternal blood of the following conditions was described: achondroplasia, sickle cell disease, hemoglobinopathies, myotonic dystrophy, congenital adrenal hyperplasia, cystic fibrosis, Huntington disease, spinal muscular atrophy and metabolic diseases. Accurate diagnosis of single gene disorder was achieved in 187/218 (86%) of the samples using fetal cells and in 258/265 (97%) of the samples using fetal DNA.

## CONCLUSION

NIPD of single gene disorders using fetal DNA obtained from maternal plasma achieved higher accuracy compared with fetal DNA obtained from fetal cells isolated from maternal peripheral blood. This higher accuracy demonstrates the potential of NIPD using fetal DNA to be clinically applicable and possibly replace the traditional methods

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# Abstract: Multidisciplinary Rounds on Labor & Delivery Improves Team Communication and Solidarity which Could Enhance Patient Safety

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#### INTRODUCTION

Unpredictable events in the context of a multi-disciplinary unit will undoubtedly pose as an avenue to error in communication, an avoidable risk that can compromise patient care. Literature on formal multidisciplinary communications is scant, but it has reportedly been used in Internal Medicine, as well as Intensive Care Units with success.

#### METHODS

During July 2011, pre-rounds and post-rounds questionnaires were distributed during multidisciplinary rounds on labor and delivery to assess the opinions of each disciplinary team (Labor & Delivery, Antepartum, NICU, Anesthesia) on their rounding experience. A scoring system using scores of 1 through 5 (completely disagree through to completely agree) corresponding with each statement on the questionnaire was used. The statements were kept the same for both pre- and post-rounds. Five statements were used. For example, "I am comfortable with my clinical knowledge of the patients".

#### RESULTS

All teams in total scored higher on the post-rounds questionnaire, as well as on each individual item, when compared to the prerounds questionnaire (n=27). However, when the data was dissected into individual specialties, Anesthesia scored relatively the same pre- and post-rounds, 82 versus 84 (n=4), respectively.

## CONCLUSION

The unit as a whole feels rounds are helpful. This may improve patient safety on labor and delivery via better communication. More Anesthesia participants are needed to decipher whether the department finds multidisciplinary rounding to be unnecessary. Future research could collect and analyze data on patient errors made before and after implementation of multidisciplinary rounds to search for an association, whether it would be positive, negative, or absent.

## Abstract: Risk Stratification of Lead Extraction in Octogenarians Using the Charlson Risk index

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## INTRODUCTION

The indications for implantable cardiac devices are expanding to larger patient (pt) populations, especially for the elderly. By 2050, elderly populations will increase by 125%. Our aim was to explore the use of comorbidity risk stratification to the degree of complications, implant durations, and extraction tool selection in octogenarian pts.

## METHODS

We performed a single center retrospective review of 247 pts that underwent lead extractions from 2009 to 2011, including 26

octogenarians. Pts were risk stratified by using the Charlson Risk Index to determine their risk class (Table 1). Chi square analysis was used to compare their post procedure complications, which we classified as none, mild, major, and death.

#### RESULTS

This population consisted predominantly of male pts (18:8 M:F) with an average age, ejection fraction, and implant duration of 83.4 years, 41%, and 51.18 months, respectively. The population was classified as medium-high risk with 18 pts having none, 3 mild, 3 major, and 2 30-day mortality complications. Tool sets for extraction consisted of 38 mechanical sheaths and 8 lasers sheaths. Correlation of risk category and complication rate showed a chi square value of 2.837 which correlates to a p value of 0.457.

#### CONCLUSION

In octogenarian pts undergoing lead extraction, comorbidity risk stratification using the Charlson Index does not correlate to the degree of post procedure complication.

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Table 1. Risk Stratification

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	Low	Med	High	Very	Total	
	Risk	Risk	Risk	High	n=26	
	n=4	n=13	n=7	n=2	11-20	
Age (yrs)	83	83.7	83.1	83.5	83.4	
Male Gender	1	10	5	2	18	
LV EF (%)	58	41	38	28	41	
Charlson RI	0	1.54	3.43	7.5	2.27	
Minor Comp	1	2	0	0	18	
Major Comp	1	2	0	0	3	
Death	0	1	1	0	2	
Laser	2	3	2	1	8	
Mech Sheath	6	19	12	1	38	

Abstract: Single cell cytomorphologic pattern of metastatic breast carcinoma in pleural and peritoneal effusions, a diagnostic pitfall

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#### **OBJECTIVES & METHODS**

Metastatic invasive ductal and lobular breast carcinomas in body cavity fluid often have distinct cytomorphologic patterns. Ductal carcinoma usually shows tightly packed clusters, so-called cannonballs, which are readily recognized. A single cell arrangement or mesothelial-like pattern is characteristic of lobular carcinoma and morphologically deceptive. However, we found that ductal carcinoma could occasionally present as single cell pattern in the effusions which might be missed without immunohistochemistry. To further characterize this unusual cytomorphologic pattern, we reviewed retrospectively pleural and peritoneal effusions from 98 patients with advanced breast carcinoma diagnosed in our institution over the last 5 years.

## RESULTS

Malignant cells were identified in 42 of 67 (63%) pleural effusions and 11 of 31 (35%) peritoneal effusions. Of 53 malignant effusions, 46 were from ductal carcinoma and 6 were lobular carcinoma. All of the lobular carcinoma had single-cell pattern

with weak or negative E-Cadherin expression. Interestingly, 8 of 46 (17%) ductal carcinoma also showed single-cell pattern and the malignant cells were revealed by immunohistochemistry for Calretinin, ER, PR, Pan-CK or MOC31. The tumor cells in these 8 cases show common features of high nuclear grades (5 grade 3, 3 grade 2) and decreased expression (weak or negative) of E-Cadherin in 6 cases.

#### CONCLUSION

Our results indicate that a significant portion of metastatic breast ductal carcinoma in effusions can present as a single-cell pattern which might be related to decreased E-Cadherin expression and high nuclear grades. Immunohistochemistry is important in these cases to avoid false negative results.

### Abstract: Team based strategy to improve compliance with national safety & amp; practice quidelines

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#### **OBJECTIVE**

We sought to develop a multi-disciplinary team-based perinatal strategy though education, monitoring and implementation of tools geared to improving compliance with national guidelines.

#### METHODS

A multi-disciplinary team consisting of Anethesia and Obstetrical attendings, residents and nursing was established to improve and monitor compliance with Joint Commission and ACOG perinatal safety and practice guidelines. These guidelines include eliminating elective delivery before 39 weeks; administering correct pre-operative antibiotic and DVT prophylaxis; appropriate use of antenatal steroids, and performing an preinduction/augmentation checklist (evaluation of fetal heart rate, Bishop score, pelvic adequacy, uterine contraction pattern and estimated fetal weight) to ensure safe utilization of these techniques.

## RESULTS

2792 deliveries were reviewed from August 2010 to June 2011. Elective deliveries without a medical indication dropped from 3.1% to 0. The percent of patients receiving antibiotic prophylaxis prior to cesarean increased from 94% to 99%. DVT prophylaxis improved from 2.7% to 87.3%. The percent of physicians performing all the required elements of the preinduction/augmentation checklist before utilizing these management techniques rose from 34% to 92.5%. Documentation regarding pelvic adequacy and estimated fetal weight had the most opportunity for improvement (69% and 53% compliance respectively). Although all pre-term patients received antenatal steroids, the percent of patients with appropriate documentation improved but fluctuated from 53% to 93%. In addition, surgical site infections dropped from 1% to 0.63% with a direct cost savings of approximately \$4100 over the time period.

#### CONCLUSION

Implementation of a multi-disciplinary team approach including residents, Anesthesia and Obstetrical attendings and nurses can be utilized successfully to develop, implement and monitor a perinatal safety strategy geared towards improving compliance with Joint Commission and ACOG practice guidelines.

Abstract: The LGL registry: A review of 79

patients with large granular lymphocytic leukemia and Rheumatoid arthritis

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#### INTRODUCTION

The purpose of this study is to analyze patients enrolled in the LGL leukemia registry in order to distinguish the similarities between LGL leukemia and rheumatoid arthritis in order to access overlapping immune mechanisms that may be responsible for neutrophil mediated destruction. <sup>1,2,3</sup>

#### **METHODS**

A retrospective chart review was performed on 79 patients enrolled in the LGL registry at Penn State Cancer Center. All patients enrolled in the study had a diagnosis of both rheumatoid arthritis and potentially LGL leukemia. Data was collected for age, sex, family history, autoimmune disease, T-Cell receptor gene rearrangement, and bone marrow invasion.

## RESULTS

Of 79 patients the mean age of onset for LGL leukemia was 60 years old with no discrepancy noted between sex; 37M, 42 F. 27 patients had rheumatoid arthritis in a first degree relative with no discrimination between maternal or paternal inheritance. 22 patients were positive for any other autoimmune process. 60 patients were positive for T-cell receptor gene rearrangement. Of the remaining patients that were negative for TCR, bone marrow infiltration was evident to conclude a diagnosis of LGL. 2 patients were positive for NK LGL (Table 1).

#### CONCLUSION

Patients with LGL leukemia and rheumatoid arthritis appear to be clinically similar with regard to age, sex, and patterns of inheritance. There also appears to be a significant overlap between other autoimmune disorders. It is conceivable that the association of LGL leukemia and rheumatoid arthritis may result from shared genetic alterations as demonstrated in this present study population.

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Table 1

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	LGL/RA	Bone Marrow				
	No (%)	No (%)				
N	79	74				
TCR +	60 (75%)	72 (97%)				
TCR-	19 (24%)	12 (16%)				

Abstract: The use of a domestic violence (DV) screening card to improve screening and referral rates for DV in a resident continuity clinic. A quality improvement project

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#### **OBJECTIVE**

The American Academy of Pediatrics recommends that pediatricians routinely screen caregivers for DV. Unfortunately, DV screening in front of children older than 3 years old is not recommended because the child may disclose conversational content to the perpetrator. Addressing this challenge, we implemented a DV screening card for resident use. We hypothesized that DV screening cards, as adjuncts to verbal DV screening, would improve DV screening and referral rates in resident clinic.

#### **METHODS**

A laminated 4x6 inch DV screening card, containing a modified version of the 4-item HITS© questionnaire (English and Spanish), was placed in patient rooms of a resident continuity clinic. Residents were instructed to administer the screening card when caregivers presented with children older than 3 years-old. Caregivers were asked to silently read the questions and respond with "yes" (positive DV screen) or "no" (negative DV screen). Changes in DV screening rates were determined by resident report through online surveys. DV referral rates were measured by absolute number of DV victims referred to the DV counselor before and after implementation of the screening card.

#### RESULTS

Forty-two percent of residents reported using DV screening cards within 3 months of implementation. Self-reported DV screening rates of caregivers with children older than 3 years-old were unchanged. Referrals to the DV counselor increased after the intervention (Fig 1). After 3 months of implementing DV screening cards in resident clinic, DV screening rates were unchanged however referrals increased.

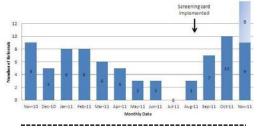
#### CONCLUSION

With more consistent use of DV screening cards, we expect improvement in DV screening and referral rates.

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Figure 1. Referrals to DV Counselor



Abstract: Treatment Challenges in the Management of Cryptococcal neoformans Meningitis in HIV Patients in a University Hospital in Philadelphia

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#### **OBJECTIVE**

To assess adherence of health care providers in a university hospital to published guidelines in the management of Cryptococcal neoformans meningitis in HIV patients.

## **METHODS**

This is a retrospective, single-center study. Data on demographics, HIV status, details on induction anti-fungal treatment, and lumbar punctures (LP) was collected on all adult HIV patients discharged with the diagnosis of cryptococcal meningitis between 2003-2010. Statistical analysis was conducted using Fisher's exact test and Mann-Whitney U test.

#### RESULTS

A total of 19 patients were included in analysis. Mean age was  $41(\pm\,9.9{\rm SD}).$  Mean absolute CD4 count was  $27(\pm\,33{\rm SD}).$  HIV status was known in 16 patients on admission but opening pressure on initial LP was documented in only 6/16 cases (38%). Patients who presented later in the study period (2008-2010) were significantly more likely to receive guideline appropriate induction therapy with amphoterecin B and 5-Flucytosine than patients presenting earlier in the study period (2003-2007)[80% vs. 22%, p=0.023] Adherence to guideline for induction therapy did not vary significantly by age, sex or renal function on admission. 10 patients (53%) had repeat LP.

#### CONCLUSION

Despite the importance of measuring opening pressure in HIV patients with Cryptococcal neoformans meningitis, a majority of patients (62%) did not have documented opening pressure. Interestingly, patients diagnosed after 2008 had a significant higher chance of receiving 5- Flucytosine. Excluding those who left against medical advice or refused LP, only a minority of patients (30%) did not have a repeat LP to document clearance of Cryptococcal neoformans from CSF.

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Abstract: Where there is sodium there may be sepsis

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## INTRODUCTION

The coincidence of sepsis and infection among patients who presented with hypernatremia was anecdotally observed to be high among patients admitted in a community teaching hospital. A

retrospective study was performed to ascertain if there is a positive correlation between presence of hypernatremia and sepsis.

#### METHODS

A Pennsylvanian community hospital's medical records from 2008 through 2010 were reviewed for patients with hypernatremia on admission. Data regarding age, sex, admission leukocyte count, admission serum sodium level, and presence of dementia were collected and analyzed with admission and discharge diagnoses.

#### RESULTS

In the three years of data 162 met the criteria for admission into the study. Among the patients which met criteria for participation, the median age was 82, 52.5% were male, 62.9% had leukocytosis on admission (mean of 14,145), mean sodium on admission was 155 mmol/l, and 86.4% had been previously diagnosed dementia. The incidence of infection among all the patients was 79.6%.

#### CONCLUSION

Our results show a striking correlation of the presence of hypernatremia on admission and sepsis. The incidence of sepsis did not increase, with any statistical significance, for increasing serum sodium, age, sex, or presence of dementia. We conclude that physicians should diligently search for infection in patients with hypernatremia on admission, if the infection is not apparent. The majority of infections diagnosed were pneumonia and urinary tract infections. Our proposed mechanism is the infection causes increased free water requirements and altered mental status leading to poor oral intake and decrease in water consumption leading to hypernatremia.

Figure 1. % Hypernatremic patients with infection

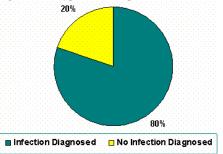
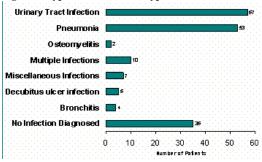


Figure 2. Types of Infection in Hypernatremia



Abstract: Why do Patients Bounce Back? Findings of Reasons for Readmission among Patients Age 65 and Older in a Community Hospital

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#### INTRODUCTION

Hospital readmissions among older patients have increased substantially over the past 30 years. The recently passed healthcare reform legislation aims at reducing readmissions and plans to penalize both hospitals and physicians who have higher than average readmission rates.

#### **OBJECTIVE**

To identify the reasons for readmission, then to examine whether there are clear factors associated with early versus late hospital readmissions. We hypothesized that patient's non-compliance is the major reason for all cause readmission.

#### METHODS

Chart review of patients age 65 and older who were admitted between December 1, 2008 and December 30, 2009.

#### RESULTS AND DISCUSSION

198 patients were included in the study. Healthcare associated infections represent the strongest predictor for hospital readmission (30%). Patients tend to return early if they have poor health on discharge and elders requiring skilled nursing and did not receive it bounce-back later. Non-compliance accounted only 14 %. Using these identified risk factors, patients potentially at high risk for

such events may be prospectively targeted to reduce readmission.

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## Case Report: A Rare Case of Cytomegalovirus Colitis Mimicking Ischemic Colitis in an Immunocompetent **Patient**

David Oustecky, MD

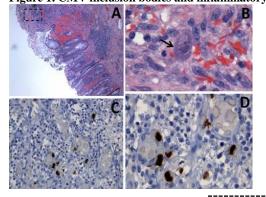
Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology

## CASE REPORT

A 78-year-old woman with a history of hypertension, umbilical hernia, osteoarthritis, postherpetic neuralgia, and asthma was admitted for diarrhea for a week and asthma exacerbation. Her diarrhea improved over the next 2 days. A week later, she had 3 episodes of bleeding per rectum with large clots. A limited colonoscopy showed multiple blood clots and stool mixed with blood. A CT scan showed distended proximal and transverse colon > 10 cm. A repeat colonoscopy because of continued bleeding showed edematous, ulcerated, friable mucosa in the sigmoid and descending colon. The biopsies showed ulcerated mucosa, granulation tissue, and acute inflammatory exudates. A laparotomy with left hemicolectomy and colostomy was done because of worsening abdominal pain and evidence of a perforation at the splenic flexure. A surgical biopsy revealed multifocal areas of ulceration, granulation tissue. hemorrhagic contents, transmural inflammation, and peritonitis. Viral cytopathic effects and positive staining consistent with CMV colitis were seen. Results of serological tests showed CMV IgG Ab 4.13; IgM Ab < 0.9; DNA PCR 16000 copies/ml; no HIV 1 or 2. She was given gancyclovir for 3 weeks.

DISCUSSION

CMV gastrointestinal disease ranges from ulcers of the oral Figure 1. CMV inclusion bodies and inflammatory infiltrates



cavity and pharynx to massive hemorrhage, toxic megacolon, and perforation of the bowels. It can also protract the course of IBD.<sup>2</sup> Endoscopy reveals mucosal erythema, ulcers, pseudotumor formation, and perforation.<sup>3</sup> Diagnosis can be established based on serological tests and biopsy. Treatment includes medical therapy supportive care and antivirals or surgery or a combination of both.<sup>4</sup>

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## Case Report: A Rare case of Myeloid Sarcoma Presenting as an Anorectal Ulcer in a Pregnant Woman

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#### CASE REPORT

A 30-year-old woman, 8 weeks pregnant with a family history of inflammatory bowel disease (IBD) and recurrent oral ulcers, presented with anal pain and bright red blood coating the stools. Examination revealed an atypical fissure. Non-operative measures failed and the symptoms worsened. Examination under anesthesia showed a large deep anal canal ulcer with no obvious fistula or abscess(Fig

1). Sigmoidoscopy revealed multiple aphthous ulcerations in the rectum and sigmoid colon consistent with IBD. Biopsies depicted ulceration with mucosal and submucosal aggregates of atypical hematopoietic cells(Fig 2). The peripheral smear showed 5% blasts. A bone marrow biopsy showed hypercellularity with 53% blasts consistent with acute monocytic leukemia (AML) on immunologic marker staining. CSF analysis showed 30% blasts. Karyotypes

were abnormal with Inv (16) (p13q22) and trisomy 8 and 22. She underwent a therapeutic abortion and was treated with idarubicin, cytarabine, and intrathecal methotrexate. She had no evidence of recurrent disease on a repeat bone marrow biopsy 9 months after completing therapy.

## DISCUSSION

Myeloid sarcoma, or chloroma, is a solid collection of leukemic cells outside of the bone marrow and can occur as solitary or multiple masses in different tissues or organs. <sup>1,2</sup> Although rare, GI tract involvement presents with abdominal pain, bleeding, perforation, obstruction or intussusception. <sup>3</sup> Endoscopic findings have been demonstrated throughout the GI tract and include erosions, ulcers and polypoid lesions. <sup>4</sup> CT imaging can show focal bowel wall thickening, exophytic lesions or a luminal mass or polyp. <sup>5</sup> Myeloid sarcoma is diagnosed with biopsy and immunohistochemical staining.

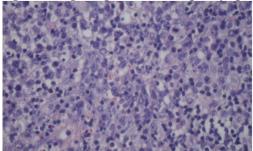
Figure 1. Large ano-rectal ulcer on exam under anesthesia



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Figure 2. Large irregular nuclei and pale cytoplasm



Case Report: A Rare Case of Pharyngitis: Lemierre's Disease

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## CASE REPORT

A 26 yo healthy male was admitted for decreased PO intake due to severe pharyngitis after failing a course of Azithromycin. Monospot testing was negative. Despite treatment with Decadron and Tylenol, fever continued over 102° F with episodic shortness of breath, progressive neck swelling and an erythematous macular rash of the face, neck and upper chest. Labs showed leukocytosis and a platelet count of 55 per mcL, improved to 100 with steroids. CT of the neck and chest demonstrated angioedema of the left side of the neck and numerous lung nodules up to 2.5 cm representing septic emboli. Ultrasound of the neck showed a thrombus in the left internal jugular vein. Blood cultures identified Fusobacterium necrophorum gram-negative rods. Lovenox, Coumadin and intravenous antibiotics were started. The patient was discharged home with IV Clindamycin and Flagyl for a six weeks course.

### DISCUSSION

Lemierre's Disease is often characterized by septic thrombophlebitis of the main neck vessels as the internal jugular vein and embolization to other organs. More cases have presented in the last few years. It is mostly due to Fusobacterium necrophorum infection with one third of the

cases from polymicrobial etiology and others from Staphylococcus aureus infections. Mortality rate is approximately 5 %. Complications are lung and brain abscesses, kidney failure, respiratory distress, septic shock and death. Treatment involves an average of six weeks of intravenous antibiotics with not enough evidence on the benefit of anticoagulation. Early detection and aggressive treatment are essential to decrease mortality.

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## Case Report: A rare condition causing a grave problem: Idiopathic hypertrophic pachymeningitis (IHPM)

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#### CASE REPORT

29-year-old Brazilian immigrant presented with complaints of one month h/o recurrent headaches associated with intermittent horizontal diplopia, ataxia, and parasthesias in upper extremities. Exam showed tetraparesis in all extremities, diffuse hyper-reflexia, and decreased sensation below C8-level. Cranial nerves exam normal. Four years ago patient had similar symptoms lasting for one week. CT, MRI was non-revealing at that time and she refused LP. Initial lab-work for CBC, CMP, ESR, CRP, ANA, RF, ANCA, Lyme's serology and RPR were negative. CT headunremarkable. MRI Brain and spine showed extramedullary dural thickening at base of skull and spine to T4 level compressing the spinal-cord. CSF analysis showed chronic inflammation and culture was negative. Diagnosis of idiopathic hypertrophic cranio-spinal pachymeningitis was made. Patient was treated with high-dose steroids and had decompression surgery involving excision of thickened meninges, C2-T5 laminectomies with dural augmentation.

#### DISCUSSION

IHPM was associated with tuberculosis and syphilis in the past and recently has been associated with some infections, autoimmune disorders and cancers. IHPM is a diagnosis of exclusion and is characterized by chronic inflammation of the dura and can mimic multiple sclerosis. Steroids and immunosuppressive agents are the main-stay of treatment in initial stages and are used to maintain remission. Radiation and surgical-decompression are used in some cases to relieve compression symptoms.

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## Case Report: A Unique Case of Refractory Iron Deficiency Anemia associated with H. pylori

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#### CASE REPORT

22 years old male with past history of insulin dependent diabetes and iron deficiency anemia presenting with fatigue, severe dyspnea on exertion and lower extremity swelling for 2 weeks .Vital signs HR 97 bpm, BP 104/54 mmHg, RR 16, O2 sat 99%. Physical examination revealed cachectic male with severe pallor, poor dentition, clear lung fields, systolic ejection murmur and lower extremity edema. Laboratory studies showing blood sugar of 658 mg/dL, hemoglobin of 4.2 g/dL, hematocrit 14.7 % and hemoccult negative. Patient admitted to the MICU, insulin drip started for treatment of diabetic ketoacidosis, then transfused 4 units of pRBCs and iron. Extensive hematological workup including bone marrow biopsy confirmed IDA with hypocellular marrow. Flow cytometry was negative for paroxysmal nocturnal hemoglobinuria (PNH). Gastroenterology workup including colonoscopy and capsule endoscopy with no lesions or source of

bleeding, EGD revealed thickened gastric folds, biopsies positive for H. pylori and negative for celiac disease. Patient was subsequently treated with triple therapy, hemoglobin improved and remained stable and patient was discharged .Outpatient follow up showed stable rising hemoglobin of 9.7 and 14.0 g/dL one/two months later.

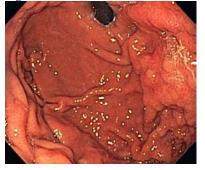
#### DISCUSSION

Unexplained IDA is a frequent indication for endoscopic evaluation of the GI tract. We present a unique case of severe unexplained IDA, refractory to previous multiple blood and iron transfusions, extensive hematologic workup without etiology, with EGD with prominent gastric folds and biopsies positive for H. pylori infection. Follow up hemoglobin normalized after H. pylori treatment and eradication. We recommend H. pylori testing/treatment for all cases with unexplained IDA.

Figure 1. Thickened gastric folds



Figure 2. Gastric nodularities



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## Case Report: Acute Pancreatitis after Starting Methimazole for Grave's Disease - A Distinct Clinical Entity

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#### CASE REPORT

A 47 year old African American female presented to our emergency department with a one month history of abdominal pain, nausea, recurrent vomiting and 21 pound weight loss. On admission, physical examination revealed diffuse enlargement of her thyroid gland, over which a bruit was heard. Thyroid function tests were as follows: TSH 0.004 miu/ml, T4 27.8 mcg/dl, T3 by radioimmunoassay > 800ng/dl, Thyroid stimulating immunoglobulin positive at 385%, thyroid peroxidase antibody negative. Her liver function tests were abnormal (Table 1). Serum lipase, amylase, acute hepatitis panel, celiac screen, SIEP, ANA, anti-mitochondrial and anti-smooth muscle antibody and alpha fetoprotein assays were normal. Serum ferritin was mildly elevated at 290ng/ml and IgA was also mildly elevated at 531mg/dl. CT and MRI revealed a 2 x 1.3 cm hepatic lesion consistent with an atypical hemangioma. Findings were consistent with Graves' disease and she was commenced on methimazole. Three days after discharge from hospital, she presented again with abdominal pain.

Her laboratory evaluation was significant for elevated amylase and lipase (Table 1) levels but CT of the abdomen showed no evidence of pancreatitis. On admission, her methimazole was held and her amylase and lipase levels trended towards normal levels. Methimazole was resumed two days after readmission and amylase and lipase rose sharply again on the day following reinstitution of her methimazole therapy (Table 1).

## DISCUSSION

Our literature search shows that this is the second case reported worldwide after Taguchi et al, and the first in the United States.

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Table 1. Trend of Lab Testing and Correlation with Methimazole Use

	7/8/11	7/7/11	7/6/11 Methim restarted	7/5/11	7/4/11 Readm Methim held	6/30/11	6/29/11
AST U/L)	164	212	223	339	403	258	221
ALT(U/L)	286	330	324	417	556	260	225
ALP(U/L)	100	104	87	91	123	80	100
Bilirubin (mg/dl)	0.6	1.1	1.1	1.2	1.2	1.0	0.9
Lipase (U/L)	257	583		127	341		42
Amylase (U/L)	159	221	78	96	196		47

## Case Report: ADEM or Aseptic Meningitis: an Enigmatous Clinical Presentation

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#### CASE REPORT

3-year-old male presented with headache and vomiting for 10 days. Headaches were not associated with vision changes, photophobia, or loss of consciousness. Two days prior to presenting, patient fell off a trampoline. Since headache continued, CT scan obtained and was normal. Patient returned to ER since symptoms continued and thus was admitted. Physical exam normal. CBC showed WBC 24,000, ESR 60, LDH 174. MRI findings consistent with ADEM (Fig 1). LP was done with WBC 33 and differential showed 71% lymphocytes and a high CSF pressure consistent with ADEM/aseptic meningitis, but protein<10 and glucose>60 favoring bacterial meningitis. Patient developed fever during hospital stay and started on ceftriaxone and acyclovir. Blood and CSF culture were negative. Lyme serology was negative. Patient was sent home on 3 weeks of antibiotics for possible Lyme meningitis.

#### DISCUSSION

ADEM usually is preceded by an infection or vaccination but presents with neurological symptoms but the development of fevers in this case is not associated with ADEM. MRI and CSF findings were consistent with ADEM /aseptic meningitis but there were absent neurologic symptoms or signs of meningismus. This patient presents as an enigma as these findings could be an early indicator of multiple sclerosis, but this is unclear and likely will not be able to be established.

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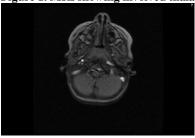
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Figure 1. MRI showing involved thalamus and basal ganglia



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## Case Report: An Interesting Case of Norwegian Scabies

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## CASE REPORT

A 49-year-old African-American male with no past medical history was admitted with cough and shortness of breath. On lung examination, he had crackles in the left lower lobe. Incidentally, multiple hyperkeratotic plaques were seen at the web spaces of his hands, wrists, axilla, nipples, abdomen, groin, ankles and feet. These non-pruritic lesions had developed three weeks prior to his presentation. Laboratory tests revealed anemia, lymphopenia and eosinophilia. Chest X-ray showed a left sided infiltrate consistent with pneumonia. Skin biopsy of one of his skin lesions showed Sarcoptes scabiei. He was placed in contact isolation and was treated with oral ivermectin and topical permethrin. He was not homeless nor an intravenous drug abuser but had multiple sexual partners in the past. HIV ELISA test was positive and his absolute CD4 count was below 50.

## DISCUSSION

Norwegian scabies, also known as crusted scabies, is a severe and a highly contagious form of infestation of the skin by the mite, Sarcoptes scabiei. This rare form of

scabies is seen in patients with HIV, leprosy or lymphoma. The lesions in crusted scabies begin as erythematous patches that develop into prominent scaly plaques characteristically seen on extremities. Pruritus is usually absent in these patients due to their diminished delayed type IV hypersensitivity reaction to the mites, their eggs or scybala (packets of feces). Due to heavy infestation of the mites, multiple doses of both topical permethrin as well as the oral Ivermectin is recommended to ensure effective mite eradication.

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Figure 1. Crusted lesion at the wrist



Figure 2. H&E stain showing Sarcoptes scabei



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## Case Report: Aneurysmal Bone Cyst of the Spine

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#### **CASE REPORT:**

A 7 year old girl presented with 1 month history of aching pain in right leg and gradually worsening gait difficulty. There was vague back pain but no paresthesias or radiating pain. She had intermittent nocturia. There was no prior trauma or febrile illness. On examination, there was weakness of dorsiflexion at right ankle with diminished ankle jerk. She had back pain with flexion. The remainder of the examination was normal. MRI spine revealed a large, well-circumscribed lytic expansile mass in right sacrum and body of S1 with multiple fluid levels and hemorrhage extending to neural foramina of L5-S1/ S1-S2 and compressing the caudal sac (Fig 1,2). She underwent L5-S1 laminectomy and resection of the tumor with fusion from L4 to sacrum with right iliac screw. Histology was consistent with aneurysmal bone cyst. Postoperatively she has some residual pain and bladder incontinence improving with rehabilitation.

#### DISCUSSION:

Aneurysmal bone cyst is an uncommon bone tumor

comprised of a rapidly proliferating blood filled cystic lesion and account for 1-2% of benign bone tumors and 11-30% occur in the spine. They present with back pain, swelling and stiffness, often in patients younger than 20 years. Spinal involvement can lead to cord / nerve root compression. A high index of suspicion and evaluation with CT/MRI is essential. Treatment options include curettage and bone grafting, excision, embolization and/or radiation. Spinal lesions require stabilization. The prognosis is good but the high rate of recurrence warrants meticulous follow-up. <sup>2,3</sup>

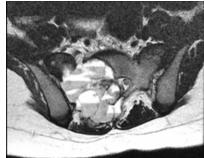
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Figure 1. MRI LS Spine – Sagittal



Figure 2. MRI LS Spine – Coronal



## Case Report: Angiosarcoma of the Gastrointestinal Tract: A Rare Case

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\*\*Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology
\*\*\*Drexel University College of Medicine: Department of Pathology and Laboratory Medicine

#### CASE REPORT

A 73-year-old woman with a medical history of blindness, hypertension and diabetes was diagnosed with new-onset atrial fibrillation requiring anticoagulation (dabigatran). During her admission, the patient had bright red hematochezia and a precipitous drop in her hemoglobin prompting transfusion. A colonoscopy was performed revealing a 1.5-cm ulcerated, heaped-up, submucosal nodule in the transverse colon which was biopsied. Microscopic examination revealed an infiltrating tumor composed of vascular channels and endothelial cells that were reactive to an endothelial marker (CD31) consistent with an angiosarcoma. Pre-operative imaging was negative for metastasis. She underwent a right hemi-colectomy with

removal of the entire tumor.

## DISCUSSION

Angiosarcomas are malignant tumors of the vascular endothelium representing about 1% of all sarcomas. Fourteen case reports of primary colonic angiosarcoma, which account for <0.001% of all colorectal cancers, have been cited in the literature. We report a case of primary colonic angiosarcoma identified at an early stage in the disease process in the setting of hematochezia from systemic anticoagulation. In the literature, older age (>50 years), larger tumor size (>5cm), and metastatic disease have been associated with a poor clinical outcome in patients with angiosarcomas. Lesions typically present with

gastrointestinal bleeding and anemia. Six of these patients survived less than 6 months often due to a delay in diagnosis resulting in poorer outcomes. We believe anticoagulation caused our patient's tumor to bleed allowing us to identify it at an earlier stage. At her 6 month follow-up visit, she was feeling well and had no complaints

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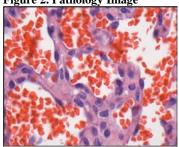
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Figure 1. Colonoscopy Image



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Figure 2. Pathology Image



## Case Report: Antithrombin III (ATIII) Concentrate for Treatment of Hepatic Sinusoidal Occlusion Syndrome in an Infant with Acute Lymphoblastic Leukemia (ALL)

Himadri Nath, MD, Stanley Calderwood, MD, Ionela Iacobas, MD Saint Peter's University Hospital: Department of Pediatrics

#### CASE REPORT

A 7-month-old male, diagnosed at three months of age with infantile ALL, developed sinusoidal occlusion syndrome characterized by jaundice, hepatomegaly, ascites and significant weight gain following high dose methotrexate. He developed a consumptive coagulopathy with low protein C, S and ATIII levels. His condition did not allow for a safe hepatic biopsy. As he was non-responsive to conservative management with fluid and sodium restriction, fresh frozen plasma and diuretics, he was administered antithrombin concentrates to maintain plasma antithrombin concentration > 30%. His condition improved significantly over the next several weeks and the hepatomegaly, ascites and coagulopathy resolved.

## DISCUSSION

Hepatic sinusoidal occlusion syndrome is characterized by jaundice, painful liver enlargement and retention of fluids

with weight gain and ascites. It is caused by chemotherapy induced injury to the vascular endothelium of the hepatic sinusoids, resulting in formation of local microthrombi, obstruction of portal blood flow, and hepatocelluar injury. Complications include portal hypertension, liver failure, consumptive coagulopathy, and multiorgan system failure. Mortality rate approaches 100% in severe cases. Sinusoidal occlusion syndrome is most often seen after high dose therapy for bone marrow transplantation, but has been reported after conventional chemotherapy, especially high dose methotrexate. Antithrombin concentrates may work by inhibiting new thrombus formation in hepatic sinusoids and re-establishing normal portal flow. Sinusoidal occlusion syndrome should remain in the differential diagnosis of children presenting with ascites and hepatomegaly. Timely initiation of directed therapy is of essence in this life-threatening condition.

## Case Report: Are Hyperacute T waves in chest pain an indication for an emergency PCI?

Swarnalatha Kanneganti, MD, Farhad Elmi, MD Easton Hospital: Department of Medicine

## CASE REPORT

A 34-year-old male arrived to the emergency room within 50 minutes after sudden onset of post-prandial mid-sternal chest discomfort. Serial electrocardiograms showed tall and peaked T waves (hyperacute) in leads V2-V4 without STsegment elevation. The patient was taken for cardiac catheterization as his chest pain persisted. A total occlusion of the left anterior descending artery was noted at the midsegment with evidence of right-to-left collateral circulation. Successful PCI was performed with door-to-balloon time of 99 minutes.

#### DISCUSSION

Traditionally, acute myocardial infarction requiring emergency percutaneous coronary intervention (PCI) is defined by ST-segment elevation or new onset left bundle branch block in an appropriate clinical setting. Hyperacute T waves have not been a sole indicator for emergency PCI in the literature. Prolonged hyperacute T waves should be considered an indication for emergency PCI in the appropriate clinical setting. This can improve the door-toballoon time with ultimate better clinical outcome.

## Case Report: Botulinum Toxin in Treating a Child with Hirschsprung's Disease

Mona Chhabra, MD, William Bernstein, MD Saint Peter's University Hospital: Department of Pediatrics

#### CASE REPORT

A 4 year old male patient with a history of Hirschsprung's disease, a leveling colostomy, and several anal dilatations presented with enterocolitis and gross fecal impaction. The patient received IV metronidazole and piperacillitazobactam. He was maintained with intermittent large red rubber catheter colonic irrigations and a nasogatrtic tube. On admission day #6, the patient was taken to the operating room for Botox A injections (20 units) into each quadrant of the internal anal sphincter. <sup>1</sup>

## DISCUSSION

Congenital aganglionosis of the distal bowel defines Hirschsprung's disease. Both myentric and submucosal plexus are absent resulting in reduced bowel peristalsis and function. On barium enema the classic finding is narrow distal colon with proximal dilatation. Anorectal manometry demonstrates absence of the inhibitory relaxation reflex of the internal sphincter after distension of the rectal lumen. Rectal biopsy is needed for the definitive diagnosis which reveals absence of ganglion cells. Several surgical procedures are described for Hirschsprung's disease such as the Swenson procedure, Duhamel procedure, modified procedures for long segment disease, anal myomectomy for ultra-short segment disease and the

latest being injection of botulinum toxin in the internal anal sphincter after the pull through procedure and for symptomatic internal sphincter hypertonicity. Botulinum toxin binds and inhibits acetylcholine esterase release at neuromuscular junctions and helps achieve temporary relaxation in pathological sphincters of children with Hirschsprung's disease.<sup>3</sup> However because the muscle paralysis by the toxin is reversible these children might relapse in the longer term and may need repeated injections.<sup>2</sup>

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## Case Report: Candida utilis: A Rare Pathogen

B. Sharmila Mohanraj, MD, Parul Kaushik, MPH, MD Drexel University College of Medicine: Department of Medicine, Division of Infectious Disease

#### CASE REPORT

A 54-year-old woman presented to the emergency department with one week of fevers and chills. She had had a recent diagnosis of a lung mass that was being treated as presumptive Actinomyces infection with intravenous ampicillin-sulbactam infusion through a Peripherally Inserted Central Catheter (PICC) in her right antecubital fossa for the past three weeks. She was afebrile on presentation. The PICC insertion site did not show erythema, tenderness or purulent discharge. Laboratory testing revealed a white blood cell count of 17,000 cells/mm3 with 80% neutrophils. Two sets of peripheral blood cultures were drawn and the patient was started on empiric antibiotics. The next day, blood cultures turned positive for yeast. Antibiotics were changed empirically to an echinocandin, and the PICC line was removed. Within two days, her white blood cell count decreased to 12,000 cells/mm3, and she reported subjective improvement. One week after the cultures were drawn, the yeast was identified as Candida utilis using VITEK 2. The echinocandin was adjusted to fluconazole based on the sensitivity pattern of Candida utilis. Subsequent blood cultures were negative; ophthalmology examination was unremarkable, and the

patient received two weeks of appropriate antifungal treatment.

#### DISCUSSION

Candida blood-stream infections are associated with immunocompromised states, indwelling central venous catheters, and pelvic surgery, with Candida albicans being the most common species isolated. Although there has been an upsurge of infections caused by non-albicans species, Candida utilis remains an exceedingly rare cause of fungemia, with only five cases reported in the literature.

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## Case Report: Constrictive Pericarditis in a 13 year-old after Mediastinal Radiotherapy

Aakanksha Gera, MD, Michele Cohen, DO, Tejal Mehta, MD Saint Peter's University Hospital: Department of Pediatrics

#### CASE REPORT

13 year-old female, known case of relapsed T-cell ALL, s/p 3 chemotherapy courses, 2 bone marrow transplants, total body irradiation in 2009, and palliative readiation of 2400 cgy for mediastinal mass and 3500 cgy for bone lesions. She presented with tachycardia and increased oxygen requirement up to 1.5L O2 followed by hypotension with BP 70s/40s. PE showed wight gain 4.8 kg in the past month, cushingoid appearance, shoddy LAD, tachypnea, decreased air entry b/l lung bases, normal S1, soft S2, distant heart sounds, hepatomegaly, and b/l LE pitting edema. Labs showed pancytopenia, BNP 250-370. CXR showed significant b/l pleural effusions. EKG showed sinus tachycardia, non-specific T-wave abnormalities and prolonged QT. Echo showed thickened pericardium, trivial pericardial effusion, impaired ventricular diastolic function, LVEF 67%, and large left pleural effusion. Findings were strongly suggestive of constrictive pericarditis, confirmatory cardiac MRI and cardiac catheterization were

not done as the patient was terminal and in hospice care.

#### DISCUSSION

Constrictive pericarditis is a well-recognized but sporadic complication of mediastinal radiotherapy in the pediatric age group. It is defined as scarred, fibrotic, and sometimes calcified pericardium due to multiple reasons including mediastinal radiotherapy, tuberculosis, and cardiac surgery. In our patient the history of mediastinal radiotherapy, physical findings, and echo with thickened pericardium, preserved EF but impaired diastolic filling, trivial pericardial and b/l pleural effusions, were highly suggestive of rare pediatric constrictive pericarditis.

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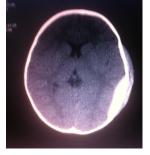
## Case Report: Delayed Onset Epidural Hematoma

Adam Isacoff, MD, Tejal Mehta, MD Saint Peter's University Hospital: Department of Pediatrics

## CASE PRESENTATION

A 2 year-old boy presented to the Emergency Department with 1 day of increased in left sided scalp swelling. Four days prior to presenting, the patient fell off of a six foot slide, landing on the left side of his body. No changes in mental status or vomiting were reported. Slight scalp swelling with no other bruising or injury was noted. The scalp swelling then drastically increased 3 days later. On physical exam, vitals were stable; a ~10 cm swelling was appreciated over the left parietal region of the skull with point tenderness noted. The rest of the exam including detailed neurologic examination was unremarkable. A head CT showed a hyperattenuating, lenticular, extra-axial collection measuring 12mm in thickness in the left parietal region, with a 5 mm midline shift to the right (Figure 1). He was immediately taken to the operating room for a frontotemporal decompressive craniotomy. The procedure was tolerated well and he was discharged home on postoperative day 4.

Figure 1. Left parietal epidural hematoma



#### DISCUSSION

Epidural hematoma is present in 0.5 -1 % of patients who have experienced traumatic brain injuries. All phases of presentation (decreased consciousness, a lucid interval, then a loss of consciousness) happen hours after the presenting injury. Epidural hematoma is caused by a direct impact injury between the inner table of the skull and dura. Blood forms from arterial or venous bleeding, most commonly a laceration of the middle meningeal artery. Focal evacuable hemorrhagic lesions occur and require immediate neurosurgical intervention.

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## Case Report: Elevated CA 19-9 in a female with multifocal biliary strictures

Maosong Qi, MD, Manoj Mittal, MD, Susan Sloan, MD Easton Hospital: Department of Medicine

#### CASE REPORT

A 57 year-old female presented with fever, chills, mild abdominal pain for 3 days. CT of abdomen showed mild central intrahepatic bile duct dilatation. Patient was admitted for treatment of acute cholangitis and further evaluation. MRCP showed multifocal areas of biliary stricture and associated dilatation (Fig. 1-A). Considering the multifocal biliary strictures, a neoplasm was highly suspected. CA 19-9 was 1755 U/mL (normal: < 37 U/mL). ERCP was then performed (Fig. 1-B). Brushings were done and cytology came out negative for malignant cells. ERCP was repeated and again cytology did not find any malignancy. CA 19-9 was repeated 5 days later and was down to 29 U/mL. Patient symptoms had improved and she was discharged to home. CEA, CA 19-9 will be checked in 2 months, and patient will be followed by gastroenterologist.

#### DISCUSSION

Elevated CA 19-9 has also been associated with benign diseases including acute cholangitis. Even though CA 19-9 can return to normal level with the subsidence of

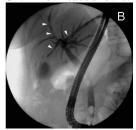
cholangitis, some data suggest cholangitis-associated elevated CA 19-9 may have increased risk for cholangiocarcinoma, and these patients require close follow up, for example MRCP in 3 to 6 months. Our patient has multifocal stricture cholangitis, and the significance needs to be addressed.

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Figure 1. A-MRCP, B-ERCP. Arrowheads show strictures





## Case Report: Epithelioid Hemangioendothelioma (EHE) in a patient with CREST syndrome

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## CASE REPORT

Patient is a 59 year-old Caucasian female with a past medical history of CREST syndrome and pulmonary hypertension who presented for further workup of pulmonary nodules that were found on a CT scan of the chest. During her follow up, an outpatient PET scan showed the largest of the lesions to be hypermetabolic. Subsequently, the patient underwent VATS which histologically showed EHE. The patient was given a brief trial of Sunitinib by oncology, but the medication was discontinued due to side effects of diarrhea.

#### DISCUSSION

EHE is a tumor that rarely presents of pulmonary origin. Calcinosis cutis, Raynaud phenomenon, esophageal motility disorder (CREST Syndrome) has a modest increase in risk of lung cancer. Our patient presents with concomitant EHE and CREST syndrome. EHE is rare

cancer that was first described in 1975 and can present with a variable clinical course: it can be asymptomatic, indolent with chronic cough, regress spontaneously, or be rapidly fatal with similar CT findings. EHE is a vascular tumor and is considered a sarcoma that does not have a well-described natural history. Chemotherapy and radiotherapy are generally considered ineffective and curative surgery is often not feasible since EHE often presents as multifocal bilateral lesions. In the pulmonary form, half of the cases die due to respiratory failure. Case reports have reported regression to chemotherapy regimens that include medications that block vascular endothelial growth factor (VEGFR), or a VEGFR Tyrosine kinase inhibitor.

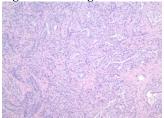
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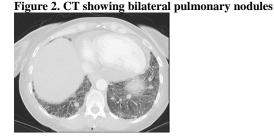
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## Case Report: Esophageal Stricture Post Alkali Ingestion

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#### CASE REPORT

A 2 year old male patient presented to the ED with persistent vomiting immediately after eating solid foods. He was reportedly tolerating Pedialyte and water. The patient had been seen in the ED one month prior to this visit due to ingestion of oven cleaner. He was evaluated at that time and discharged home. The current physical exam was normal. Due to history, a barium swallow was ordered which showed an esophageal stricture in the distal esophagus (Figure 1).

#### DISCUSSION

Caustic ingestion injury to the digestive tract remains a significant problem despite various efforts to minimize hazards of caustic household products. Alkali substances, such as drain cleaner, are commonly accidentally ingested as they are tasteless. They can produce liquefaction necrosis and stricture formation. Symptoms include vomiting, drooling, refusal to drink, dysphagia, dyspnea, abdominal pain, hematemesis and stridor. Presence of 3 symptoms is predictive of severe disease. Approximately

Figure 1. Esophageal Stricture



20% develop strictures. The classification of esophageal injury classified based on endoscopy findings post 12-48 hrs ingestion are grade 0, negative finding; grade 1, injury limited to erythema and edema; grade 2, ulcerations with necrotic tissue and white plaque; and grade 3, injury involving deep ulcerations, white plaque, and necrotic changes. Grade 0 and 1 esophageal injuries are defined as low-grade injuries, while grade 2 and 3 esophageal injuries are defined as high-grade ones. Dilution by water or milk is recommended but neutralization, gastric lavage and induced emesis should be avoided. Strictures are treated with dilation, surgical resection or stenting.

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Case Report: Fatty metamorphosis or calcification of the infarcted myocardium on Cardiac MRI

Swarnalatha Kanneganti, MD, Farhad Elmi, MD, Mahesh Krishnamurthy, MD Easton Hospital: Department of Medicine

#### CASE REPORT

A 52-year-old female had an extensive anterior wall

myocardial infarction with late presentation. She required percutaneous coronary intervention to the left anterior

descending artery with 2 drug-eluting stents. There was suspicion of a left ventricular apical thrombus for which the patient was placed on oral anticoagulation with warfarin. Two months later, the patient presented with chest pain. EKG did not show any acute changes and serial cardiac enzymes were negative. There was no reversible ischemia on adenosine-sestamibi SPECT stress test. However, the transthoracic 2D Echocardiogram showed a linear calcified structure along the distal half of the septum butting into the left ventricular cavity. This was thought to be a thrombus along the distal septum, however an aneurysm could not be excluded. This finding was new when compared to the prior echocardiogram performed at the time of acute myocardial infarction. Cardiac MRI was obtained,

Fig 1. Echo showing LV linear calcified structure



revealing calcification or fatty metamorphosis of the distal septum and a left ventricular apical true aneurysm. No intra-cavitary lesion was noted.

#### CONCLUSION

We present a case of an extensive anterior wall myocardial infarction with late presentation, complicated by fatty metamorphosis and left ventricular apical aneurysm of the infarct territory. Calcification of infarcted myocardium soon after myocardial infarction is a rare finding. Consideration of this entity, while providing follow up care to the patient with acute MI, can be of clinical value. Further studies are required to assess the clinical and prognostic significance of fatty metaplasia.

Fig 2. MRI with septal calcification or fatty metamorphosis



Case Report: Fibromuscular dysplasia presenting as a renal artery infarction in a 35 year-old female with no predisposing factor

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## CASE REPORT

A 35 year-old female with a past medical history significant for polycystic ovarian syndrome not requiring medical therapy presented to the ED complaining of 3 days progressive, intermittent, left flank pain radiating into the lower back. She never smoked or drank alcohol. Family history was unremarkable. Other than being sinus tachycardic at 120/min the rest of the vital signs were within normal range. Patient physical exam was significant for left flank tenderness without rebound. Laboratory data were all unremarkable including normal renal function and a negative pregnancy test. CT scan of the abdomen revealed a segmental left renal infarct. Hypercoagulability panels were obtained and intravenous weight-based heparin was initiated. A renal arteriogram confirmed the stenosis of a sub-segmental artery to the left kidney corresponding to the infarct area. All hypercoagulability tests came back normal, as did a rheumatologic work up for vasculitis and the hepatitis panel. Trans-esophageal echocardiography did not identify any cardiac source of emboli. Patient was discharged home on Coumadin without further incidence.

## DISCUSSION

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory arterial disease that most commonly affects the renal and internal carotid arteries. <sup>1</sup> Although it is a known cause of renovascular hypertension in younger patients, a presentation of renal artery infarction is rare. <sup>2</sup> The rare reports of FMD induced renal artery infarcts were commonly associated with hypercoagulability states or an embolic source. <sup>3</sup> To the best of our knowledge this is the first case reported with no predisposing factors.

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## Case Report: Heerfordt's Syndrome: Rare variant of Sarcoidosis

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\*Abington Memorial Hospital: Department of Medicine, Division of Pulmonology
\*\*Abington Health Lansdale Hospital: Department of Medicine

#### CASE REPORT

 $55\ \text{yr}$  old AAF presented with c/o fevers with chills, dry

mouth, difficulty swallowing, dry eyes, rash on legs and swelling of both sides of face along with weakness of face

progressively getting worse over 1 month period. ROS: 10lb wt loss. PMH: None. Social and family history was not contributory and she was not on any outpatient medications. PE: Sick appearing, Temp 101F. HEENT: Injected sclera, PERLA, bilateral firm non-tender parotid enlargement. Heart and lung exam normal. Extremities: Erythema nodosum on bilateral lower extremities. Neurological exam: bilateral facial nerve weakness (LMN type). Labs: CMP, CBC, ESR, CRP were normal; Calcium 10.4; AST 80, ALT 68, AKP 128. CXR normal. ANA, SSA, SSB, RF, HIV were negative. Initially the DDx included SICCA syndrome secondary to Sjogren's, viral infections, and atypical presentation of Sarcoidosis. Considering that the rheumatology work-up was negative up to this stage, further tests were ordered to investigate the atypical presentation of Sarcoidosis. CT chest: Mediastinal and hilar lymphadenopathy. ACE level elevated at 222. MRI brain normal. Hilar lymph node biopsy revealed noncaseating granulomas.

#### DISCUSSION

A diagnosis of Heerfordt's syndrome (uveoparotid fever) was made as the patient had non-suppurative parotitis, uveitis, fever and cranial nerve paralysis. This is usually a self-limiting disorder, but she was started on IV methylprednisolone 40 mg every 6 hours as there was involvement of the facial nerves. Her symptoms rapidly improved in 2 days and she was sent home on a prolonged slow taper of steroids.

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## Case Report: Herpes Esophagitis in an Immunocompetent Patient Causing Anemia and Exertional Angina

Shahruq Sarela, MD

Allegheny General Hospital: Department of Internal Medicine

#### CASE REPORT

A 96-year-old Caucasian female with PMH of colonic AVMs, partial colectomy with colostomy, CKD, s/p CABG presented with new onset reddish-brown stool from her colostomy bag for 2 days. She reported exertional chest discomfort with walking. Prior to this event she had no orthopnea, paroxysmal nocturnal dyspnea, or lower extremity edema. In the ER she was guaiac positive with a hemoglobin of 9.0 from a baseline of 11.0 two months prior. She had a normal point of care troponin, INR 1.1, Bun 78, Cr 1.6, WBC 6.9, and PLT 203. EKG showed sinus bradycardia, LVH, and ischemia. Her home medications were ASA, Aldactone, HCTZ, Lisinopril, and Plavix for her cardiomyopathy. EGD showed a clean base ulcer at the gastro-esophageal junction. Ostomy scope revealed few non-bleeding angioectasias in the cecum. Ulcer biopsy was positive for herpes esophagitis. She reported no odynophagia or dysphagia complaints. She was treated with Valtrex and transfused 2 units PRBCs.

#### DISCUSSION

It was proposed that the patient had an upper GI bleed secondary to herpes esophagitis. Usually this is self-

limiting in immunocompetent patients and resolves gradually within 6 - 42 days (mean, 17.5 days) after presentation. Severe complications include bleeding and esophageal perforation; therefore treatment is recommended. Given the patient had CKD, it was ideal to use Valtrex over Acyclovir. Both drugs are effective forms of treatment. The anginal pain was thought to be from outstripping the collateral CABG network which resolved after transfusion of 2 units of PRBCs.

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Case Report: Hydroxyurea Treatment for Stuttering Priapism in a Prepubertal Male with Sickle Cell Disease

Anitha Sathynarayana Singh, MD, Stanley Calderwood, MD, Ionella Iacobas, MD, Kanika Shanker, MD Saint Peter's University Hospital: Department of Pediatrics

## CASE REPORT

A 4 year old African-American male with Sickle Cell Disease (SCD) developed priapism during hospitalization for streptococcus pneumonia sepsis. The acute episode was treated with analgesics, hydration and packed red blood cell transfusion. Following this intial episode, he developed stuttering priapism persisting over the next 2 years requiring frequent emergency department visits. On 2

occasions he required aspiration of blood from the corpus cavernosa. In view of the recurrent nature of the disease, he was started on oral hydroxyurea with a target a fetal hemoglobin (Hb F) level >30%, after which stuttering pripapism resolved, and there have been no further episodes. Notably, there have also been no further admissions for acute vaso-occlusive crisis.

#### DISCUSSION

Priapism is an unwanted often painful and sustained erection lasting longer than 1 hour. It may cause impotence due to fibrosis of the corpus cavernosa. Stuttering priapism (repeated episodes) has been reported in prepubertal males with SCD. Treatment of acute episodes includes hydration and analgesics, and occasionally aspiration of blood from the corpus cavernosa in non-responsive patients. Due to the risk of impotence, prevention of episodes is highly desirable. Stuttering priapism is an under-investigated and poorly-managed urologic problem in prepubertal children with SCD. The efficacy of hydroxyurea is promising and warrants further multicenter studies.

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## Case Report: Iatrogenic duodenal perforation treated with endoscopic placement of metallic clips

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\*\*Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology

### CASE REPORT

A 50-year-old woman who underwent pancreaticojejunostomy nine years prior for chronic pancreatitis was hospitalized with acute relapsing pancreatitis (ARP). Magnetic resonance cholangiopancreatography revealed a dilated pancreatic duct and subsequently an ERCP was performed. After passage of the side viewing endoscope to the ampulla, the papilla was examined at which time bleeding was seen coming from the duodenal wall opposite the papilla. Examination of this area revealed a 1 cm defect compatible with perforation likely caused by the shaft of the endoscope in the presence of a fixed duodenum from her previous surgery. Fluoroscopy showed free air in the peritoneal cavity and peritoneal leakage of contrast injected into the duodenum confirmed a leak (Figure 1). The side viewing endoscope was then exchanged for a gastroscope and the perforation was closed with 8 hemostatic endoclips (Boston Scientific, Resolution) (Figure 2). Subsequent contrast injection into duodenum showed no leak. The patient consented to conservative treatment of the perforation and was started on broad-spectrum antibiotics and nasogastric tube was placed. A follow-up upper GI series was normal and CT of the abdomen 2 weeks later showed persistent free air in the peritoneum. Initiation of

feeding precipitated an attack of ARP which was managed with total parenteral nutrition and later placement of jejunal feeding tube. She was discharged pain free and stable on tube feeding after 4 weeks. The patient was subsequently seen in the outpatient clinic without noted complications related to the perforation.

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Figure 1. Fluoroscopy contrast leak



Figure 2. Multiple endoclips



Case Report: Infected Urachal Cyst resulting in an Umbilical-Urachal Sinus Tract in an Adult

Ami Khatri, MD

Drexel University College of Medicine: Department of Family, Community, & Preventive Medicine

## CASE REPORT

A 25 yo male presents with drainage from his umbilicus.

Patient was afebrile and physical exam revealed purulent drainage coming from the umbilicus. CT abdomen/pelvis

showed a 7.7x6.8x4.8 midline collection consistent with an infected urachal cyst resulting in an umbilical-urachal sinus. Imaging could not distinguish an infected urachal cyst and urachal cyst adenocarcinoma and biopsy was recommended. Urology was consulted and cystoscopy revealed a small defect at the bladder dome. Bladder biopsy was not performed because the patient was lost to follow-up.

#### DISCUSSION

Urachal abnormalities are common in infants. Described here is a case of an infected urachal cyst resulting in an umbilical-urachal sinus tract. Urachal abnormalities remain rare in adults, the most common presentation being an infection of the urachal cyst. Histolologically the urachus is comprised of three layers, the outermost layer being continuous with the detrusor muscle of the bladder. During fetal development, the bladder descends into the pelvis pulling the urachus along resulting in formation of the urachal canal. Throughout development, the canal obliterates and eventually forms a fibrous tract. At the end

Figure 1. CT showing sinus tract ascending to umbilicus

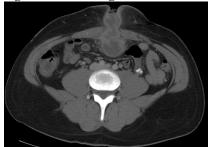


of development, the urachus attaches the umbilicus to the dome of the bladder. A defect resulting in partial obliteration of the urachal canal can lead to abnormalities such as a patent urachus, urachal cyst, and a urachal sinus which drains into the umbilicus. Computed tomography and ultrasound are used to identify urachal abnormalities. The treatment is complete primary surgical excision due to the risk of malignant transformation.

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Figure 2. CT showing sinus tract from umbilicus to bladder



Case Report: Inferior Vena Cava (IVC) Leiomyosarcoma: A Rare Entity

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## CASE REP

A 64-year-old male with a history of hepatitis C presented with chronic intermittent right flank/abdominal pain for several months. Upper and lower GI endoscopies were unremarkable. An abdominal CT scan was negative except nonspecific periportal/paracaval adenopathy. The patient had cholecystectomy for a hypocontractile gallbladder but symptoms persisted. A subsequent CT scan showed a 3.8 x 3.0 cm ovoid mass within the retroperitoneum, inseparable from the IVC and right adrenal gland. MRI of the abdomen confirmed these findings. The patient was transferred to a tertiary-care center for surgical resection which involved right nephrectomy and adrenalectomy along with a polytetrafluoroethylene graft replacing IVC. The pathology report confirmed the grade-3 IVC leiomyosarcoma. The patient died from perioperative complications.

#### DISCUSSION

IVC leiomyosarcoma is a rare malignant tumor, constituting 0.5% of adult soft tissue sarcomas.<sup>1,2</sup> The incidence of this tumor is not well described in the literature due to its rarity (approximately 300 cases reported). The initial diagnosis is often delayed in daily clinical practice due to nonspecific clinical features.<sup>3</sup>

Complete en-bloc resection provides the best chance of cure but the prognosis remains poor. The optimal treatment strategy is still controversial in case of recurrence. Post-resection management of IVC is contentious as different strategies such as primary repair, ligation, patch repair, or reconstruction have been used with variable results. <sup>4,5</sup> Although adjuvant therapy is beneficial, clear evidence is lacking and more studies are needed to establish its significant role.

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Figure 1. CT showing IVC leiomyosarcoma



of the Inferior Vena Cava. Journal of Gastrointestinal Surgery 2008;12:2141-8.

Figure 2. MRI showing IVC leiomyosarcoma



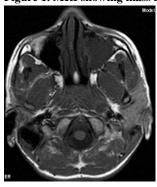
Case Report: Large Latero-Cervical Mass in a Teenager Proves NOT to be Lymphoma

Tayleen Bhatia,MD, Megha Sharma,MD, Stanley Calderwood,MD, Ionela Iacobas,MD Saint Peter's University Hospital; Department of Pediatrics

#### CASE REPORT

A thirteen-year-old male presented with a latero-cervical mass and no history of fevers, weight loss or night sweats. Initially, CT neck showed lymphadenopathy and left submandibular sialadenitis. The mass continued to grow and re-evaluation three weeks later revealed a left posterior cervical mass 7x5cm, firm, multi-lobulated, non-tender, and an enlarged submental lymph node 4.5x3cm. Repeat CT neck showed multiple necrotic cervical lymph nodes and mentioned opacification of maxillary sinus most probably due to an occupying lesion. MRI brain/neck revealed a heterogeneously enhancing mass involving left ethmoid air cells and left upper nasal cavity/osteomeatal complex extending into left maxillary sinus and into medial aspect of the orbit and confluent jugular chain lymphadenopathy. Lymph node biopsy was diagnostic for rhabdomyosarcoma. Metastatic work-up was completed. Patient was classified as parameningeal alveolar rhabdomyosarcoma, stage III, grade 3a and started on chemo and radiation therapy and is doing well.

Figure 1. MRI showing mass lesion in left maxillary sinus



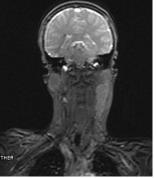
#### DISCUSSION

A teenager presenting with large, fast-growing lymphadenopathy is highly suspicious for lymphoma. We report the case of a rhabdomyosarcoma with typical clinical picture of a hematological malignancy at onset. Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma. Still, our patient had no epistaxis, nasal congestion or vision complaints that may point towards a paranasal/orbital concern. As in our case, sometimes the metastatic site dominates the clinical picture and the primary tumor may be found only incidentally with significant long-term influence on prognosis and management.

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Figure 2. MRI showing left jugular adenopathy



Case Report: Lyme Disease Presenting with Facial Palsy and Myocarditis Mimicking Myocardial Infarction

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## CASE REPORT

A 45-year-old female presented with sudden onset of

severe chest discomfort and associated diaphoresis and headache. She was afebrile with a blood pressure of

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154/108 and heart rate of 86. The patient received sublingual nitroglycerin with subsequent relief of the pain. Initial EKG showed normal sinus rhythm with 1 mm ST-elevations in lead II and lead aVF and 1 mm ST-depression in lead V1 with associated T-wave inversion. Initial Troponin I and CK-MB were elevated at 7.82 and 55.2 respectively and, 6 hours later, Troponin I increased to 13.44 and CK-MB to 75.7. Cardiac catheterization revealed normal coronary arteries and a normal ejection fraction. During hospitalization, she developed right-sided facial palsy. Her Lyme titers were positive. After treatment with ceftriaxone, her symptoms resolved.

#### DISCUSSION

Lyme disease may involve the skin, nervous system, heart, and joints. <sup>1</sup> Cardiac manifestations are relatively infrequent and, typically, present with a fluctuating degree of atrioventricular block and, rarely, myocarditis, with or without, pericardial involvement. <sup>2</sup> The most common neurologic manifestations are cranial neuropathy, particularly facial palsy, lymphocytic meningitis and motor or sensory radiculoneuritis. <sup>3</sup> This is an unusual case of Lyme myocarditis associated with markedly elevated

Figure 1. EKG at presentation



Troponin I and normal left ventricle function. Such high level of Troponin I related to Lyme myocarditis has not been previously reported in the literature. Lyme myocarditis should be considered in the differential diagnosis in patients presenting with clinical symptoms suggestive of acute coronary syndrome.

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Figure 2. EKG after treatment



Case Report: Negative Pressure Pulmonary Edema after Robotic-Assisted Radical Prostatectomy

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## CASE REPORT

A 48-year-old man was admitted for a robotic-assisted radical prostatectomy for prostate cancer. His past medical history included hypertension, dyslipidemia gastroesophageal reflux. The patient underwent a cholecystectomy in the past without significant postoperative events. He denied smoking, alcohol intake or illicit drug abuse. His routine pre-operative assessment was revealed as low risk for the planned surgical procedure. The patient received Desflurane and Propofol for induction of anesthesia, Midazolam for sedation, and Fentanyl and Phenylephrine as adjuvants. The operation was completed successfully. Immediately after extubation, however, the patient developed stridor, hypoxemia, and severe respiratory distress; he was re-intubated. Chest examination revealed new-onset bilateral crackles. Chest radiograph showed pulmonary vascular congestion. The patient was continued on pressure support ventilation and successfully extubated after 12 hours.

## DISCUSSION

Negative pressure pulmonary edema (NPPE) is the term describing that unique type of pulmonary edema resulting from forceful breathing with obstructed airways resulting in increased intrathoracic negative pressure followed by increased venous return. Several cases have been reported describing different etiologies for the upper airway obstruction. Rapid recognition of NPPE is very critical, as misdiagnosis carries a 40% morbidity and mortality. NPPE should be suspected in any patient with respiratory distress associated with tracheal stridor, hypoxemia, and hypercapnea with radiographic evidence of pulmonary edema. The initial treatment is relief of the airway obstruction via intubation or tracheostomy. The uses of dexamethasone and diuretics have not shown to be efficacious in prospective studies, and may be deleterious.

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## Case Report: Painless Acute Aortic Dissection (AAD): A Question Without an Answer

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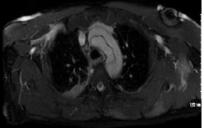
#### CASE REPORT

A left-handed previously healthy 63-year-old male presented with left side weakness and aphasia of unknown duration. His BP was 150/100 mmHg, HR was 63 bpm, regular and pulses were equal on both sides. No carotid bruit. CT head without contrast, Chest Xray, and EKG all were normal. Intravenous TPA therapy was withheld only because of the unknown symptom duration. He was started on aspirin. Ultrasound of the neck subsequently showed bilateral common carotid artery dissections, and then emergency MRA of the chest showed type-A aortic dissection (Fig 1). The patient was sent for emergency thoracic surgery. He subsequently improved and was ultimately discharged to a neuro-rehabilitation unit.

#### DISCUSSION

Acute ischemic stroke occurs in one-half of patients with AAD presenting with neurologic symptoms. Sudden-onset chest pain reported in two-thirds of patients. Classic physical findings such as aortic regurgitation and pulse deficit were noted in 31.6% and 15.1% of patients,

Figure 1. MRA showing aortic dissection



respectively. Initial chest radiograph and electrocardiogram were frequently not helpful. <sup>1,2</sup> Missing painless AAD presenting with neurologic symptoms will not only delay the proper surgical intervention but is potentially catastrophic in the thrombolytic era. <sup>3</sup> In the sight of the previous data , high index of suspicion will decrease but will not eliminate the incidence of missing painless AAD. In our case, in spite of proper history taking, complete physical examination and initial imaging studies, the presence of bilateral common carotid dissections was the first and only indication of ADD.

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## Case Report: Palmoplantar Pustulosis

Smitha Ballyamanda, MD, Rosemary Harris, MD Drexel University College of Medicine: Department of Family, Community and Preventive Medicine

#### CASE REPORT

An 18 year-old female presented to the office with erythema, itching, scaling and vesicular lesions of bilateral palms and soles of feet which gradually worsened over the past 3 months. She smokes ½ pack of cigarettes per day. The physical exam revealed bilateral erythematous palms with thickened silver scales covered in 1-2 mm vesicular lesions draining a yellowish fluid. The patient was initially treated with oral antibiotics for concerns of an infection along with low dose topical steroids. Patient reported continued discomfort and pain. She had a skin biopsy of her palm which demonstrated palmoplantar pustulosis. She was treated with Clobetasol proprionate cream with an occlusive plastic wrap over the palms and soles of feet. She was also counseled on the importance of smoking

cessation.

#### DISCUSSION

Psoriasis is a skin condition affecting the life cycle of skin cells, causing cells to build up rapidly on the surface of the skin forming well demarcated round or oval lesions. Palmoplantar pustulosis affects the palms of the hands and soles of the feet with small, deep, yellow pustular lesions. Biopsy is characterized by neutrophilfilled intra-epidermal vesicles. Palmoplantar pustulosis can present as crops of sterile pustules and may persist for many years, more common in current smokers or eversmoker. It is possible that activated nicotine receptors in sweat glands cause an inflammatory process. Treatment options include topical steroids, Acitretin tablets, Psoralen

and ultraviolet A (PUVA), a process of photochemotherapy and immunomodulators. The rate of reoccurrence is high.

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## Case Report: Posterior Reversible Encephalopathy Syndrome (PRES): a catastrophic but reversible side-effect of bevacisumab

Ramya Mallareddy, MD, Prasad Ancha, MD Easton Hospital: Department of Medicine

#### CASE REPORT

A 68-year-old female with history of non-small cell carcinoma of lung with metastasis to bone presented with seizures, left sided neurological deficits and confusion, one day after her chemotherapy. Patient had been receiving docetaxel and bevacisumab for 2.5 years. Patient had no history of hypertension but admission blood pressure was 160/90 mmHg. MRI of the brain showed prominent vasogenic edema involving parietal and occipital lobes symmetrically suggestive of PRES (Fig 1). There were no such findings in MRI brain done 7 months prior (Fig 2). EEG showed diffuse delta wave activity over both hemispheres. Patient was treated with antihypertensive medications and supportive measures. Chemotherapy was held. Patient's neurological deficits improved significantly by the third day and had no further deterioration.

## DISCUSSION

In this case, bevacisumab was suspected to be the cause of PRES<sup>1</sup> and other potential etiologies have been ruled out, mainly uremia, sepsis and hypertension. Bevacisumab is an anti-vascular endothelial growth factor agent. PRES is thought to be caused by vasospasm due to damage of the

vascular endothelium and failure of cerebral autoregulation causing vasogenic edema leading to typical MRI findings of hyperintensity in bilateral posterior cerebral white matter symmetrically, as seen in this patient.<sup>2</sup> Although prognosis is good in such patients, treatment is mainly supportive with tight blood pressure control and withdrawal of the offending agent.<sup>3</sup> Further studies are essential to evaluate the possibility of recurrence of PRES in such patients after resuming chemotherapy. Elevated index of suspicion is necessary to diagnose PRES and for institution of timely management.

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Figure 1. MRI brain at presentation

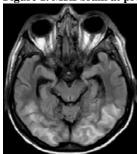
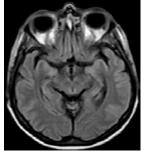


Figure 2. MRI brain 7 months prior



Case Report: Postpericardiotomy syndrome in an infant

Saurabh Patel, MD Saint Peter's University Hospital: Department of Pediatrics

## CASE REPORT

A 4 month-old male status-post correction of absent right

pulmonary artery 20 days prior, presented with history of cough, vomiting for one day, decreased oral intake

DrexelMed Journal, Volume 6 (1); Spring 2012

and urine output for 2-3 days. Patient had normal ECHO 10 days prior. Father is known case of CHF and concentric LVH. On examination child appeared withdrawn and dehydrated. Patient had tachycardia, normal heart sounds, no murmur, capillary refill was 3 sec. Patient was tachypneic, no wheezing, minimal grunting. During hospital course in matter of 6 hours, cardiorespiratory status worsened. Chest X-ray showed massive cardiomegaly (Fig 1). Echo showed massive pericardial effusion (Fig 2). US guided drainage was done and child was stabilized.

#### DISCUSSION

Postpericardiotomy syndrome is an inflammatory reaction of pericardium that develops after surgery involving pericardiotomy, with incidence 25% to 30%.<sup>3</sup> Onset is few weeks to few months after cardiac surgery. It is rare in age less than 2 years. Presentation includes fever and chest pain, on physical examination, pericardial and pleural

Figure 1. Chest X-ray showing cardiomegaly



friction rubs and hepatomegaly. Tachycardia, tachypnea, rising venous pressure is present, and falling arterial pressure with a paradoxical pulse are signs of cardiac tamponade. Chest x-ray shows cardiomegaly. EKG shows persistent ST–segment elevation and flat or inverted T waves. Echo is most reliable test. Management includes bed rest, hydration, NSAID in moderate case, in severe case moderate doses of corticosteroids may be indicated. Emergency pericardiocentesis may be required if cardiac tamponade is present. Recurrences can occur up to 21%. In recurrence pericardiectomy may be necessary.

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Figure 2. US showing pericardial effusion



## Case Report: Pott's Puffy Tumor (PPT): a forgotten complication of chronic sinusitis

Sivagama Ramasundaram, MD, Osamuyimen Igbinosa, MD, David Alcid, MD Saint Peter's University Hospital: Department of Medicine

## CASE REPORT

51-year-old previous healthy man presents with 4-month history of right-sided superomedial orbital swelling that slowly enlarged. He had 2-day history of progressive right-sided medial swelling that dramatically increased over what was previously present. He denied having any significant pain, but reported somewhat worsening diplopia over 24 hours that prompted emergency room visit. He was afebrile; had an obvious right medial orbital bulge measuring 3 cm with outward displacement of the right globe (Figure 1). Orbital swelling was firm, fluctuant but nontender; laboratory values were normal. CT scan is shown on figure 2. There was an immediate release of greenish mucopurulent secretion on aspiration under local anesthesia. Culture grew Fusobacterium nucleatum. Clindamycin was started with intention to treat for 6 weeks.

#### DISCUSSION

PPT is characterized by swelling of the scalp secondary to

subperiosteal abscess in frontal bone. PPT was originally described by the British surgeon, Sir Percival Pott (1714-1788). Incidence of PPT has progressively declined to the point of rarity since the advent of antibiotics<sup>2</sup>, but not to extinction as we have demonstrated. It has been associated with subdural empyema and intracerebral abscess. This entity is usually not recognized initially and easily confused with neoplasms or local eye pathology.

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Figure 1. Lateral view of right medial orbital bulge



Figure 2. CT scan showing mass eroding through sinus wall



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## Case Report: Proximal protection device with flow reversal system for safer carotid artery stenting (CAS)

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#### CASE REPORT

69 year-old male patient with history of HTN, DM was noted to have carotid bruit on examination. Doppler study revealed elevated velocity in left internal carotid artery 530/205 cm/sec. Carotid arteriogram revealed 95% stenosis of proximal left internal carotid artery. Due to high risk features for carotid endarterectomy, patient underwent CAS. Neurological examination was documented within 24 hours before and after CAS. During the procedure, occlusive balloons were inflated in external and common carotid arteries with resulting flow reversal. Procedure was performed with balloon pre-dilatation of the lesion followed by stent deployment under fluoroscopy. Finally, aspiration was done to remove any debris prior to deflating the occlusive balloons. Excellent results with 0% residual stenosis and brisk cerebral perfusion were noted.

## DISCUSSION

Proximal protection device with flow reversal system is a novel approach to minimize stroke risk during CAS. To prevent periprocedural strokes during CAS, distal protection with filters are used. Proximal balloon occlusion is an alternative which may be more effective. <sup>1,2</sup> Potential

disadvantages of filters are needing to cross the lesion before protection is installed which may allow small particles to pass through or alongside the filter. Potential advantages of proximal balloon occlusion are placement before crossing the lesion, inducing flow reversal by occluding external and common carotid artery thereby directing emboli away from the brain,<sup>3</sup> and debris is aspirated before deflation and restoration of flow to the brain.

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Figure 1. Flow reversal system

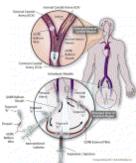


Figure 2. Proximal protection balloons in ECA and CCA



Case Report: Recurrent Gastrinoma and Diffuse Cutaneous Metastasis, a unique case

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- \*Drexel University College of Medicine: Department of Medicine, Division of Internal Medicine
- \*\*Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology

#### CASE REPORT

A 65 year old male with history of pancreatic gastrinoma resected in 1983 presented with abdominal pain 22 years later. An upper endoscopy revealed a 1 cm Barrett's but otherwise unremarkable. Four months later a repeat EGD revealed grade IV erosive esophagitis and multiple ulcers in the duodenum. Nine days later the patient developed worsening dysphagia despite PPI therapy. A subsequent endoscopy revealed a pinpoint esophageal stricture. A barium esophagogram showed a 9 cm long stricture and esophageal dilation was performed. The patient underwent weekly therapeutic dilation and required 3 removable esophageal stents. Ultimately the patient developed numerous cutaneous metastases. Six months later the patient succumbed to his metastatic disease.

Figure 1. Cutaneous metastatic gastrinoma



#### DISCUSSION

Gastrinoma is a rare tumor with an incidence of 1-4/million. Approximately 80% arise within the "gastrinoma triangle" bounded by the porta hepatis, the neck of the pancreas, and the third portion of the duodenum. Over 2/3 are malignant and commonly metastasize to the lymph nodes and liver while metastasis to bone and skin is rare. Gastrinoma patients sometimes develop ZES and present with multiple GI symptoms; however, Barrett's esophagus and esophageal strictures are uncommon. Although gastrinoma patients present with plethora of gastrointestinal symptoms, esophageal complications are rare. This is a unique case of known gastrinoma patient developing rapid progression of Barrett's esophagus to refractory esophageal stricture. This case also presents a rare example of cutaneous metastatic gastrinoma.

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## Case Report: Recurrent Syncope - Think Outside the Box

Thein Aung, MD, Jacek Jodelka, MD, Mahesh Krishnamurthy, MD Easton Hospital: Department of Medicine

## CASE REPORT

A 74 year-old male presented with multiple episodes of lightheadedness and syncope. His past medical history was significant for smoking and nasopharyngeal cancer treated with radiation and chemotherapy. Examination revealed orthostatic changes in blood pressure (BP- 168/100mmHg lying down and 102/44mmHg standing up). The rest of his physical exam and extensive laboratory, cardiac and neurological testing were negative. A diagnosis of baroreceptor failure was made on the basis of his history of radiation, tilt table test, depressor response to a small dose of clonidine and acceleration of pulse rate with mental arithmetic. The patient was advised a variety of nonpharmacologic measures (increasing dietary salt and caffeine, TED stockings) and advised physical maneuvers to decrease fluctuations in blood pressure. He was treated with alprazolam, clonidine and fludrocortisones and was doing better on follow-up.

## DISCUSSION

Baroreceptor failure is caused by improper regulation of autonomic neurons in the carotid arteries. It is an under recognized cause of labile blood pressures with orthostatic changes and recurrent syncope. It presents with postural light headedness, labile hypertension, orthostatic hypotension and syncope. Prompt diagnosis is extremely important as the treatment strategies differ from the conventional management of other conditions presenting with orthostatic hypotension associated syncope. Both the hypertension and hypotension are often difficult to control with conventional therapy. Clonidine is the antihypertensive of choice for supine hypertension. Fludrocortisone and benzodiazepines are helpful in some cases. Physical maneuvers like periodic squatting help maintain blood pressure when upright.

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Figure 1. Causes of Recurrent Syncope

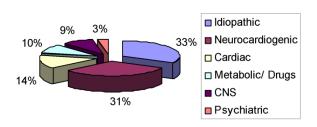
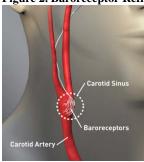


Figure 2. Baroreceptor Reflex



Case Report: Stroke in Fabry's Disease: A Disease Without a Clear Prophylaxis

Mariam Kemal, MD, Mohamed Sheta, MD Monmouth Medical Center: Department of Medicine

## CASE REPORT

A 53 year-old right-handed male with a history of Fabry's disease diagnosed 30 years earlier after development of peripheral neuropathy in addition to strong family history, presented with right hemiparesis and 2 days of lethargy. For his Fabry's disease, he received enzyme replacement therapy (Agalsidase beta) for 3 years, but it was discontinued 6 months prior to this admission. At presentation, MRI showed extensive bilateral cerebellar and pontine hypodense areas as well as hydrocephalus. An emergent external ventricular drain was placed, but the clinical course rapidly deteriorated and the patient expired. A limited brain autopsy showed acute anoxic change in the cerebellum and slight to moderate arteriosclerosis with mineralization of vessel walls in basal ganglia and cerebellum.

#### DISCUSSION

Fabry's disease is an X-linked recessive lysosomal storage disease caused by alpha-galactosidase A deficiency. Stroke occurs in 5.6% of patients; cerebral involvement is mainly due to vasculopathy, and 87% of the strokes are ischemic. Carrier females may also develop cerebrovascular complications. A first stroke tends to occur in patients in their early 40s. <sup>1,2,3</sup> In one study, brain MRIs performed in 50 patients showed increased disease burden with age, but

no patients younger than 26 years had lesions on MRI and all patients older than 54 years had cerebrovascular involvement.<sup>4</sup> Anti-platelet agents, statins, and blood pressure control are recommended for prevention of cerebrovascular accidents but no clear proof of their benefits exists. The role of enzyme replacement therapy remains unclear. Pharmacological chaperones are still under investigation.<sup>5</sup>

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Case Report: SUNCT - A Typical Case Of Short-lasting, Unilateral, Neuralgiform Headache Attacks

Aparna Kumar, MD Abington Memorial Hospital: Department of Medicine

## CASE REPORT

69 year-old male presented with an intense, acute, sharp pain behind his right eye lasting 5-15 seconds per event, with frequency of every 10 minutes. Symptoms started in the morning. It was associated with redness and lacrimation of the right eye. Patient had a root canal surgery a few weeks prior. There was no history of trauma or any other neurological symptom. On examination he was alert and oriented. There was right eye lacrimation and injection; pupils were equal and reactive with no ptosis; no other focal neurological signs. Patient did not respond to treatment with 100% oxygen and intranasal and

intravenous sumatriptan. Imaging studies were negative for mandibular abscess, carotid dissection and cranial venous thrombosis. MRI brain showed normal orbits and an old small parietal infract. Patient's TSH, ESR and ANA screen were normal. Patient was treated with intravenous phenytoin which relieved his symptoms. He was discharged on carbamezepine. Patient had resolution of symptoms on his follow up after a few weeks with no reoccurrence.

#### DISCUSSION

SUNCT is a rare headache syndrome which is a subtype of trigeminal autonomic cephalalgia characterized by

paroxysmal short acting unilateral pain in the ocular/periocular area. It is associated with ipsilateral conjunctival injection, lacrimation and nasal stuffiness. The etiology and pathogenesis are unknown. Incidence of SUNCT is 1:100,000 in USA. This case illustrates a classical presentation of an unusual cause of a very common symptom—headache. It is important to be aware of as the treatment of SUNCT is very different from other causes of headache.

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S. Jeffrey Yakish, DO, Arvin Narula, MD, Simon Smith, DO, Andrew Kohut, MD, Steven Kutalek, MD Drexel University College of Medicine: Department of Medicine, Division of Internal Medicine

Case Report: Superior Vena Cava Echocardiography Detecting Lead Fibrosis prior to Device Extraction

#### CASE REPORT

A 76 year-old man received an ICD four years ago for ventricular tachycardia. The patient developed a pocket infection which prompted a device extraction. Prior to extraction transthoracic echocardiography was performed by placing the transducer in the right supraclavicular fossa between the two heads of the sternocleidomastoid muscle. The color Doppler demonstrated turbulent flow at the junction of the superior vena cava (SVC) and innominate vein. The first two leads were able to be removed with gentle traction, while the third demonstrated heavy fibrosis in the right atrium with direct traction. Specialized tools including a locking stylet and a laser sheath were required to break up the fibrosis and remove the lead.

## DISCUSSION

When device extractions go awry, the effects may be life threatening with complications that include laceration of

Figure 1. Supraclavicular view of Innominate and SVC

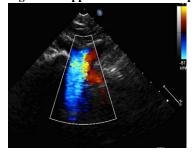


the right atrium, SVC or innominate vein. Currently there is no non-invasive way to predict the difficulty of extraction. Our case illustrates transthoracic echocardiography as a useful tool in assessing fibrosis preoperatively. Transthoracic echocardiography, in the SVC window, was able to detect turbulent flow by color Doppler that correlated with fibrosis later confirmed during extraction. If SVC echocardiography is sensitive for detecting fibrosis it may serve as a screening tool to guide future management of device extractions.

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Figure 2. Doppler of SVC - mosaic pattern = turbulent flow



## Case Report: The Role of Clinically Applied Medical Geography in Diagnosis

Osamuyimen Igbinosa, MD, David Alcid, MD Saint Peter's University Hospital: Department of Medicine

## CASE REPORT

50-year-old woman presented with rigor, headache and fever of 5-days duration, accompanied by nausea, myalgia, and loose brown stool. She lives in New Jersey. She recently returned from a 2-week trip to Machu Picchu where she had many insect bites. She did not take malaria prophylaxis. On physical exam: temperature 103.8F, pulse 108/min, respiratory 22/min. She was awake with rigor. No

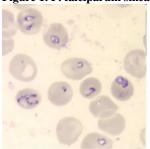
jaundice, tachycardic but regular; spleen tip was palpable and no focal neurological deficit. Laboratory data revealed; Hgb 9.8, WBC 4800, 75% polys 25% lymph's, platelet count 48,200, BUN 28, creatinine 1.9. Smear positive for Plasmodium falciparum (Fig 1). Patient was started on quinine and doxycycline, however she continued to be sick. Platelets dropped to 38,900, fever and rigors continued after 3 days of therapy. Tropical disease specialist was

consulted, smear reviewed by consultant turned out to be Babesia spp (Fig 2). She was started on atovaquone and azithromycin. She began to defervesce after 24 hours.

#### DISCUSSION

We present our experience in the care of a febrile Chinese woman who had just returned from Machu Piccho, Peru. This case highlights importance of knowledge about world geographical prevalence of diseases. It is not uncommon for laboratory technicians to misinterpret babesiosis for malaria but clinicians should be guided based on pretest

Figure 1. P. falciparum smear with 2% parasitemia

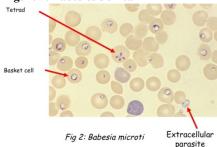


probability of diseases. Our patient likely acquired babesia in New Jersey. Machu Picchu stands in the middle of a tropical mountain forest located 2,430 meters (7,970 ft) above sea level; the risk of malaria in travelers is very low.

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Figure 2. Babesiosis smear



Case Report: Tubal Occlusion with Adiana Prior to In Vitro Fertilization in a Patient with Hydrosalpinx and Multiple Abdominal Surgeries

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## CASE REPORT

32 year-old Caucasian nulligravida female presented with complaints of infertility for one year. Infertility work up and all tests were normal with exception of a hysterosalpingogram which revealed bilateral hydrosalpinx. The patient's past medical history is significant for Crohn's disease, anemia, and hypothyroidism. Gynecologic history includes regular menses occurring every 26 days with no significant dyspareunia or dysmenorrhea. She has a past surgical history of two prior bowel resections secondary to Crohn's and had an attempted exploratory laparoscopic surgery which was aborted due to extensive adhesions. The patient underwent a hysteroscopic Adiana tubal occlusion procedure. The patient's 3 month follow-up HSG showed bilateral tubal occlusion. The patient then went on to have successful IVF treatment followed by a normal spontaneous vaginal delivery at term.

## DISCUSSION

To our knowledge, no prior cases of IVF following Adiana tubal occlusion have been reported in the literature. We believe this method to be safe and a time-/cost-effective alternative to laparoscopic salpingectomy. Adiana is sa afe alternative to Essure for transcervical sterilization in

patients with hydrosalpnix with multiple abdominal surgeries. It can also be used in patients who have nickel allergy who are not candidates for Essure hysteroscopic tubal occlusion. This case illustrates the successful use of Adiana for distal occlusion of hydrosalpinx in a patient who plans to undergo IVF. This procedure will further enhance success in IVF pregnancy in patients who are poor candidates for laparoscopic and abdominal procedures, though more studies are needed.

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## **Case Report: Uterine Rupture**

Monica Suliman, MD, Robert Massaro, MD Monmouth Medical Center: Department of Obstetrics and Gynecology

#### CASE REPORT

A 32 year-old G2P1001 female at 39 weeks gestational age with gestational diabetes controlled on Glyburide 1.25mg twice daily presented complaining of contractions. Patient had a history of a prior full-term normal spontaneous vaginal delivery in 2007 and no significant past medical or surgical history. Patient was noted to be 3/80%/high upon admission and progressed to dilation of 9cm over six hours without any augmentation. It was noted that patient had persistent worsening variable decelerations to the 60's that resolved back to baseline. Secondary to non-reassuring fetal heart tracing, a primary cesarean section was performed. Upon entrance a subserousal hemorrhage on the uterus was visualized with a low vertical uterine rupture in which the infant was delivered with appars 9, 9 weighing 3995gms. Patient recovered without any further complication.

#### DISCUSSION

Uterine rupture occurs in approximately 1:1,235-3,000 of

pregnancies with 90% of them occurring in patients with prior cesarean scars. <sup>1</sup> It is estimated that uterine rupture occurs in 1:12,960 deliveries with an unscarred uterus. Common etiologies of uterine rupture in an unscarred uterus include grand-multiparity, macrosomia, and labor augmentation. <sup>2</sup> This case presents a rare instance of uterine rupture in a female with an unscarred uterus and no risk factors including no augmentation to labor. Of interest, this patient did not show any hemodynamic compromise. As rare as uterine rupture is it may occur in patients with no risk factors as highlighted in this case.

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## Case Report: Vocal Cord Dysfunction (VCD)

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#### CASE REPORT

A 12-year old boy presented to the ER with 1 hour of acute onset shortness of breath triggered by brushing of teeth. On physical exam vitals were stable. Patient had a diagnosis of asthma. Over the past year, patient had multiple hospitalizations, one PICU admission and multiple ER visits for similar symptoms. There is strong family history of GERD. Bronchoscopy was consistent with mild laryngomalacia, omega shaped epiglottis and mild bronchomalacia. Bronchial washings showed numerous lipid laden macrophages. Upper GI series showed severe reflux. Spirometery showed a normal FEV1: FVC. Pulmonary function test showed flattening of the inspiratory limb of the flow volume loop at the beginning of the episode which became normal as the episode waned suggesting VCD.

#### DISCUSSION

VCD produces abnormal adduction of the vocal cords during inspiration. It mimics persistent asthma<sup>1</sup> with

symptoms of sudden onset of dyspnea not responsive to bronchodilators and is often treated with high-dose inhaled or systemic corticosteroids, bronchodilators, hospitalizations, tracheostomies and intubation. In VCD FEV1:FVC is normal with flattening of the inspiratory Flow Volume Loop. We suggest that refractory asthma with poor response to beta-agonists or inhaled corticosteroids be investigated further for VCD. Gastroesophageal reflux, psychiatric conditions, emotional stress, upper airway inflammation, strenuous exercise, exposure to irritant fumes, and environmental allergens could lead to VCD.

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Figure 1. Flattening of inspiratory portion of flow-volume loop



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## Case Report: Water Immersion Endoscopy and the Use of Underwater Argon Plasma Coagulation in the Control of Upper Gastrointestinal Bleeding

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- \*\*Drexel University College of Medicine: Department of Medicine, Division of Gastroenterology and Hepatology

#### CASE REPORT

An elderly gentleman with a history of arteriovenous malformation disease presented with acute anemia. Endoscopy was performed and clotted blood was found in the gastric lumen which was cleared using extensive irrigation and suction. An actively oozing fundic AVM was identified (F3,F4). The lesion was identified while bleeding and it stopped bleeding on its own. A large amount of water was instilled and the lesion was visualized underwater (F3,V1,V2). The area was treated safely and successfully with Argon plasma coagulation (APC) (F4,F5,V3), which resulted in typical coagulation pattern following treatment.

#### DISCUSSION

Underwater Endoscopy (UE) is a commonly used technique to study the gastrointestinal tract. <sup>1,2</sup> Water as a medium between the scope and the mucosa improves images by reducing glare and reducing total reflections which reduces image quality (F1,F2). Arteriovenous malformation bleeding can be very difficult due to the spread of blood over a large part of the stomach as lesions continue to ooze. Continuous irrigation is employed to keep the area of oozing clear of blood for targeting therapy.

Water immersion endoscopy has been employed for better visualization and targeted therapy. Water immersion helps by weighing down the dependent portion of the stomach, hence decreasing motion. It also helps in localizing the area by avoiding dispersion of blood as it oozes due to the difference of the fluid densities. This case shows the ease of localizing the bleeding lesion and the first description of safe and successful use of APC underwater.

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Editor's Note: See Medical Essay on p.69 re: QR code use

Figures - Scan QR Codes for Picture/Video Links









## Case Report: When a Simple Earache is not that Simple

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## CASE REPORT

A 37 year-old male was seen in the ER complaining of severe left ear pain and fever. Examination of his left ear revealed purulent discharge in the external auditory canal with erythematous tympanic membrane. His physical examination was also significant for conjunctival pallor and hepatosplenomegaly. CBC revealed Hgb 7.7, WBC 2.9 x  $10^3$ , and platelet 99 x  $10^3$ . CMP showed creatinine of 1.33 with total protein of 10.3. SPEP and serum immunofixation showed an IgG kappa monoclonal protein of 4.6 in the gamma globulin region. CT of the abdomen showed numerous metastatic lesions of the liver and spleen, and the

highly aggressive appearance of multiple lytic bone metastases. Bone marrow biopsy revealed more than 70% of monoclonal plasma cells.

## DISCUSSION

Earache is a common medical problem and most of its causes are benign. We present a case of earache which is caused by a very uncommon underlying condition. This case is different from a typical multiple myeloma in that the presenting signs and symptoms of the patient have more features of leukemia (severe infection, pancytopenia, and significant involvement of liver and spleen) than those seen

in multiple myeloma. This highly aggressive clinical presentation suggests a more aggressive form of multiple myeloma, namely plasma cell leukemia. Since early detection of such diseases is crucial to initiate proper treatments, physicians should be able to recognize when an earache is not a simple infection, and may be the presenting feature of an underlying sinister illness.

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Figure 1. Axial CT View



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Figure 2. Coronal CT View



## Article: A Rare Case of Metachronous Skip Metastasis of Pancreatic Cancer to the Colon

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#### ABSTRACT

Pancreatic cancer metastasis to the non-adjacent colon is rare, even more so without liver metastasis. The patient was two years status post curative Whipple's procedure. This case is both extremely rare and unique because it is the first to demonstrate metachronous metastasis originating from primary pancreatic malignancy to non-contiguous colon sans liver involvement. The histological determination of the tissue of origin of the metastatic lesion, using immunohistochemical staining, is also described.

## INTRODUCTION

Pancreatic cancer is the fourth leading cause of cancer mortality in the US¹ with 5-year mortality even after curative resection at about 15%.² This mortality rate is closely related to the stage of the disease, including the presence or absence of metastasis. Common sites of metastasis include peripancreatic lymph nodes, liver, lung and peritoneum.³ Distant involvement of the colon is rare. This report presents a first-in-literature case of metachronous metastatic pancreatic cancer to the colon, causing large bowel obstruction, two years after curative resection of the primary pancreatic tumor. The definitive determination of the tissue of origin of the colonic mass was done using immunohistochemical staining.

## CASE REPORT

An 85-year-old Caucasian female with past medical history of stage IIA pancreatic cancer, status post pancreatico-duodenectomy 24 months prior, presented to our emergency room with a 1-week history of constipation and intermittent abdominal pain. Physical exam was significant for left lower quadrant tenderness and an epigastric hernia but no organomegaly or palpable mass. Initial laboratory data including liver function tests were essentially within normal limits except for a mildly elevated serum creatinine of 1.16. The patient had a CT scan of the abdomen and pelvis which revealed partial colonic obstruction at the mid-sigmoid with wall thickening and pericolic adenopathy, raising suspicion of a neoplasm in the sigmoid colon. There were no other lesions seen in the abdomen including within the residual pancreas and liver. Subsequent barium enema showed complete colonic obstruction to retrograde filling at the mid-sigmoid colon. Serum carcinoembryonic antigen (CEA) level was elevated at 4.3. A Hartmann's operation with adjacent lymphadenectomy was performed. There were no lesions found in any other abdominal viscera. She was discharged home 11 days after surgery with plans made for oncologic follow-up and a subsequent operation for bowel recontinuity.

## **PATHOLOGY**

Gross: There was a circumferential ulcerated mass measuring 2x3cm in greatest dimension within the 14x7cm specimen of sigmoid colon. There was puckering of the serosa and thickening of the mucosal surface corresponding to the location of the mass; however, there was no definitive extension of the mass through the wall of the colon. Histology: Extensive lymphatic invasion was seen in the wall of the colon with extension into and ulceration of the mucosa; however, no clear adenomatous change was noted within the colonic mucosa. Mucosal and radial margins were tumor-free. Hematoxylin and eosin stains showed identical histologic patterns with the patient's previously resected pancreatic adenocarcinoma. Lymph nodes were negative for metastatic carcinoma. Immunohistochemistry: Immunohistochemical staining showed strong positivity for cytokeratin 7 (CK7) and focal weak positivity for cytokeratin 20 (CK20). The tumor tissue was also positive to CA 19-9 testing. The histopathological pattern was compared to that of the patient's previously resected pancreatic cancer and results were essentially identical. The final diagnosis was moderately differentiated metastatic pancreatic adenocarcinoma involving serosa, wall of colon, and extending into colonic mucosa with ulceration and extensive angiolymphatic and perineural invasion.

## DISCUSSION

Pancreatic cancer is the second-highest cause of mortality among cancers of the gastrointestinal tract. It carries the poorest prognosis of all major cancers, mostly due to late-stage presentation, with more than half of patients presenting initially with locally invasive disease or metastastatic dissemination. Metastatic pancreatic cancer is associated with an overall 5-year survival of 5%. While pancreatic cancer can metastasize to the lungs, abdomen, regional lymph nodes, and peritoneum, it most commonly metastasizes to the liver<sup>3</sup> and rarely disseminates beyond

DrexelMed Journal, Volume 6 (1); Spring 2012

these areas. Even after curative resection of pancreatic cancer, local recurrence, liver metastases, and peritoneal spread are the most common recurrent patterns.<sup>5,6</sup>

Metachronous metastasis of pancreatic cancer to colon is extremely rare with no reported cases in literature. There have been previously reported cases of metastatic cancers between the pancreas and the colon. We found three of these on literature review. There were two cases of synchronous colon to pancreas metastases and one case of metachronous metastasis of colon cancer to the pancreas. Of note, none of these cases involved metachronous pancreatic metastasis to the colon.

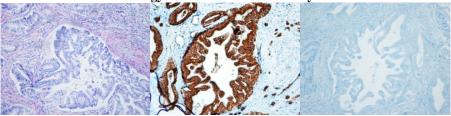
To our knowledge, our case is the first in literature because it involved metachronous pancreatic cancer to the colon without evidence of liver metastasis. Metastatic disease involving the colon especially without liver involvement from pancreatic cancer is an unusual clinical event. <sup>16</sup> This case provides additional clinicopathologic insight into the unique site and mode of metastasis of pancreatic cancer. We also outlined the utility of proper immunohistological staining techniques in proper identification. We tested the resected colon cancer for CK-20, CK-7 and CA 19-9 positivity, with the results outlined above. This served as an adequate histopathological correlate to microscopic findings and ultimately proved the site of origin to be pancreatic.

#### CONCLUSION

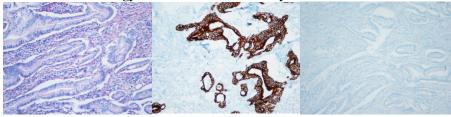
Metastatic pancreatic cancer to the colon is rare. Identification of the primary tissue of origin and type of malignancy is essential to determine the appropriate management required. Confirmation of the histological features can only be made using specific immunohistological stains as outlined in the above case, when the tumor was found to be of pancreatic origin.

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## Pancreatic Cancer Histology & Immunohistochemistry







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Article: Anemia in Women of Child-Bearing Age in a Resident Community Care Clinic: A Study to Help Identify and Reach the National Goals

Kaihong Mi, MD, Lan Cao, MD, Mahesh Krishnamurthy, MD, Ghada Mitri, MD, Rajen Oza, MD, Susan Sloan, MD Easton Hospital: Department of Medicine

## **OBJECTIVE**

To investigate the prevalence of iron deficiency anemia in women of child bearing age (18-49 years old) in our Community Care Clinic for 2010 and to compare the results to the national 2010 goal. Our goal is to improve the treatment of iron-deficiency anemia and quality of care provided to a vulnerable patient population.

#### **METHODS**

Utilizing a retrospective design to determine the prevalence of iron-deficiency anemia among women aged 18-49 in our Community Clinic, charts for all women aged 18-49, who visited the Clinic between January 1, 2009 and December 31, 2010, were reviewed. Exclusion criterion included women who had no CBC done during the mentioned period. Hemoglobin value, mean cell volume, erythrocyte distribution width and iron studies results were recorded. In addition, a diagnosis of iron-deficiency anemia was also made if hemoglobin concentration or hematocrit increased after a course of therapeutic iron supplementation. If the woman had several CBC tests between 2009 and 2010, the data from the most recent test was recorded. Additional data drawn from the patient chart included history of iron supplementation, age, and race.

## **RESULTS**

244 charts of women 18-49 years old (yo) in our community care center were reviewed for the presence of iron deficiency anemia. The majority were Caucasian. The three age groups (18-30yo, 31-40yo, and 41-49yo) were distributed equally (Fig 1). We found that 29% of women between 18-49yo had anemia with Hemoglobin less than 12.1mg/dl. 14% had microcytic anemia, of which 54% were iron deficient, mounting up to 7.8% of our study group. The status of 46% of the microcytic anemia was unknown because confirmatory iron studies were not available and there was no evidence for hemoglobin or hematocrit improvement after a course of iron supplements (Fig 2).

African American and Asian women were the two groups with the highest rate of iron deficiency anemia, 15% and 16.7% respectively. It is worth noticing that 55% of the African American women had anemia, which is significantly higher than the other groups (Fig 3A). The number of patients in each age group (18-30yo, 31-40, and 41-49yo) was relatively equal. Iron deficiency was seen highest in the age group 31-40 years, which accounts for nearly 60% of all confirmed iron deficiency anemia patients (Fig 3B).

We also reviewed whether iron deficiency anemia patients were taking iron supplementation. About 84% of iron deficiency anemia patients were taking iron supplemental pills daily, while 5% were taking folic acid and 10% were taking nothing (Fig 4). In patients with microcytic anemia without iron studies, 6% were taking iron and 12% were taking multivitamin.

#### CONCLUSIONS/DISCUSSION

Data collected in the National Health and Nutrition Examination Survey (NHANES) suggests that low iron levels are a serious problem for many people, including women of childbearing age, preschool children, and the elderly. As a result, the US government has encouraged fortification of grain and cereal with iron to correct this deficiency. In the past three decades, increased iron intake among infants has resulted in a decline in childhood iron deficiency anemia in the United States. However, the prevalence of iron deficiency among all young children remained the same, and the prevalence of iron deficiency among females of childbearing age actually increased. Iron deficiency has remained prevalent among low-income women during pregnancy with no improvement noted since the 1970s. NHANES III indicated that 11% of non-pregnant women 16-49yo had iron deficiency and that 3%-5% also had iron deficiency anemia. As a result of this deficiency, one of the national health objectives for 2010 was to reduce by 3%-4% iron deficiency in these vulnerable populations. The 2010 National health objectives for the prevalence of iron deficiency in females aged 12-49 years is less than 7%.

Our study was completed in a Resident Community Care Clinic. The majority of the clinic patients were from low social economic status. 7.8% of our study group was confirmed as iron deficient anemia by either iron study or evidence for hemoglobin or hematocrit improvement after a course of iron supplements. The actual number may be double since near half of the microcytic anemia in this study group does not have documented iron studies or evidence of hemoglobin or hematocrit improvement with iron supplements. The prevalence of iron deficiency anemia in our clinic was much higher than the 2010 national health objective of the prevalence of iron deficiency. This study revealed at least 2 areas in which our clinic can improve: 1) Iron studies should be ordered for all patients with microcytic anemia; 2) Increasing patients' compliance in finishing the ordered tests and taking iron pills. We are pursuing strategies to decrease the incidence of iron deficiency anemia in our community and improve the standards of care. The underlying etiology for non-adherence and improving compliance is our next study.

Overall, small population studies like ours may help in identifying trends that would then help us in devising strategies to improve the health of our communities. Our hope is that with adequate identification and treatment, we will be able to achieve and better the national health objectives for iron deficiency in a vulnerable population.

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Figure 1. General Study Group Information

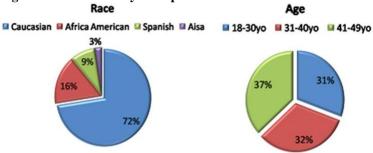


Figure 2. Prevalence of Anemia

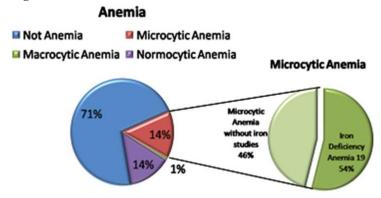


Figure 3. Anemia by Race (A) and Age (B)

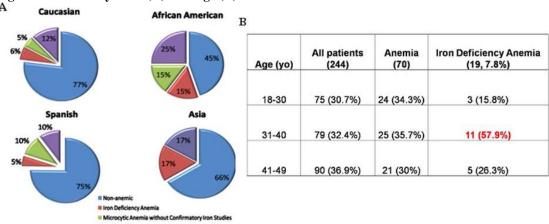
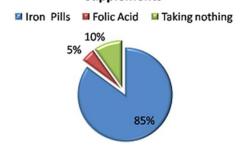


Figure 4. Supplement Use in Iron-Deficient Patients Iron Deficiency Anemia on supplements



## Article: CT-imaging to Evaluate Radiation-induced Pre-clinical Myelopathic Changes

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## **OBJECTIVES**

Radiation myelopathy is a relatively uncommon, but potentially devastating, complication of therapeutic irradiation. It incorporates a spectrum of syndromes, ranging from transient radiation myelopathy to chronic progressive radiation myelitis. The literature indicates that cord damage is related to a number of factors, some of which include dose fraction size, total dose, treatment time, length of cord irradiated, and individual patient sensitivity. In addition, both pathologic as well as radiologic (MRI) changes to the spinal cord have been well described in patients with radiation-related myelopathy. The management of such patients remains difficult; however the early detection of radiation-induced spinal cord damage may facilitate treatment in preventing the clinical symptoms of myelopathy. Although MRI surveillance of the spinal cord is not routinely done in asymptomatic patients, CT scans are used for active surveillance and are readily available. The aim of our study was to determine whether a CT can be used to assess post-radiation therapy, pre-clinical changes to the spinal cord.

## MATERIALS/METHODS

We identified a total of 39 patients with head-and-neck or lung cancer who were treated with external beam radiation between January 2006 and November 2010 at Hahnemann University Hospital. On each treatment planning-CT, the isocenter was identified, and the maximum cord dose at that level was determined. We then identified the axial cut corresponding to the isocenter (in-field) as well as a cut outside the RT field (control) on a pre- and post-treatment CT (PrT-CT and PoT-CT) surveillance scan. We measured the antero-posterior and lateral cord diameter on 3 contiguous axial cuts for both the PrT-CT and PoT-CT and calculated the mean cord area and percent change for each patient. Pearson correlation was used in our analysis.

#### RESULTS

Median age was 71 (range 46 - 90), median tumor dose was 63.0 Gy (range 23.0 - 74.0 Gy), median cord dose was 38.55 Gy (range 3.66 - 45.11 Gy), and median time to PoT-CT was 249 days (range 19 -1335). At last follow-up (median 319 days, range 43 - 1383), there were no reports of myelopathy. In-field mean percent change of PrT to PoT cord area was -5.66% (range -63.28 to 28.24, SD=19.68). Correlation of percent change in cord area to cord dose was -0.204 (p=0.213). Controlling for time to PoT-CT, the correlation was -0.183 (p=0.271). Sub-analysis for acute (<90 days post-RT) and late (>90 days post-RT) changes was also not statistically significant (p=0.469 and p=0.462). For the control, the mean percent change in the spinal cord area was smaller at -1.12% (range -23.55 to 33.08, SD=10.28). There was no correlation between the change in cord area inside compared to outside the RT field (Pearson coefficient 0.187, p=0.254).

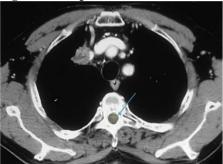
## **CONCLUSIONS**

As technological advances in radiation translate to improved survival and allow for re-irradiation of tumors, incidence of post-RT myelopathy is likely to increase. Based on our experience, post-RT surveillance CT-scans are inadequate at showing radiation changes to the spinal cord. There does appear to be a larger surface area variability in the irradiated spinal cord, even with cord doses <45 Gy, as compared to the non-irradiated cord. A study comparing MRI versus CT imaging is necessary to determine if an MRI would be better at evaluating post-radiation changes to the spinal cord and predicting potential post-RT myelopathies.

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Figure 1. Sample Measurement – CT



**Table 1. Study Members Descriptive Statistics** 

	<u> </u>		1			
	N	Range	Minimum	Maximum	Mean	Std. Deviation
Age	37	44	46	90	69.78	11.405
Treatment code 3D=1, IMRT=2	37	1	1	2	1.11	.315
Total dose to tumor	37	51	23	74	57.62	12.632
Total cord dose	37	41.45	3.66	45.11	27.5314	16.90812
# Fractions	37	29	10	39	30.24	7.765
# elapsed days	37	85	15	100	50.00	16.750
# days post treatment	37	1316	19	1335	375.49	331.359
Valid N (listwise)	37					

Table 2. Cord Dose & % Change in Cord Area – Acute

		# days post	percent change cord area (pre to	
		treatment	post tx)	Total cord dose
# days post treatment	Pearson Correlation	1	.418	664
	Sig. (2-tailed)		.302	.072
	N	8	8	8
percent change cord area (pre to post tx)	Pearson Correlation	.418	1	142
to poor on	Sig. (2-tailed)	.302		.738
	N	8	8	8
Total cord dose	Pearson Correlation	664	142	1
	Sig. (2-tailed)	.072	.738	
	Ν	8	8	8

Table 3. Cord Dose & % Change in Cord Area – Late

			percent change	
		# days post	cord area (pre to	
		treatment	post tx)	Total cord dose
# days post treatment	Pearson Correlation	1	.223	150
	Sig. (2-tailed)		.246	.436
	N	29	29	29
percent change cord area (pre to post tx)	Pearson Correlation	.223	1	294
(pro to post or)	Sig. (2-tailed)	.246		.121
	N	29	29	29
Total cord dose	Pearson Correlation	150	294	1
	Sig. (2-tailed)	.436	.121	
	N	29	29	29

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## Article: Direct Bilirubin Levels are usually Unnecessary in the Newborn Nursery

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#### INTRODUCTION

The evaluation and treatment of jaundice in the newborn period is to prevent severe hyperbilirubinemia and its associated sequelae, which include encephalopathy, athetoid cerebral palsy, auditory dysfunction, paralysis of upward gaze, and intellectual dysfunction. Hyperbilirubinemia in the first week of life reflects transition from an intrauterine to an extrauterine pattern of bilirubin transport and metabolism. After birth unconjugated bilirubin levels increase and are reflected in total serum bilirubin. The predominant cause of unconjugated neonatal hyperbilirubinemia is physiologic jaundice seen in 13% of breast-fed newborns in the immediate postnatal period. Additionally, in 15% of pregnancies, ABO blood group incompatibility will later lead to isoimmune hemolytic disease of the newborn. Neither of these etiologies results in elevation of conjugated bilirubin. The 2004 American Academy of Pediatrics Clinical Practice Guidelines for Treatment of Neonatal Jaundice (=35 weeks gestational age) recommends measuring direct bilirubin levels in four circumstances: 1) when phototherapy is to be instituted, 2) if the patient is persistently jaundiced after 2-3 weeks of life, 3) an unexplained rise of the serum bilirubin without a plausible cause (such as ABO incompatibility), and 4) sepsis. The AAP does not recommend measuring direct bilirubin in the first week of life in well-appearing late preterm or term infants. We have observed that obtaining direct bilirubin alone or along with total bilirubin is done frequently by pediatricians in the regular nursery in Monmouth Medical Center.

#### **PURPOSE**

To quantify the occurrence of direct hyperbilirubinemia through a retrospective chart review of newborn infants born at our level 1 newborn nursery.

## **METHODS**

We retrospectively reviewed all infant's charts admitted into the newborn nursery at Monmouth Medical Center who had a direct bilirubin measurement from January through August 2009. It is a Level 1 nursery, admitting newborns from the delivery room if they satisfy the following criteria: 1) gestational age =35 weeks, 2) birth weight = 2000 grams, and 3) no abnormalities during delivery and initial assessment after birth. Infants were selected through a database search using current procedural terminology (CPT) code 82248 (direct bilirubin). The first 1,000 patients who had a total and direct bilirubin initially assessed on either the first, second, or third day of life secondary to jaundice were included in the analysis (n=1000). Through manual review of charts, the following patient characteristics were obtained: 1) gestational age, 2) total and direct bilirubin values, 3) initiation of phototherapy, 4) direct antiglobulin testing (DAT), and 5) complete blood count (CBC) for DAT positive infants. Direct hyperbilirubinemia was defined using guidelines from the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN). The society defines abnormal conjugated bilirubin as a direct bilirubin =1 mg/dL if total bilirubin is <5 mg/dL or direct bilirubin is >20% of the total bilirubin measurement = 5 mg/dl.

## **RESULTS**

1000 newborn charts were reviewed. Demographic breakdown (n (%)): male 529 (52.9%) female 471 (47.1%) Caucasian 795 (79.5%), Hispanic 132 (13.2%), African American 55 (5.5%), Asian 18 (1.8%), Gestational Age, 35-36 6/7 weeks 49 (4.9%), 37 -41 6/7 weeks 940 (94.0%), 42+ weeks 16 (1.6%). Of 1,000 newborns, none had an elevated direct bilirubin, as defined by NASPGHAN clinical guidelines (Table 1). In the study population, only 2 newborns had initial direct bilirubin values > 1.0 mg/dl (1.1 mg/dl, 1.4mg/dl), with total bilirubin values 12.8 mg/dl and 12.2 mg/dl respectively. Their ratios of direct to total bilirubin were well below 20% (Table 2). Only 43 infants had direct bilirubin measurements prior to phototherapy (Table 3). Of the newborns who received phototherapy, 43 patients had subsequent prior direct bilirubin measurements, with 7 patients having direct bilirubin values > 1.0 mg/dl, with none of their direct bilirubin values being greater than 20% of the total bilirubin, indicating normal levels (Table 4).

## DISCUSSION

Direct bilirubin levels are measured commonly in our newborn nursery population despite recommendations to the contrary. Increasing total bilirubin levels in the first three days of life is an expected occurrence in many neonates, but the likely cause is due to indirect hyperbilirubinemia. Direct hyperbilirubinemia has been noted in various

disease states, with an overall incidence in 1 in 2500 newborn patients.<sup>5</sup> Three common causes of cholestatic jaundice in the neonatal population are biliary atresia (25%), neonatal hepatitis (15%), and alpha-1 antitrypsin deficiency (10%). NASPGHAN recommends a direct bilirubin measurement at two weeks of life if a newborn appears jaundiced to rule out biliary atresia. The management of biliary atresia involves re-establishment of bile flow (Kasai portoenterostomy) and the long term survival of the infant's native liver is improved when the procedure is performed before 45 days of life. Other causes include extrahepatic obstruction from common duct gallstones and choledochal cysts, hypertyrosinemia, galactosemia, inborn errors of bile acid metabolism, and Alagille syndrome, which are extremely rare to diagnose in the first three days of life in a well appearing baby.<sup>2</sup> During the study period, no abnormal direct bilirubin measurements were identified in the well appearing infants in the first 72 hours of life, or in the subgroup of newborns receiving phototherapy in the first week of life. Measuring direct bilirubin values in our laboratory costs \$1 if a total bilirubin measurement is already performed. This would result in unnecessary added expense in the management of well newborn infants. Despite the recommendations of the AAP, not all pediatricians obtain a direct bilirubin measurement before initiating phototherapy in our nursery. Phototherapy is the management of choice for unconjugated hyperbilirubinemia. Hence, measuring direct bilirubin prior to initiating phototherapy will confirm unconjugated hyperbilirubinemia as well identify infants at risk for Bronze baby syndrome. <sup>6,7</sup> Bronze baby syndrome is a benign condition where an infant presents with grayish-brown skin discolorations. It is an uncommon side effect noted in infants with cholestatic jaundice who received phototherapy. The serum of the patients who developed bronze baby syndrome has elevated levels of unconjugated and conjugated bilirubin. Another observation we made in the study was that all patients had direct antigen testing (DAT) irrespective of their mothers' blood group or Rh status. Of 1000 patients, only 36% were DAT positive. DAT is often positive predominantly in the setting of ABO or Rh incompatibility. 8 ABO incompatibility exclusively occurs only in O group mothers delivering A/B group infants. Rh incompatibility occurs exclusively only in Rh negative mothers with Rh positive infants. Hence cord blood DAT should be ordered only in infants of O group or Rh negative mothers. This would be an evidence-based and cost effective approach.

## CONCLUSION

Direct bilirubin measurements in the jaundiced newborn are a low-yield laboratory examination in the regular newborn nursery in the absence of specific risk factors. Pediatricians should adhere to AAP recommendations and not routinely obtain direct bilirubin levels on well appearing newborn's for the evaluation of neonatal jaundice.

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Table 1. Bilirubin Analysis

	Number
T. bilirubin ≥5.0 mg/dL	656
T. bilirubin <5.0 mg/dL	344
Direct Bilirubin≥1.0mg/dL	2
Direct Bilirubin < 1.0mg	998
Direct Bilirubin >20% of Total Bili	0

**Table 2. Initial Direct Bilirubin ≥ 1.0 mg/dl** 

	Direct bili (mg/dl)	Total bili (mg/dl)	Direct/Total Bilirubin
Patient 1	1.1	12.8	8.59%
Patient 2	1.4	12.2	11.47%

Table 3. Analysis of Bilirubin before Phototherapy

N of phototherapy	100
Total Bilirubin ≥5.0mg/dl	100
Direct Bilirubin measured	43
Direct Bilirubin > 20% of Total Bilirubin	0

Table 4. Direct Bili ≥1.0 mg/dl prior to Phototherapy

	Direct bili	Total bili	Direct/Total Bili
Patient 1	1.0	22.4	4.46%
Patient 2	1.1	24.4	4.50%
Patient 3	1.0	20.3	4.92%
Patient 4	1.4	13.0	10.76%
Patient 5	1.0	14.3	6.99%
Patient 6	1.0	13.1	7.63%
Patient 7	1.0	16.6	6.02%

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## Article: Endovascular Repair of Aneurysmal Aberrant Artery in Adult Pulmonary Sequestration

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## **ABSTRACT**

A case of intralobar sequestration supplied by a 3-cm aneurysmal vessel originating from the abdominal aorta is reported here. This was discovered in an abdominal CT scan performed due to patient's presenting symptom of severe, persistent abdominal pain. The vessel was repaired using an endovascular technique and embolized using an Amplatzer ® vascular plug (AVP) (AGA Medical Corp, Plymouth, Minnesota). The patient's symptoms improved post-operatively and she was discharged after 1 day.

## INTRODUCTION

Pulmonary sequestration refers to a pulmonary lobe or portion of a lobe that is supplied by an anomalous systemic artery, usually branching from the abdominal aorta. The lobe drains into either systemic or pulmonary veins. Sequestration can be intralobar, in which the sequestration is situated inside the visceral pleura of a normal lobe, or extralobar, where the sequestration is surrounded by its own pleura. The portion of pulmonary tissue supplied by this anomalous artery is non-functional and its airways are not connected to the tracheobronchial tree. Intralobar sequestration is most frequently diagnosed in childhood and rarely found in patients greater than 40 years old. Furthermore, only a few exceptional occurrences of intralobar sequestration supplied by a small aneurysm of the aberrant artery have been reported.<sup>1-3</sup>

## CASE REPORT

A 43 year-old Caucasian female was admitted for evaluation of epigastric pain of 1 day duration and a 1 year history of alternating constipation and diarrhea. She received an abdominal CT as well as an EGD, both of which were unremarkable. The patient was diagnosed with gastroenteritis and discharged, but returned to the ED 15 days later with persistent epigastric pain that now radiated to the left upper quadrant, as well as 1 episode of vomiting and a 4 day history of constipation. A repeat CT scan showed no abdominal pathology. An abdominal vessel aneurysm of approximately 3cm was noted. This vessel appeared to be extending through the diaphragm into the lower lobe of the right lung in the posterior basal segment (Figure 1). A CT arteriogram was performed and confirmed these findings (Figure 2). Since a complete abdominal workup was negative for this patient, the patient's pulmonary sequestration was considered the etiology for her abdominal pain. Embolization of the abdominal aortic aneurysm was performed using an Amplatzer ® vascular plug (AVP). An AVP 2 of 12 mm was advanced from the left femoral artery and deployed distal to the aneurysm. After this, flow through the anomalous vessel was markedly diminished. Then, an AVP 2 of 6mm was advanced and deployed proximal to the aneurysm. Within 5 minutes, a final hand injected digital subtraction run was performed and showed cessation of flow through the entire segment including the aneurysm. In Figure 3, the presence of dense clips can be seen indicating the location of both the distal and proximal AVPs. As is shown, flow has ceased after placement of these AVPs. The post-operative course showed an improvement of the patient's abdominal pain and the patient was discharged on post-procedure day 1.

#### DISCUSSION

Pulmonary sequestration is a rare congenital malformation in which a pulmonary lobe or portion of a lobe is supplied by an anomalous systemic artery. Most patients with pulmonary sequestration are diagnosed in their early life or during the first decade because of the early appearance of symptoms, including feeding difficulty, cyanosis,

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and dyspnea, or because of symptoms arising from the associated congenital abnormalities. In adults, the presenting symptom is usually persistent and recurrent pulmonary infections. <sup>8</sup> Due to the risk of infection, the current recommendation is resolution of sequestration to prevent future complications. <sup>8</sup> Current standard of care in adults has been the use of lobar resection via a transthoracic approach. <sup>1-4</sup> However, new research using endovascular techniques has indicated that this may be equally effective while causing less side effects and posing a lesser risk to patients. As was the case in this patient, very few cases of aneurysmal supply to a pulmonary lobe have been reported. In previous instances, the lobe and anomalous artery were resected through a transthoracic approach as per current recommendations.<sup>1-4</sup> Furthermore, while AVPs have been used to embolize aberrant arteries in pulmonary sequestration in adults in a small number of cases,<sup>5</sup> this is the first incidence of their use to embolize an aneurysm in pulmonary sequestration. Embolization has been an accepted alternative to surgery in pediatric pulmonary sequestration.<sup>6</sup> However, major concerns with embolization in adults include possible incomplete occlusion of vascular supply, subsequent evolution of sequestered tissue, possible recurrence of symptoms, migration of embolization material, or continued flow resulting in rupture of aneurysm. The use of AVPs, however, allows for a higher likelihood of complete occlusion of the vessel and reduces risk of distant migration of embolization material. In conclusion, pulmonary sequestration supplied by an aneurysmal arterial branch may be successfully managed by means of percutaneous endovascular treatment using AVPs with minimal complications. However, wider experience and longer follow-up are needed to propose endovascular treatment as an equally effective alternative to surgery in treatment of pulmonary sequestration in adults.

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Figure 1. Aneurysm and Vessel Extension



Figure 2. CT Arteriogram Confirmation

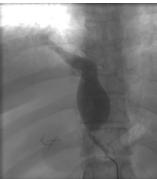
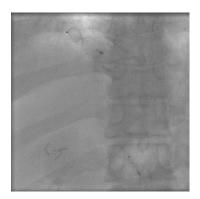


Figure 3. Cessation of Flow



# Article: Fecal Incontinence: Prevalence, Severity, and Quality of Life Data from an Outpatient Gastroenterology Practice

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#### ABSTRACT

<u>Background</u>: The prevalence of fecal incontinence (FI) varies tremendously as a result of diverse or inadequate data collection methods. Few office-based studies have assessed the prevalence of FI and none have looked at modifiable risk factors or effect on quality of life.

<u>Objective</u>: Our aim was to determine the prevalence of FI after direct questioning. We assessed symptom severity and impact on quality of life, and identified modifiable risk factors that could prevent the need for invasive procedures.

<u>Methods and Main Outcome Measures</u>: Five hundred patients who visited our inner-city, university-based gastroenterology practice were asked about symptoms of FI. Patients with symptoms then completed a demographic questionnaire, the FI Severity Index and the FI Quality of Life Scale. We also retrospectively reviewed 500 charts to identify the frequency of patient-physician reporting of FI in a non-structured office interaction.

Results: Of the 500 patients that were directly questioned, 58 (12%, 43 women, 15 men) admitted to FI compared to 12 (2.4%) in the retrospective arm. Patients with FI had a reduced quality of life predominantly in regards to coping and embarrassment. Forty-one of the 58 patients (71%) reported altered stool form. Individuals with loose/watery stool reported the lowest quality of life scores compared to those with formed (p=0.005), alternating (p=0.05), and all other stool consistencies combined (p=0.006). While the average severity score was similar between men and women, women had a significantly lower average quality of life score (3.04 versus 2.51; p<0.03).

<u>Conclusions</u>: Identification of FI increases when patients are directly questioned. Identifying and treating patients with loose stool is a potential strategy to improve quality of life in this patient population. In men and women with similar severity of FI, women have a significantly lower quality of life.

## INTRODUCTION

FI is defined as the inadvertent passage of stool, soiling, or excessive escape of flatus. The prevalence of FI varies among studies because of differing definitions of this disorder, patients' reluctance to report symptoms, and inadequate data collection methods. In the general community, the prevalence ranges from 0.4% to 18%. <sup>2,3-12</sup> Office-based studies document that 13% to 29% of patients in primary care and specialty clinics admit to FI when asked. <sup>7,10-11,13-14</sup> Studies from obstetrics and gynecology, urogynecology, and antenatal outpatient clinics report prevalence rates between 5.6% and 29%. <sup>9,14-15</sup> Johansen and Lafferty are the only group who has sampled patients from primary care and gastroenterology outpatient offices. However, this study was limited to a predominantly Caucasian population and did not evaluate the effect of FI on quality of life (QOL). <sup>16</sup>

The difference in prevalence of FI between men and women has similarly yielded variable results. The discrepancies are largely due to inconsistent data collection methods, variable ages, and reluctance to report symptoms. A large-scale systematic review found that 0.8% of men and 1.6% of women aged 15 to 60 years reported FI. In those older than 60 years, the prevalence increased to 5.1% in men and 6.2% in women. The most recent epidemiologic survey cites a similar prevalence in men and women (8.9% vs. 7.7%), whereas other studies cite a higher prevalence in men (20% vs. 11%, P<0.015).6,17 None, however, has examined the gender-specific effect on symptom severity and QOL.

It is evident from the aforementioned studies that the prevalence rises when FI is directly addressed. If incontinence does not pertain to the patient's chief complaint, however, many physicians may not inquire. Unfortunately, this perpetuates the "don't ask, don't tell" cycle. Better attempts to identify and treat patients with FI are essential, especially among physicians such as internists, gynecologists, and gastroenterologists, who are the most likely to treat this devastating condition.

We hypothesized that we could increase the identification of patients with FI by direct questioning during a routine gastroenterology office visit. We also hypothesized that we could determine if the severity of FI symptoms would correlate with quality of life. Our study intended to investigate these two hypotheses by adding the symptom of FI to our standardized gastrointestinal review of symptoms. We intended to evaluate QOL and severity of FI symptoms by administering validated questionnaires. In addition, we aimed to identify modifiable risk factors within this population that could be addressed and treated early to prevent the need for invasive procedures.

#### **METHODS**

The study was conducted at a Drexel University College of Medicine outpatient office. This office is an inner-city, single-specialty, university-based practice comprised of 8 gastroenterologists and 6 gastroenterology fellows who see outpatients with all types of gastrointestinal and liver disorders. Two of the 8 gastroenterologists specialize in motility disorders. The Drexel University College of Medicine Institutional Review Board approved this study.

The patients from this practice reflect an inner-city tertiary-care population. More than half of our patients are African American and have an annual income of \$25,000 or less. Approximately one-third of our patients have attended college or higher educational degrees. An equal number of men and women attend our practice. New patients are routinely asked to fill out a detailed but general review of systems while they wait to see our physician. During the visit, our history and physical forms (both new and follow-up) prompt our physicians to directly discuss a focused gastroenterology review of systems. Our gastrointestinal review of systems includes nine upper GI symptoms (dysphagia, odynophagia, heartburn, dyspepsia, abdominal pain, nausea, vomiting, melena, and weight loss) and eight lower GI symptoms (diarrhea, constipation, changes in bowel movements, decreased stool caliber, tenesmus, urgency, bleeding, and pain). FI is not included in either review-of-systems intake form. For this study, FI was added to the focused gastrointestinal review-of-systems intake form. Prior to the start of the study, all gastroenterologists were notified about the addition of FI to the standard review of systems intake form and were reminded to ask patients about this symptom. No restrictions were placed on the language physicians used to discuss this symptom with patients, with some gastroenterologists referring to "fecal incontinence" while others using terms such as "leakage or soiling."

During our study period, 500 consecutive patients (both new and returning) visiting our gastroenterology practice were asked about FI during the gastrointestinal review of systems. Patients who said yes were asked to enroll in the study and underwent informed consent procedures. Physicians then verbally administered three questionnaires to the enrolled patients. The first questionnaire included demographic factors; known medical, surgical, and obstetric risk factors; medications; duration of symptoms; need and frequency of pad or diaper use; and whether FI had ever been addressed by their health care providers. The second questionnaire consisted of the FI Severity Index (FISI), which we used to assess the frequency and type of stool loss. The third questionnaire consisted of the FI-specific American Society of Colon and Rectal Surgeons quality of life questionnaire (FIQL), which evaluates the impact of FI on coping, embarrassment, depression and lifestyle. We chose to use these scales because they allow subjects to weigh their answers. In turn, an external weighing scheme is employed for analysis. In addition, the validated FISI and FIQL questionnaires are easy to use, concise, reliable, and validated. 2,19

Our study also included a retrospective arm aimed to identify the number of patients who reported FI before it was included in the standardized gastrointestinal review of systems. In this part of the study, any documentation of FI would be the result of independent questioning by the physician or voluntary admission by the patient. Every third chart from our file room was selected until we reached 500 acceptable charts. Patients who were not seen within 3 years of the study were eliminated from this part of the study. Patients were excluded if they had been seen in our practice within 10 weeks prior to the study period. This prevented overlap with patients from the prospective arm. The selected charts were then completely reviewed for documentation of FI during any visit. Information regarding severity and effect on QOL was not assessed in this group because of the retrospective nature of the review.

The data obtained was analyzed statistically using Pearson's  $\chi 2$  test along with 95% confidence intervals (CIs) to compare groups of interest (men, women, and combined). Logistic regression analysis was used to identify independent associations with gender variables. A P value of <0.05 was considered statistically significant. We performed additional analyses using Microsoft Excel (Redmond, WA) t tests for comparison of subgroup variables.

#### RESULTS

Five-hundred individuals were approached over 3 consecutive months in the prospective arm of the study. Fifty-

eight (11.6%) reported symptoms of FI, including 74% of women (43) and 26% of men (15) with an average age of 51.7 years (range 22–84 years) and an average age of onset of 48.6 years (range 17-80 years). Approximately 90% of the patients who reported FI had a high school education or higher. Seventy-two percent of patients with FI had either medical or surgical risk factors for FI. Patient characteristics, duration and frequency of incontinence are summarized in Table 1.

Only 2 of the 58 patients (3%) in the prospective arm presented to the office with a chief complaint of FI. The remaining 56 (97%) reported incontinence only on direct questioning. Thirteen of the 58 individuals (22%) had discussed their symptoms with a physician in the past (Fig 1). On retrospective chart review, only 12 (2.4%) of 500 patients had any mention of FI anywhere in their outpatient charts. Gastroenterologists that specialized in motility disorders were not more likely to ask about symptoms of FI in either arm of the study.

Twenty-eight of the 58 patients with FI (48%) reported a poor quality of life (FIQL score <2.5). Each of the four FIQL scales was independently scored. Patients with FI had a significantly lower coping score than either lifestyle score (2.31 vs. 2.92, p<0.002) or depression score (2.31 vs. 2.98, p<0.003). Similarly, FI patients had a significantly lower embarrassment score than either lifestyle (2.37 vs. 2.92, p<0.006) or depression score (2.37 vs. 2.98 p<0.001) (Fig 2). When combined QOL scores were compared among groups, patients with loose/watery stools had significantly lower QOL scores when compared against the groups with formed stools (P=0.005), alternating loose/formed stools (P=0.05), and all groups combined (P=0.006) (Table 2).

Thirty-two of 58 patients (55%) had high severity scores (FISI score >25). When severity scores were compared among groups, patients with formed stool had relatively lower severity scores than all other stool-consistency groups alone and combined, although the findings were not significant.

At the time of visit, 41 of the 58 patients with FI (71%) reported altered stool form (loose/watery, hard, or alternating consistency). Of FI patients with loose/watery stool, six of 23 (26%) were taking laxatives and eleven of 23 (48%) were receiving no medical therapy at all, and only seven of 23 FI patients (30%) reported using antidiarrheal agents (Table 2).

Severity and QOL scores were evaluated separately for women and men in our cohort. The average FI severity score was not significantly different between men and women. However, women had a significantly lower average quality of life score than men (3.04 versus 2.51; P<0.03).

Pearson correlations were calculated for men, women, and men and women combined in regards to severity and QOL. For men, no significant correlation was found between severity and QOL (r = -0.09; P = 0.75; 95% CI -0.58 to 0.44). Conversely, a moderate correlation was found among women (r = -0.595; P < 0.001; 95% CI -0.68 to -0.36). As the severity of FI increases, therefore, the QOL decreases in women. A moderate correlation between severity and quality of life was also found when men and women were analyzed together (r = -0.505; P < 0.001; 95% CI -0.68 to -0.28) (Figs 3-5).

## DISCUSSION

Although estimates of the overall prevalence of FI range from 0.4 to 18% in the general community, it is clear that the reported prevalence rises when patients are directly questioned about symptoms of FI. In this study, 12% of patients reported FI when directly asked, whereas a prevalence of only 2% was revealed in our retrospective arm. This latter finding is surprising given that we conducted the study in an academic practice with expertise in motility disorders. Furthermore, this is testament to the fact that if doctors don't ask, patients don't tell.

Additionally, it was interesting that our FI patients had a reduced QOL predominantly as a result of issues with coping and embarrassment rather than depression and lifestyle issues. Although not officially validated, we ultimately chose to illustrate FIQL as a composite score of these main scales because we believe that the 4 scales of FIQL are simply facets of a total picture. In fact, Rockwood does state that with the FIQL "there is a sense that the overall quality of life is being assessed which is not true of other specialized scores such as those that assess depression or functional status." This approach should be further investigated.

While previously published studies examine the prevalence of FI among men and women or women alone, none focus on the gender-specific effect on QOL. Although no significant differences in severity scores were found

between men and women, women with FI were found to have a significantly lower QOL. In addition, QOL significantly correlated with severity in women but not in men. This finding raises multiple questions. Are women more embarrassed by this issue than men, thereby more negatively affecting their quality of life? Conversely, are men more embarrassed by this issue and thus more reluctant to report their symptoms or admit their deteriorating QOL? Or are men less emotionally bothered by the soilage?

There are many risk factors for FI; however only stool consistency is easily modifiable. That said the best initial approach to fecal incontinence is to identify and target treatment for bowel consistency. Patients with formed stool reported lower severity scores than patients with loose, hard or alternating bowel patterns. In addition, patients with loose stool had a significantly lower QOL than patients with alternating and formed stool consistencies. There are studies revealing that treating diarrhea-associated fecal incontinence with loperamide or fiber supplements is effective in the short-term. This is important considering nearly 75% of patients who reported incontinence to loose/watery stool had not been taking appropriate anti-diarrheal therapy, and approximately 25% were actually taking laxatives. It is therefore plausible that by simply inquiring about stool consistency, specifically diarrhea, and treating appropriately a physician may avoid the need for additional testing and referrals while simultaneously contributing to a patient's quality of life.

Our study had some potential limitations. For one, we did not administer an overall quality of life questionnaire to our patients in an attempt to limit the number of surveys that the patients had to complete. As a result, we are unable to compare the quality of life of our FI patients to other populations such as healthy individuals or those that suffered from urinary incontinence. In addition it could be considered a limitation that we did not use a strict, consistent definition for FI when we approached our patients about this disorder. On the other hand, given the varied levels of education and communication skills of our patients, along with the sensitive nature of this issue, we felt that tailoring the individual interview instead of using a defined wording for FI was most appropriate. Therefore, asking patients directly about FI can lead to increased identification of this debilitating condition. This can be accomplished by prompting physicians to inquire about FI in a targeted review of gastrointestinal symptoms. Furthermore, identifying and treating abnormal stool consistency in patients with FI is a potential strategy to reduce severity and improve quality of life. This intervention is especially important in women, who are more likely to be adversely affected by the severity of their symptoms. Patient and physician education should be stressed to shed light on this difficult and debilitating condition.

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DrexelMed Journal, Volume 6 (1); Spring 2012

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**Table 1: Patient Characteristics** 

Patients, n	58
Female gender, n (%)	42 (74)
Patient Age, years (range)	45 (22-84)
Onset Age, years (range)	49 (17-80)
Education Level, %	
Less than High School	9.3
High School	51.8
Attended College or Greater	38.9
Patients with risk factors for FI, n (%)	72
Hemorrhoids	21 (36.0)
Hysterectomy <sup>a</sup>	12 (29.0)
Irritable bowel syndrome	10 (17.0)
Episiotomies <sup>a</sup>	7 (17.0)
Forceps deliveries®	7 (17.0)
Diabetes Inflammatory bowel disease	9 (15.5)
Anorectal surgery	3 (5.0) 3 (5.0)
Spinal surgery	3 (5.0)
Rectal Prolapse	2 (3.4)
Anal fissures	2 (3.4) 2 (3.4) 2 (3.4)
Scleroderma	2 (3.4)
Radiation therapy (abdomen, pelvis)	2 (3.4)
Rectal/Vaginal surgery <sup>a</sup>	1 (2.4)
Pelvic or Rectal cancer	1 (1.7)
Daily Incontinence, %	
Solid	17
Liquid/Mucous	25
Reason for visit, n (%)	
Upper GI complaints	13 (22.0)
Lower GI complaints	36 (62.0)
Liver	5 (8.6)
Anemia	2 (3.4)
FI	2 (3.4)
Duration, n (%)	
Less than 5 years, n (%)	49 (84.5)
Greater than 5 years, n (%)	9 (15.5)
Use of pads, n (%)	20 (34)
Use of diapers, n (%)	8 (14)

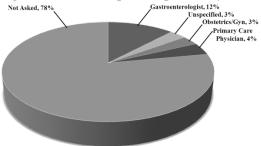
Table 2. Stool consistency, medication use, severity, & QOL

Table 2. Stool consistency, medication use, severity and quality of life scores

Consistency	Medications at the time of visit	Severity score	Quality of Life Score
	None – 11 (48%) Laxative – 6 (26%) Anti-diarrheal – 7 (30%)	29.5	2.26 (1.17 – 3.92)
Formed – 17	None – 9 (53%) Laxative – 6 (35%) Anti-diarrheal – 2 (12%)	23.8	2.94 (1.33 – 3.88)
$Hard-6^{a} \\$	None – 1 (17%) Laxative – 5 (83%) Anti-diarrheal – 1 (17%)	29.7	2.90 (1.48 – 3.96)
Alternating – 12	None – 4 (33%) Laxative – 5 (42%) Anti-diarrheal – 3 (25%)	29.8	2.80 (1.34 – 4.07)

<sup>&</sup>lt;sup>a</sup>One person took both laxatives and anti-diarrheals.

Figure 1: Percent of pts who previously discussed FI w/a physician



 $<sup>^</sup>b$  Significant difference between quality of life (QOL) in loose/watery stool consistency vs. all other groups combined, p=0.006, and separately (vs. formed, p=0.005, vs. alternating, p=0.05). No significant difference vs. hard stool, p=0.06.

## Figure 2. FI QOL scores

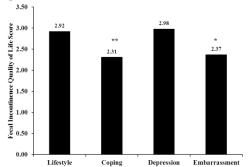


Figure 2. FIQL Scores. \*Significantly lower than the lifestyle and depression scales (p<0.006): nificantly lower than the lifestyle and depression scales (p<0.003

## Figure 3. QOL as function of severity of FI - men

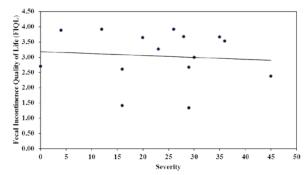


Figure 3. Quality of life as a function of severity of fecal incontinence in men (p=0.75)

Figure 4. QOL as function of severity of FI – women

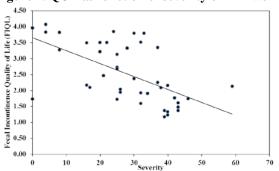


Figure 4. Quality of life as a function of severity of fecal incontinence in women (p<0.001).

Figure 5. QOL as function of severity of FI – both

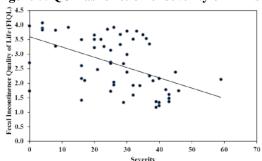


Figure 5. Quality of life as a function of severity of fecal incontinence in men and women

## Article: Metastatic diffuse gastric carcinoma presenting as an acute cholecystitis – report of an unusual case with review of literature

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Diffuse gastric adenocarcinoma or "linitis plastica" is characterized by a thickened and rigid stomach wall due to the diffuse infiltration of signet-ring cells into the lower parts of the mucosa and the upper layers of the submucosa of the stomach. Gastric cancer is known to commonly metastasize to the liver, lungs, and ovaries, but metastasis to the gallbladder is rare. We now report an unusual case of a 79-year-old Caucasian female with diffuse gastric adenocarcinoma that metastasized to the gallbladder, who presented with symptoms mimicking acute cholecystitis. Right upper quadrant ultrasound findings were consistent with cholecystitis, although cholelithiasis was not noted. Cholecystectomy revealed a hardened mass in the gallbladder infundibulum measuring 4.3 cm, and perigastric lymph nodes and gallbladder were removed for pathology studies. Histological exam showed rare signet-ring cells in the perigastric lymph nodes and the submucosa of the gallbladder wall, sparing the serous and mucous layer. Because the mucous and serous layers were spared, this indicated that the signet-ring cells were of metastatic origin. In order to determine whether the adenocarcinoma was of gastric origin, an esophagogastroduodenoscopy with biopsy was performed. Signet-ring cells were also noted in the gastric mucosa, which confirmed that this was a case of diffuse gastric adenocarcinoma with metastasis to the gallbladder. The majority of reported cases in the literature discuss cases in Asia of gastric cancer metastasis to the gallbladder, but this is the first reported case in the United States.

## INTRODUCTION

Diffuse gastric adenocarcinoma, sometimes referred to as "linitis plastica", is cancer of the stomach that is not

associated with Helicobacter pylori. Its name, linitis plastica or "leather-bottle stomach" refers to the thickened and rigid stomach wall due to the diffuse infiltration of signet-ring cells into the lower parts of the mucosa and the upper layers of the submucosa of the stomach. Because the thickening of the stomach, which interferes with its peristalsis, patients may present with symptoms ranging in intensity from a vague sense of postprandial fullness to a severe, steady pain, diarrhea, weight loss, cachexia, and vomiting. It usually presents in an advanced stage and carries a poor prognosis. Diffuse gastric adenocarcinoma most commonly metastasizes to the liver, lungs or ovaries. Only a few studies in Asia report metastasis to the gallbladder, and it has not been previously reported in the United States.

#### CASE REPORT

A 79-year-old Caucasian female presented to the Emergency Department complaining of an acute onset of right upper quadrant pain. The pain was characterized as sharp, radiating to her back, and was associated with nausea. The patient had an episode of the same symptoms one month prior to presentation which resolved on its own. The patient underwent a right upper quadrant ultrasound which noted a thickened gallbladder wall and showed findings consistent with acute cholecystitis. However, no stones were noted on the ultrasound. A laparoscopic cholecystectomy was elected and attempted. Visualization of the gallbladder revealed enlarged lymph nodes of the gallbladder and along the hepatoduodenal ligament. The gallbladder infundibulum also appeared extremely hardened. Due to the inability to visualize the anatomical structures as well as a high suspicion for malignant etiology of the disease, the patient subsequently underwent an open cholecystectomy. The gallbladder infundibulum contained a hard mass, and the patient was found to have extensively enlarged perigastric lymph nodes as well as lymphadenopathy into the omental tissue. Biopsies of the perigastric lymph nodes and gallbladder were performed. CT scan also revealed a thickened stomach wall with air in the midline, and multiple enlarged nodules between the liver and stomach, suspicious of gastric malignancy with metastasis (Fig 1). In order to determine whether the adenocarcinoma was of gastric origin, an esophagogastroduodenoscopy with biopsy was performed.

The gallbladder showed no external masses or focal mucosal lesions, but the wall was thickened over an area of 4.3 cm in greatest dimension. The tissue submitted as perigastric lymph nodes were two portions of adipose tissue with areas of induration. The gastric mucosal biopsies were from an area that was described as diffusely irregular and firm. The gallbladder showed a poorly differentiated adenocarcinoma, including tumor cells with a signet-ring morphology, involving the external aspect of the gallbladder, including the subserosal connective tissue and muscle wall (Fig 2). The mucosa was uninvolved by carcinoma or dysplasia. This distribution of tumor strongly suggested a metastasis. Immunohistochemical stains were performed to further characterize the carcinoma. The positive results for the polyclonal and monoclonal carcinoembryonic antigen stains confirmed the presence of an adenocarcinoma. Thyroid transcription factor-1, a marker for thyroid and lung carcinomas, was negative. Markers for breast origin, including gross cystic disease fluid protein-15, estrogen receptor and progesterone receptor, were all negative. The positive cytokeratin 7 and negative cytokeratin 20 results suggested stomach, pancreas, and biliary tract as the most likely primary sites. The tissue submitted as perigastric lymph node showed mesentery extensively involved by poorly differentiated adenocarcinoma. The gastric body mucosal biopsy showed poorly differentiated adenocarcinoma in the gallbladder.

#### DISCUSSION

Signet-ring cell carcinoma is an extremely uncommon primary carcinoma in the gallbladder with five published case reports. <sup>3-6</sup> Given the rarity of primary signet-ring cell carcinoma of the gallbladder, it is important to determine whether a signet-ring cell carcinoma is primary or metastatic. In our case, adenocarcinoma involved the external aspect of the gallbladder, and the absence of mucosal involvement by carcinoma or dysplasia strongly suggested metastasis. Metastasis to the gallbladder is rare in clinical practice and is usually encountered incidentally at surgery or during autopsies. <sup>7</sup> One study found that it accounts for 4.8% of all pathologically diagnosed gallbladder malignancies, and the most common primary malignancy was from stomach. <sup>8</sup> Signet-ring cell adenocarcinoma of the stomach is a subtype of diffuse gastric adenocarcinoma. It is characterized by round tumor cells that contain abundant intracytoplasmic mucin and nuclei flattened against the periphery of the cells. Signet-ring cell adenocarcinoma is thought to be on the rise in US, even though there is an overall decline in gastric cancer rates. <sup>9</sup> The majority of reported cases of metastatic adenocarcinoma to the gallbladder are those from gastric primaries in Asia (Table 1)<sup>8,10-11</sup> and only one patient was noted to have signet-ring cell pathology. <sup>8</sup> There are no known reports of patients in the United States with signet-ring cell adenocarcinoma of the stomach metastasizing to the gallbladder. In our case, the clinical presentation was unique in that it mimicked a benign condition of acute cholecystitis. While both the clinical and radiological studies pointed to a benign condition, the histological study showing signet-ring

cell pathology encroaching the outer layers of the gallbladder wall uncovered an underlying malignant etiology. Metastasis to the gallbladder is an extremely rare clinical finding since diffuse gastric cancer most commonly metastasizes to the liver, lungs or ovaries. The clinical presentation of a secondary tumor can often mimic acute or chronic inflammation of the gallbladder. Therefore, a patient with metastatic gallbladder tumor can present with cholecystitis, as seen in our case. In addition, ultrasound studies of gallbladder wall thickening without evidence of cholelithiasis may suggest metastatic gallbladder tumor since primary gallbladder carcinoma appears to be associated with gallstones in approximately 80% of primary gallbladder malignancies. This case of metastatic diffuse signet-ring carcinoma to the gallbladder is a unique one in that the patient was an elderly Caucasian female who presented with symptoms mimicking acute cholecystitis. To our knowledge, this is the first reported case in the United States, and it provides additional clinicopathological insight into the sites of metastasis of diffuse gastric adenocarcinoma.

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Figure 1. CT showing thickened stomach wall and multiple enlarged nodules



Figure 2. Signet-ring cell carcinoma in the gallbladder subserosa

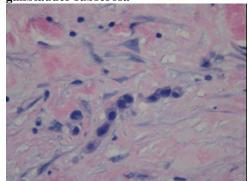


Figure 3. Poorly differentiated adenocarcinoma in the gastric mucosa

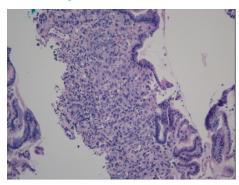


Table 1. Reported cases of gastric carcinoma metastasis to gallbladder

Reference	Country of study	Type of gastric cancer	Presentation	Diagnosis
Yoon et al <sup>8</sup>	South Korea	Adenocarcinoma, signet ring cell carcinoma	Cholecystitis, abdominal pain, jaundice, weight loss	Carcinoma confirmed by pathology
Masamune et al <sup>10</sup>	Japan	Moderately differentiated adenocarcinoma	RUQ pain, fever, jaundice	RUQ US showed thickening of gallbladder, dilation of intrahepatic ducts and common bile ducts, smooth focal elevation of gallbladder
				Carcinoma confirmed by pathology
Kanno et al <sup>11</sup>	Japan	Poorly differentiated adenocarcinoma	Epigastric discomfort, abdominal full feeling	RUQ US showed multiple echogenic lesions of the gallbladder
				Carcinoma confirmed by pathology

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## Article: MR Imaging Findings of Occult Ipsilateral and Contralateral Breast Cancer in Women with Recently Diagnosed Breast Cancer in a Community Setting

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#### INTRODUCTION

Breast MRI is a valuable tool for pre-operative evaluation of patients with newly diagnosed breast cancer. It can provide a more accurate assessment of the extent of disease than the conventional modalities such as mammography or ultrasound. The reported sensitivity of MR imaging for the visualization of invasive cancer has approached 100%. While the effect on long-term survival has not yet been determined and the false positive rate of MRI can be high and variable, <sup>2,3</sup> recent studies suggest the overall discriminating ability of MRI is significantly better than conventional imaging. <sup>4,5</sup> In this report, we present a retrospective review of our experience with MR-guided detection of mammographically and sonographically occult lesions in women who underwent pre-operative MR imaging for recently diagnosed breast cancer.

## MATERIALS & METHODS

Retrospective review was performed on records of patients with newly diagnosed breast cancer who underwent preoperative staging breast MRI from January 2007 to June 2010. These examinations were performed at either Monmouth Medical Center or at the hospital's associated outpatient imaging center, Shrewsbury Diagnostic Imaging. All imaging performed at Monmouth Medical Center was on a 1.5 T MRI system (GE Signa EXCITE);a dedicated 8-channel breast coil was used with bilateral acquisition. Vibrant software was used which allowed to acquire thinner slices with more detail in regards to breast imaging. The post contrast protocols consisted of four contrast-enhanced acquisitions. Gadolinium was administered with a mechanical injector at a dose of 0.2 mL/kg unless MultiHance was used where we then used .07mL/kg. The dynamic acquisitions were under 2 minutes. Slice thickness was less than 3 mm. The imaging performed at Shrewsbury Diagnostic Imaging was done using a 1.5 T Siemens Espree Ultra Short Bore Magnet (more commonly known as a Hi-field Open). A 7-channel dedicated breast coil was used for bilateral axial acquisitions. Post contrast imaging consisted of three one-minute acquisitions unilaterally in the sagittal plane of both left and right breasts run consecutively, using 3DFlash with Fat Sat. Slice thickness was 3 mm. Post-contrast imaging began approximately 23 seconds after injection and a power injector was used for all cases. For the purpose of this study, results of the diagnostic MR images were reviewed. Those reports that suggested a lesion or multiple lesions separate from the index lesion as well as those that had lesions more extensive than initially suspected on mammography or ultrasound were identified. Pathology of these additional lesions was reviewed. Any questionable lesions that did not clearly demonstrate a separate additional lesion or a more extensive finding on pathology were not included as the final positive findings. Only lesions examined by pathologists at Monmouth Medical Center were included in our analysis.

#### RESULTS

Between January of 2007 and June of 2010, 166 patients underwent breast MRI at Monmouth Medical Center or Shrewsbury Diagnostic Imaging following a new diagnosis of breast cancer. MR imaging detected additional

suspicious previously unsuspected lesions in 76 women. These women either underwent additional biopsy of these lesions or went onto surgical excision. Correlation with the pathology reports demonstrated 16 women (9.6%) to have clinically occult cancer in the ipsilateral breast and two women (1.2%) to have clinically occult cancer in the ipsilateral as well as the contralateral breast. Of these occult cancers, invasive ductal carcinoma was present in 9 women (5.4%), DCIS was present in 10 women (6.0%), and invasive lobular carcinoma was present in two women (1.2%). High risk lesions such as atypical lobular hyperplasia were present in two women (1.2%) and LCIS was present in two women (1.2%). The positive predicted value of an ipsilateral or a contralateral occult cancer detected on MR imaging for this study was 21.1%.

## DISCUSSION

Despite high sensitivity, pre-operative breast MRI has not been without controversy due to a variety of factors including high false positive results, variable practice standards and interpretation guidelines.<sup>4</sup> In this study, we found that 10.2% (17 of 166) of patients with recently diagnosed breast cancer had mammographically and sonographically occult additional cancer on either ipsilateral or contralateral breast. Additional ipsilateral breast cancer was found in 9.6% of women in our study group and 1.2% were found to have a synchronous contralateral cancer. 1.2% of women had high-risk markers only. Rates of detection of additional ipsilateral cancer in other reports have ranged between 6% and 27% and of contralateral cancer have ranged between 3% and 9%. 6-9 Our experience with breast MRI in the community setting reveals a diagnostic frequency in identifying additional ipsilateral occult lesions consistent with that of published reports. The contralateral occult cancer detection rate however is lower. The reason for this decreased detection rate for contralateral cancer on MRI is likely because at our institution, until March of 2010, bilateral global breast ultrasound was routinely done in patients with newly diagnosed breast cancer before the pre-operative MRI. There was a low threshold for a biopsy on those who were found to have suspicious additional lesions on ultrasound. Pre-operative breast MRI was done after the bilateral ultrasound and subsequent biopsies, several of which yielded additional cancer. Therefore, this likely has artificially lowered our detection rate of occult cancer with MRI. What this means, however, is that despite doing a bilateral global ultrasound prior to pre-operative breast MRI in these patients, MRI was able to detect additional cancer in 9.6% in the ipsilateral breast and 1.2% in the contralateral breast. Since March of 2010, we have abandoned the practice of pre MRI global ultrasound and began performing breast MRI following the diagnosis of breast cancer. We conclude that breast MRI is an invaluable tool in detecting mammographically, sonographically and clinically occult additional breast cancer when used in a pre-operative evaluation. Our study has shown that community hospitals are able to reproduce cancer detection rate on MRI that is comparable to that found in larger university hospitals. Additional work and continued long-term follow-up is necessary to determine the ongoing role of breast MR imaging in the preoperative assessment of women with proven breast cancer.

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## **Article: Rare Case of Stercoral Perforation Requiring Subtotal Colectomy**

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## INTRODUCTION

Stercoral perforation is recognized as a rare cause of colonic perforation most commonly seen in elderly, debilitated, and/or institutionalized patients on multiple medications. Drugs such as narcotics, anticholinergics, antacids, and NSAIDs have been implicated as causes of fecal impaction resulting in stercoral perforation. We report a case of stercoral perforation in a previous heroin user on methadone maintenance. To our knowledge, this is the first reported case of subtotal colectomy performed for drug-induced stercoral perforation.

## CASE REPORT

A 41 year-old female presented with a 2 day history of abdominal distention and generalized pain, worse with motion, accompanied by nausea, anorexia, non-bilious vomiting, and 1 week history of obstipation. Past medical history was significant for type 2 diabetes and current methadone maintenance therapy at 180mg. On physical examination, she was afebrile, tachycardic, and tachypneic. Her abdomen was diffusely tender with maximal tenderness on her left side with rebound, guarding, and hypoactive bowel sounds. A rectal exam was unrevealing for abnormality. Laboratory examination showed WBC 22,200/ml with bandemia, glucose 267mg/dL, and elevated lactic acid 4.7mmol/L. CT scan revealed perforation with moderate free air and thickening of peritoneal surfaces. An exploratory laparotomy showed a large perforation in the sigmoid colon measuring 8-10cm with proximally dilated colon studded with fecalomas. The widest point, measuring 19cm in circumference, had a thinned out bowel wall. Attempted salvage by left hemicolectomy failed as further exploration revealed extensive serosal hemorrhage of the transverse colon, prompting our decision to pursue a subtotal colectomy with end ileostomy. Microscopic examination showed multiple ischemic mucosal ulcers consistent with sterocoral perforation with transmural necrosis and acute necroinflammatory changes in the margins. After recovering from complications of sepsis and acute respiratory distress syndrome, the patient was discharged home.

## DISCUSSION

Stercoral perforation has been defined as perforation of the large bowel due to pressure necrosis from fecal masses.<sup>3</sup> Multiple cases of colonic perforation have been reported since its first description in 1894. A prospective study revealed stercoral perforation accounts for 1.2% of all emergency colorectal surgeries and 3.2% of all colonic perforations, suggesting its true incidence may be higher than previously thought.<sup>4</sup> Most patients report a history of chronic constipation as well as straining bowels as a precipitant event. There have been case reports of stercoral perforation in association with multiple drugs,<sup>1,2</sup> which are thought to affect colonic motility rather than the colonic wall itself.

Patients typically present with a picture of peritonitis, and early surgical intervention is critical to a good outcome, with the procedure of choice with the lowest mortality (23%) being resection of the diseased segment of colon, end colostomy, and Hartmann's closure of the rectum.<sup>5</sup> Even if stercoral ulcers are not visible, the substantially dilated colon, especially with multiple fecalomas, should be removed up to a subtotal colectomy to prevent a second perforation at a later time.<sup>4</sup>

## CONCLUSION

Stercoral perforation of the colon is a rare but serious complication of chronic constipation, and subtotal colectomy can be an option with good outcomes.

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Figure 1. Resected bowel and CT showing free air and colon perf/thickening







Fig a: Resected specimen Extensive purulent material with fibrinous exudates on large bowel with multiple large fecaliths of varying size in the peritoneal cavity. Serosal hemorrhages(arrow) Fig b & c: CT scan Abdomen Pelvis with free air (star) and colon perforation and thickening (thick arrow)

## Medical Essay: Quick Response (QR) codes for poster presentations

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We have been working with endoscopic underwater techniques for some time and been sharing our findings primarily through poster presentations. During these presentations, something seemed to be lacking, the still images did not serve justice to the nature of what we were trying to demonstrate. We needed to show moving videos, but the motionless images on the posters were simply unable to provide us with that platform. We would like to share an idea that one of us (Dr. A. Prushani) recently proposed and executed. His notion applied new technology that makes the presentation of multimedia possible in poster format. The method involves the use of Quick Response (QR) codes, which is a publicly available technology developed in the early 1990s. It is a 2-Dimensional bar code that can carry several thousands of alphanumerical data. A QR code scanner application installed on a smartphone or a tablet PC is necessary to interpret the information stored in QR code format. Recently some have proposed using this technology in medical handouts, newsletters, scientific articles and even business cards. <sup>1,2</sup> Here is how it is done: Once a video is ready, upload it onto any website that would host and play videos such as http://youtube.com/. A title and keywords can be added for future searching purposes. After the video is uploaded, a permanent link is provided for that video. Copy that link into your clipboard and paste it onto a publicly available QR code generator. For our project, we used http://qrcode.kaywa.com/. Next, one can save the code in .JPG format to his or her computer (from there it can be added to any poster as an image). On a smartphone or tablet, find a freely available QR reader. We used 'Neoreader' for iPhone, however, there are many more available on the Android market and Blackberry App World as well as the Apple App Store. Everything is now set. When scanned, the QR code will redirect the smartphone to the prerecorded multimedia file available on the Internet. We have foreseen multiple other uses for this technology in the field of medicine. One could apply QR codes to point at certain journal articles in reference lists; this makes it much easier and more fun to check references. They can also load high resolution images, auscultatory findings and sound files, or even the full electronic copy of a poster. This is a simple, economic, and easy way of including multimedia on poster presentations, which will definitely help scholars better express their findings during conferences and meetings. We presented this method for the very first time at the 2011 American College of Gastroenterology Meeting in Washington D.C. with great success and appreciation.

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Figures - Scan this Image using the instructions in this essay!





## Poem: My Answer

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During our community medicine rotation, we identify and assess marginalized patient populations, their health needs, and the resources available to help them. I worked with several homeless women and children struggling to survive and desperately seeking refuge, peace, and security. This poem is a story about a woman I met during my rotation; her story is quite familiar to those of us that train and practice medicine in the inner-city.

## My Answer

I don't remember my mother... I never met my father... Raised in foster care, Life was quite unfair

I felt worthless
Look at me! I'm a mess
I was first raped at thirteen
Bruised and beaten, people can be so mean...

In fact, I have never known love I've known anger, sadness and pain, but not love. What a tragedy —my life You can't imagine all my strife

I just needed to escape the pain
I just couldn't take the constant rain.
Drugs were my answer
Addicted—it's a disease you know... like Cancer.

I long for my next high Without it I feel like I might DIE! How else can I forget my pain? It's my umbrella in the rain... How else can I forget my pain?

## Review Article: A Case of Idiopathic Chronic Colonic Pseudo-Obstruction

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#### CASE REPORT

A relatively healthy 81 year-old male underwent a right total knee arthroplasty with the resulting complication of a paralytic ileus, requiring a consultation with general surgery. Abdominal films demonstrated dilation of both small and large bowel; the patient had associated nausea and abdominal pain. The patient had a medical history of hypertension, hypercholesterolemia, and coronary artery disease. A coronary artery bypass had been performed more than five years earlier. He had no allergies and did not report consuming alcohol or tobacco.

The patient's ileus at that time was treated conservatively with an NG tube and erythromycin IV, to which the patient responded positively. As is standard after an orthopedic surgery at our institution, he was discharged to an inpatient physical therapy rehabilitation center. The patient later required a skilled nursing facility for further physical therapy. As the patient did well in the rehabilitation facility and his ileus had clinically resolved, he was given a regular diet and sent to the SNF.

The patient returned to the hospital six months later and presented to the gastroenterology team with the complaint of chronic diarrhea that developed about the time of his dismissal from the hospital. Stool studies were negative and the patient's symptoms failed to alleviate after a course of metronidazole. The patient underwent a colonoscopy to determine the etiology of his loose stools. The colonoscopy report noted diverticula with no evidence of inflammation and no other pathology of the bowel. Random biopsies were taken of the entirety of the large bowel which came back grossly normal with the exception of some occasional lymphocytes in the lamina propria. The patient was eventually discharged home on asachol and Questran, though the final pathology came back negative for inflammatory or lymphocytic colitis.

Nine months later the patient again returned to the Emergency Department with the same complaints of chronic diarrhea and abdominal distention, with an acute exacerbation in symptoms over the previous two weeks. The patient complained of worsening abdominal distention, nausea, vomiting and of diarrhea occurring around three times daily. The patient was also having shortness of breath, dizziness, headache, and profuse sweating along with his abdominal distention. Furthermore, the patient had abdominal pain that was constant, dull and aching, mostly noted in his epigastric area and right upper quadrant. KUB revealed grossly dilated transverse colon. A CT scan without contrast revealed distention of the ascending and transverse large bowel with a "misty" mesentery, yet no identified colitis, obstruction or stricture. Ogilvie's syndrome was suspected. A rectal tube was attempted, but failed to relieve the abdominal distention. Gastroenterology took the patient for an emergent colonoscopy to perform decompression of the bowel. The colonoscopy revealed that the patient once again had diverticula that did not look inflamed, and no stricture was found. A follow-up abdominal x-ray showed alleviation of the colonic dilatation, and the patient's symptoms improved. The patient was slowly advanced in diet on the hospital floor and discharged several days later with a flat abdomen. The final discharge diagnosis was an episode of Ogilvie's syndrome.

Just over a year later, the patient again presented with abdominal pain, but this time in the left lower abdomen. The patient still complained of chronic abdominal distention and diarrhea. The patient had a CT which again revealed dilated ascending and transverse colon but now with a sigmoid diverticulitis. After being admitted and treated for acute diverticulitis the patient's left lower quadrant pain eventually resolved, but his abdominal distention remained the same. Surgery was consulted regarding the patient's chronic abdominal distention. Due to the chronic nature of the patient's abdominal distention and the similarly chronic symptomatology that resulted from it, the decision was made to take the patient to surgery. In the operating room, the patient's ascending, transverse, and a portion of the descending colon were massively dilated, while the entirety of the small bowel was found to be of normal caliber and with very good observed motility. The distal descending colon and sigmoid were also noted to be of normal caliber and function. There seemed to be a single point of transition between the distended and normal bowel calibers at the proximal end of the descending colon that was marked with overlying indurated mesenteric fat. It was decided intraoperatively to resect the pathologic portion of large bowel only, from the cecum to the proximal descending colon. The entirety of the resected bowel was sent to pathology with the expectation that a tumor or diverticular stricture would be found at the transition point. The patient was sent to the ICU where he recovered rapidly and was subsequently transferred to the regular floors. The patient's abdominal distention had completely

resolved after the procedure and normal bowel function soon returned. The patient recovered well and was eventually discharged to a nursing home to complete his physical rehabilitation after surgery.

The pathology report noted that the largest diameter of the colon was 27.5cm at the cecum. An area of indurated mesentery at the distal end of the specimen was found that was consistent with the transition point noted in the operating room. However, no obvious diverticular stricture or mass was found at that point, and the pathologists noted that ganglion cells were present throughout the resected specimen. The patient was then diagnosed with an idiopathic chronic colonic pseudo-obstruction.

DISCUSSION - Ogilvie's syndrome versus Chronic Idiopathic Colonic Pseudo-obstruction.

The infamous Ogilvie's syndrome, described by Sir Heneage Ogilvie in 1948, is a disease that is characterized by acute colonic distention without a mechanical obstruction. It is often associated with elderly post-operative populations, especially in those patients in their sixth or seventh decade of life undergoing cardiac or orthopedic procedures. Diagnostic criteria for Ogilvie's syndrome include acute distention of the colon, a cecal diameter greater than 9 cm, and the absence of any mechanical obstruction. The exact etiology of the disease is unknown, but it has been theorized to be a disregulation in the autonomic nervous system of the bowel. This syndrome has been associated with multiple disease states, including CHF, Parkinson's, electrolyte imbalances and cancers. Medications such as narcotics and antidepressives have been associated with the syndrome as well. The standard treatment is to make the patient NPO and to decompress both the stomach with a nasogastric tube and, if necessary, the colon with a rectal tube. In refractory cases, prokinetics and cholinesterase inhibitors such as neostigmine have been successfully used to treat colonic distention, though not without the risk of serious side effects such as bradycardia. Decompression via colonoscopy has become an important treatment and diagnostic of Ogilvie's, but when all other modalities fail, surgery becomes the only hope for cure. Untreated Ogilvie's is very morbid and can result in cecal perforation, which could cause sepsis and, very possibly, the death of the patient.

Chronic pseudo-obstruction is less well defined than Ogilvie's. It is a chronic symptomatic dilatation of the bowel, involving either the small or large bowel, or both, and has multiple proposed etiologies. Symptoms of chronic colonic pseudo-obstruction include nausea, vomiting, abdominal pain, constipation, diarrhea, abdominal distention, feeding intolerance, malnutrition, and failure to thrive. Chronic pseudo-obstructive syndromes are known to be highly morbid diseases, as can be seen from the large list of symptoms.<sup>4</sup> There are two broad categorizations of chronic intestinal pseudo-obstruction; primary, or "idiopathic", disease of unknown etiology, and secondary disease, related to a known cause. Breaking down secondary chronic intestinal pseudo-obstruction further, we find three proposed broad etiologic categories: neuropathic, myopathic and mesenchymopathic. <sup>5</sup> Neuropathic causation includes inflammatory or degenerative processes that destroy enteric ganglia. Myopathic causes of chronic pseudoobstruction are characterized by muscle fibrosis and destruction. Mesenchymopathies are characterized by destruction of the pacemaker of the GI tract, which are the interstitial cells of Cajal.<sup>4</sup> The diagnosis of a chronic pseudo-obstructive disease requires that mechanical obstruction be ruled out, which can be done with a CT with oral contrast, a barium enema, and/or endoscopy. Motility and manometry studies can be helpful in eliciting a pathology resulting from a known disease, such as gastroparesis in diabetics. Histology can help determine causation of symptoms as well, the most obvious example being that of Hirchsprung's disease, in which aganglionosis of the colon arrests colonic movement and leads to megacolon with all of its inherent morbidities. Finally, the duration of symptoms helps to reveal whether or not the disease is truly an acute process or a chronic pathological state

In an attempt to elucidate the nature of the disease, there have been several studies attempting to illuminate the causes of idiopathic chronic pseudo-obstruction. Guarino et al. performed an experiment in which the isolated circular muscle cells from normal colons were compared to those of patients with chronic intestinal pseudo-obstruction in regards to contractility when exposed to acetylcholine. The experiment found that the smooth muscle cells in affected patients contracted much less than in their healthy counterparts. Struijs et al. discovered an absence of interstitial cells of Cajal in a case of a child with chronic pseudo-obstruction, and hypothesized that this lack could have been responsible for the condition. More relevant to this specific case study, Loening-Baucke et al. found decreased bowel muscle contraction, frequency, and amplitude and increased rectal wall elasticity in patients with idiopathic chronic colonic pseudo-obstruction as measured by motility studies. Also noted in this study was that the anal pull-through pressures were not significantly different between experimental and control groups. Interestingly, it was noted in the research that the only relief for five of seven patients with idiopathic pseudo-obstruction came via surgical intervention.

This begs the question of what treatments are effective for chronic idiopathic colonic pseudo-obstruction. The initial treatment proposed by O'Dea et al in a 2008 study was the same as for an acute colonic pseudo-obstruction: NPO, decompression via insertion of rectal and/or NG tubes, and nutritional support if needed. In addition, they proposed the use of a cholinesterase as medical therapy. Their proposal was that the basic mechanism for pseudo-obstruction was an imbalance between the excitatory (parasympathetic system, using acetylcholine as a neurotransmitter) and inhibitory (sympathetic system, using nitric oxide as a neurotransmitter) auto regulation of the bowel. An overabundance of sympathetic activity or a lack of parasympathetic activity was thought to cause the clinical symptoms of disease. The authors hypothesized that the success of neostigmine in treating Ogilvie's was due to the fact that it supplied parasympathetic activation of the colon, and that such stimulation could alleviate chronic intestinal obstruction as well as colonic pseudo-obstruction. Their study attempted to demonstrate that a cholinesterase inhibitor would improve acute recurrent and chronic idiopathic colonic pseudo-obstruction as well as chronic slow-transit constipation. The results of their study showed that pyridostigmine administration improved the symptomatology of pseudo-obstruction, but had no discernible effect on slow-transit constipation. In fact, five of the six members of the slow transit constipation group needed surgical intervention while only 2 of the 7 members of the chronic pseudo-obstruction group went on to need surgery. The authors concluded that it was likely that a lack of parasympathetic activity was the cause of pseudo-obstruction and that therefore a longer acting cholinesterase could be used as a first line medical management for this disease.

For those that fail medical therapy, Thompson et al. have demonstrated an uncomplicated procedure that may spare the patient a major surgery and still effectively treat the symptoms of disease. In a 2004 case study, Thompson et al. demonstrated the safety and effectiveness of placing a percutaneous endoscopic colostomy in three patients with chronic intestinal pseudo-obstruction. These "colostomies" were, in fact, standard 16-french gastrostomy tubes that were placed in a similar fashion to PEG tubes and then converted to Mic-Key tubes six weeks later. The tubes were essentially used for venting purposes, and the authors demonstrated symptomatic improvement and avoidance of major surgery for all three recipients.<sup>8</sup>

Isolated chronic idiopathic colonic pseudo-obstruction is a rare disease, as can be attested to with the relative paucity of research and studies in current literature. A well-defined separation of chronic idiopathic colonic pseudo-obstruction from a recurrent Ogilvie's was not forthcoming in the literature. As Ogilvie's is defined as an acute process, it must be considered that a certain length of time or set number of recurrences should change the diagnosis from Ogilvie's into that of a chronic idiopathic colonic pseudo-obstruction. It is interesting to note a study by O'Dea et al. in which a well-known medical therapy for Ogilvie's was shown to be effective in chronic idiopathic colonic pseudo-obstruction, as this suggests a similar pathophysiology for each disease. It would be interesting to repeat the Loening-Baucke et al. experiment while comparing muscular contraction between patients with Ogilvie's syndrome and those with chronic idiopathic colonic pseudo-obstruction. It may be that instead of two separate disease states, these are simply two different spectra of the same illness. However, more research will need to be performed comparing the two diseases before such propositions be entertained.

Until the pathologic mechanisms of both diseases are found and a medical therapy is proven to work for chronic idiopathic colonic pseudo-obstruction, the disease should be treated conservatively. NPO, rectal tubes, NG tubes, and supportive measures should remain the initial treatment while simultaneous search for an underlying cause is sought. A cholinesterase may be tried if there is no response to conservative therapy, but if this also fails a consultation with a surgeon would be prudent. Surgical correction is a definitive treatment to pseudo-obstruction and should be considered in any recalcitrant Ogilvie's as well as any chronic colonic pseudo-obstruction, especially if symptoms become life threatening or are intolerable. More study is needed in order to more clearly define the disease state of chronic idiopathic colonic pseudo-obstruction, and thus aide the physician in treating this rare yet morbid disease.

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Figure 1. KUB-paralytic ileus



Figure 2. KUB on readmission



Figure 3. KUB showing dilation



Figure 4. CT revealing dilated colon

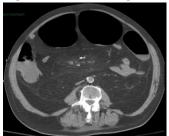


Figure 5. Preoperative patient



Figure 6. Intraoperative view



Figure 7. Gross Pathology



# Review Article: Congenital Absence of Inferior Vena Cava as a Rare Cause of Recurrent Deep Vein **Thrombosis**

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# **ABSTRACT**

Absence of a segment of inferior vena cava (IVC) is a very rare congenital abnormality which is increasingly recognized to be associated with recurrent deep vein thrombosis (DVT) in young patients. We are reporting on a case of a 28 year-old male who presented with recurrent episodes of DVTs and failed conventional treatment with anticoagulation. The decision for an IVC filter placement was made. However, absence of the infrarenal segment of

the IVC was recognized on venography. We will discuss the clinical recognition of this rare entity and provide a review of cases described in the literature.

### INTRODUCTION

IVC absence is a rare genetic abnormality believed to effect between 0.3-0.6% of the general population. While this condition can remain asymptomatic, it can manifest as DVT in the lower extremities due to increased venous pressure and stasis created by less efficient venous return through retroperitoneal collaterals. In young adults under 30 years of age who experienced DVT, IVC congenital absence syndrome is believed to be an under-recognized predisposing factor present in as many as 5% of patients. 

In young adults under predisposing factor present in as many as 5% of patients.

#### CASE REPORT

Our patient is a 28 year-old male who was working at Wegman's when he noticed that he was beginning to develop right calf discomfort, worse on ambulation. This patient has a history of multiple DVTs in the past for which he has been treated with anticoagulation. Due to the previous failed attempts to prevent clot formation in this patient with anticoagulation therapy, the decision was made to place an IVC filter. During the operation, no IVC could be located for filter placement and the procedure was abandoned. CT images showed a very prominent azygous vein identified extending from the level of its transverse arch through the diaphragmatic hiatus where it was fed by collaterals. Dilated common iliac veins were evident bilaterally right greater than left, with cephalic continuation via dilated collateral vessels. Iliac veins were not sufficiently well opacified to assess for internal defects. The IVC reconstituted at the level of hepatic veins. Patient was noted to have a 1cm hypodensity involving the lateral midpole right kidney and a urinary bladder that appeared thickened, possibly reflecting cystitis or hypertrophy. The patient also exhibited a left kidney asymmetrically enlarged compared with a somewhat atrophic right kidney, which is a commonly associated condition with IVC absence syndrome. This might lend evidence to the potential etiology of this condition.<sup>6</sup>

### DISCUSSION

IVC absence syndrome is a rare but increasingly recognized cause of DVT in younger adults. IVC absence syndrome is believed to cause blood clots by creating a less efficient return of blood flow to the heart. An inadequate blood return through collaterals may increase the venous blood pressure and stasis in the veins of the legs, thus facilitating DVT. <sup>4,5</sup> IVC absence has been described in coincidence with clotting defects, although in most of the cases including our patients, screening for thrombophilia revealed no alteration. The etiology of IVC absence syndrome is unknown but is believed to either result from a failure of the development of the right supracardinal vein or intrauterine/perinatal thrombosis. <sup>4</sup> It has been noted in numerous patients as well as our own that the right kidney often exhibits degeneration in addition to IVC absence. In an article published by Gayer et al., all 9 patients reviewed showed this association. <sup>2</sup> It has been hypothesized that the correlation between these two conditions might lend credence to the early thrombosis theory as both conditions could result from a clot within the IVC adjacent to or with migration from the left renal vein. <sup>3</sup> This theory is also supported by the case of a 12 year-old girl described by Ramanathan et al. who was noted to have acquired this condition following a DVT within 48 hours of delivery. <sup>6</sup> IVC absence can remain asymptomatic throughout the life of the patient but should be considered as a possible cause of DVT in a younger patient who has no underlying malignancy or hypercoagulable condition.

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Figure 1. Venogram of suprahepatic IVC dissipating into collaterals





Figure 3. Dilation of Iliac vessels from venous hypertension



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Review Article: Non-acute Compartment Syndrome of the Lower Extremity as a Presentation of Cutaneous T-Cell Lymphoma: A Review of Literature and Case Report

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#### ARSTRACT

In all facets of orthopaedics, compartment syndrome can be met due to a myriad of pathologies. A patient presented with symptoms over a subacute time course that raised suspicion for compartment syndrome in his lower extremity. Non-Hodgkin's lymphoma, specifically Mycosis fungoides, was ultimately discovered as the malady resulting in the disorder. When treating extremity pain, weakness, and numbness, it is important to maintain a wide differential diagnosis including that of compartment syndrome.

### INTRODUCTION

Compartment syndrome is a disorder commonly encountered in the hospital setting, particularly related to trauma. Fascial compartments provide an anatomical constraint that resist intracompartmental pressures. As increasing tissue pressures near the diastolic blood pressure, microvascular flow is impaired and irreversible cellular damage ensues. The most common causes of compartment syndrome are acute traumas including fractures, crush injuries, and circumferential burns. There are numerous other causes described throughout the literature. However, only one case found through a literature search described a compartment syndrome secondary to Non-Hodgkin's lymphoma. The lymphoma in this case was suggestive of Waldenstrom's macroglobulinemia, a plasma cell dyscrasia. Mycosis fungoides, another rare Non-Hodgkin's lymphoma, is a cutaneous T-cell lymphoma most often presenting with patch and plaque lesions on non-sun-exposed areas of the skin. We report a case of non-acute compartment syndrome in a patient with a known history of active Mycosis fungoides.

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### CASE REPORT

A 66 year-old man presented to our Emergency Department after increasing pain and swelling in his right leg over the course of the past month. He denied any trauma or falls. The swelling began in his right foot, accompanied by minimal to moderate pain and nonspecific numbness throughout the foot. Over the course of the month, the pain and swelling ascended to include his entire right leg circumferentially. Initially the patient's primary care physician eliminated the diagnosis of deep vein thrombosis via lab tests and ultrasound. Multiple lower extremity ultrasounds over the month, including in the Emergency Room, were negative for an occlusive DVT. However, they did show an infiltrating mass in his posterior right calf. Neither orthopaedic surgery nor vascular surgery had previously been consulted for the patient's leg pain. On this visit, another ultrasound revealed a non-occlusive thrombus along with an extraluminal mass impeding blood flow to the leg. Of note, the patient had recently been diagnosed with the Tcell lymphoma, mycosis fungoides, after a colonoscopy with biopsy was performed. The treatment regimen of UV phototherapy proposed by dermatology was being held until further workup of his leg pain was performed. At presentation, the pain in his leg had progressed over the previous week to the point where it had become intolerable and impelled the patient to be seen in the ED. Upon physical exam, the patient's entire right leg below the knee was swollen and discolored when compared to his left leg. It was tender to palpation and noticeably firm and cool. His skin integrity was intact with no visible defects. He had no motor function and had been non-ambulatory for 3 weeks. Pulses were palpable in the right femoral and popliteal arteries, but the dorsalis pedis and posterior tibial arteries were not palpable. Using a Doppler, a signal was unable to be obtained in the area of the dorsalis pedis, but a weak signal with poor wave form was obtained from the posterior tibial artery. Ankle-Brachial Index (ABI) for the left leg was equal to 1, and the ABI for the right leg was 0.2. Firm, non-tender lymph nodes were palpable in the right inguinal region. Compartment pressures were not measured at that time because apparent compartment syndrome was not deemed to be acute based on the patient's history. Nineteen days after being admitted from the ED, the patient was taken to the operating room to undergo an above-the-knee amputation of his affected leg, which was deemed non-viable. In the OR, a Stryker (Mahwah, NJ) needle was used to check compartment pressures twice in each compartment of his right leg prior to incision. Pressures were the following: anterior compartment 30, 31; lateral compartment 27, 26; posterior deep compartment 12, 12; posterior superficial compartment 11, 10. At the time of measurement the blood pressure ranged from 110/60 to 114/66. Upon incision, all muscles of the four compartments of the leg were dusky and non-contractile. An encapsulated tumor was discovered in the anterior compartment which tracked from the mid-calf through to the posterior thigh and into the pelvis. Pathology revealed Mycosis fungoides.

# DISCUSSION

A compartment syndrome secondary to an infiltrating malignancy has been well documented. <sup>1,6</sup> Fewer reports exist of the compartment syndrome being the result of a Non-Hodgkin's lymphoma. <sup>9</sup> In one case of note the compartment syndrome was an acute process and the diagnosis of the lymphoma was made from specimen obtained from the fasciotomy to relieve the increased intra-compartmental pressures. <sup>6</sup> This is in contrast to the diagnosis of Mycosis fungoides made in this instance prior to the onset of compartment syndrome-like features. In theory, a compartment syndrome could occur from a slow growing tumor causing a mass effect or an accumulation of fluid over time. The authors are unaware of any prior instances in the literature of lymphomas causing a compartment syndrome-like effect over a non-acute course of time. However, the end result is similar if one overlooks the time course: muscle and nervous tissue necrosis secondary to impaired vascular perfusion. <sup>4,5</sup> The authors are reluctant to classify this case as a chronic or subacute compartment syndrome because these terms are often applied to compartment syndromes secondary to an exertional etiology. <sup>3,10</sup> In these instances, the term chronic or subacute is used to signify the recurring nature of the disorder.

#### CONCLUSION

In this case the compartment syndrome most likely developed in concordance with the slow-growing tumor in the compartment of the leg. As it grew it likely occluded blood vessels and caused a mass effect within the enclosed fibro-osseous compartment, as occurs in other forms of compartment syndrome. Recognition of this non-acute process should at least be in the back of a physician's mind when evaluating a patient with any malignancy with the ability to infiltrate any commonly-affected myofascial compartment. A slow onset should not preclude the need for both non-surgical and surgical management to avoid complications associated with compartment syndrome.

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Figure 1. Right leg ultrasound - popliteal fossa – mass mainly posterior and slightly circumferential

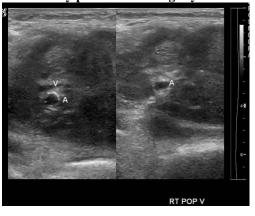
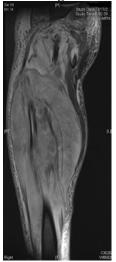


Figure 2. MRI right thigh. Mass measures 57.9mm x 166.4mm posteriorly



Figure 3. MRI right calf large heterogeneous mass with a large of amount of tissue swelling and edema distally.



# Review Article: Subacute Stent Thrombosis – A Case Report and Review of the Literature

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#### CASE REPORT

A 51 year-old female patient with known history of hypertension, diabetes mellitus, old CVA, and coronary artery disease with previous stent in mid-left anterior descending (LAD) artery presented with an acute coronary syndrome in the setting of diabetic ketoacidosis. After initial management with hydration, insulin and electrolyte correction, she underwent cardiac catheterization which revealed a critical LAD in-stent stenosis. She underwent successful percutaneous coronary angioplasty and stent deployment in the proximal LAD with a bare metal stent (Vision 3.0x 15mm) with excellent results (Figure 1). On day 4, while she was still in the hospital on clopidogrel 75mg daily and aspirin 81mg daily, she developed acute onset of chest pain with diaphoresis. An EKG revealed new ST-segment elevation in anterior precordial leads with marked elevation of cardiac markers (troponin 100). Urgent cardiac catheterization was performed and coronary angiography demonstrated total thrombotic occlusion of the LAD (Figure 2). Treatment with percutaneous coronary intervention (PCI) with balloon dilatation and aspiration thrombectomy was performed. Intracoronary and intravenous abciximab was administered. The final result was excellent with TIMI-3 distal flow with no residual stenosis or thrombus in the LAD. The patient was started on double dose clopidogrel 150mg daily and aspirin was continued. Her subsequent course in the hospital remained uneventful and she was discharged home. Genetic testing was done in an out-of-hospital laboratory revealing clopidogrel 2C19 genotype \*1/\*2 / Phenotype: intermediate metabolizer.

## DISCUSSION AND REVIEW OF THE LITERATURE

Stent thrombosis (ST) is an infrequent (0.5-1% of patients within 1 year) but life endangering complication of coronary artery stent implantation. The criteria of diagnosis and definition of ST created variance in the incidence of ST by different authors. To settle the problem, a joint panel called 'Academic Research Consortium (ARC)' was formed which developed the ARC definition of ST depending upon 'event certainty' and 'time frame'. <sup>1,2</sup> Depending upon the event certainty, ST is divided into definitive, probable, or possible. The definitive diagnosis of ST is made only after confirmation of ST by angiography during life or autopsy after death. In the absence of these definitive procedures, ST is either is probable or possible. Probable is considered when acute MI is diagnosed in the territory of the target vessel where stent had been implanted and probable MI being diagnosed in view of ECG findings, echocardiography, and biomarkers or death within 30 days. An unexplained death after 30 days is considered to be due to possible ST.

ST occurs more frequently in complex patients and lesions, especially in those with acute coronary syndromes, diabetes mellitus, chronic kidney disease, diffuse disease, small vessels, and bifurcation lesions requiring multiple stents.<sup>3,4</sup> Additionally, premature discontinuation of dual antiplatelet therapy within 6 months has been strongly associated with ST, especially in the setting of trauma or performance of surgical procedures. These patient-related factors can be critical in clinical decision making relative to the overall revascularization strategy: whether to implant a bare metal stent (BMS) or drug eluding stent (DES), or to instead consider coronary artery bypass grafting. Understanding the risk of ST according to these patient-related factors facilitates the use of procedural strategies to minimize the risk of ST, especially in high-risk patients.

Procedural factors associated with ST include the stent type selected (whether BMS or DES, and even the specific DES used), as well as whether the stent is adequately expanded and apposed to the vessel wall and is placed in a vessel with sufficient runoff to support adequate flow through the stent. Colombo and colleagues demonstrated with intravascular ultrasound that stent under expansion and/or malapposition occurred not infrequently after stent deployment and was associated with ST.<sup>5</sup> On the basis of these and other observations, adequate stent sizing and high-pressure stent deployment and post dilatation to ensure expansion are considered essential to minimize ST. Although the use of adjunctive intravascular ultrasound to ensure appropriate sizing and expansion has not been proven in randomized trials to be essential, intravascular ultrasound can be useful to confirm stent apposition and expansion, which in observational studies has been linked to lower ST at both 30 days and 1 year.<sup>6</sup>

# Mechanism of ST

Most drug-eluting stents are composed of three parts – metal platform, antirestenotic drug and polymer which serves for loading and modifying drug release. ST may be the end result of inflammation and injury due to either the drug

or polymer. The most powerful predictor of ST is the lack of endothelization of DES. The lack of neointimal coverage is accompanied with direct contact of blood with the strut. The exact mechanism of delayed or incomplete endothelization is not completely understood. Endothelium-dependent vasomotor dysfunction has been implicated. Increasing attention has been paid to the potential effect of polymers on subsequent hypersensitivity and inflammation. Concerns about the effect of polymers on vascular repair have driven efforts to either design DES with biodegradable polymers or develop nonpolymeric drug delivery. The LEADERS (Limus Eluted from A Durable vs. ERodable Stent coating) trial randomized patients to the conventional "biostable" durable polymer (Cypher) sirolimus eluding stent, SES or the Biomatrix biolimus A9-eluting stent with a biodegradable polylactic acid polymer. At 4-year follow-up, biodegradable polymer, biolimus eluding stent, BES maintained non-inferiority and improved long term clinical outcomecompared to SES. Finally, completely bioresorbable stent platforms are currently undergoing clinical trials. As presented at the AHA 2011 Meeting, angiographic follow-up at 2 years in the ABSORB (A Bioabsorbable Everolimus-Eluting Coronary Stent System for Patients With Single De-Novo Coronary Artery Lesions) trial of an everolimus-eluting platform demonstrated complete stent resorption, arterial healing, and apparent restoration of normal vascular function with no ST at 5 years.

#### DES vs BMS

Surrogate markers (death or MI) have been used for ST in multiple meta-analyses involving both randomized controlled trails (RCT) and observational data. Several conclusions regarding these outcomes following DES versus BMS can be drawn. RCT and observational data differ regarding the frequency of reported clinical outcomes. Although RCTs have demonstrated similar incidences of death and MI for both stent types, observational (real-world) datasets demonstrate an apparent reduction in mortality favoring DES. Both RCTs and observational studies demonstrate a substantial reduction in target vessel revascularization with DES.

## Clinical Profile of ST

Stent restenosis and ST have emerged as the chief complications of stent implantation. Restenosis presents as increasing angina requiring repeat revascularization. ST on the other hand is a life-endangering condition with catastrophic consequences. It usually presents as an acute myocardial infarction (39%) or sudden death (48%). MI in most cases presents as STEMI and can be associated with cardiogenic shock. STEMI with ST is a new group of high risk patients and their management should be improved with the use of modern intra-aortic balloon pump (IABP), intravascular ultrasound, thrombectomy devices and extensive use of GpIIb/IIIa inhibitors and antithrombotic drugs. No differences were found in the clinical profile and outcome of ST patients with DES and BMS. One wonders why stent thrombosis is so dreadful with a higher mortality and inferior revascularization results when compared with STEMI in de novo coronary arteries. The reason appears to be the unduly large thrombus burden in ST cases. Despite maximal inflation pressures during PCI and optimization efforts, a significant remaining resistant thrombus can often be visualized in patients treated for BMS or DES thrombosis. It is possible that thrombus is not only larger but also qualitatively resistant to mechanical and pharmacological interventions.

## Treatment of ST

Treatment of patients suffering from ST is a medical emergency of utmost urgency. Patients who have undergone PCI with stent implantation may report with severe substernal chest pain, acute left ventricular failure, cardiogenic shock or life threatening arrhythmias. Mechanical flow restoration should be achieved at the earliest. Besides antiplatelet and antithrombotic drugs, aspiration devices usually should be used during PCI and intravascular ultrasound guidance is indicated for proper stent optimization. The treatment may be followed by dual antiplatelet drug therapy in higher maintenance doses. In patients with acute coronary syndromes, the rates of ST have also been reduced by replacing clopidogrel with more potent antiplatelet agents such as prasugrel and ticagrelor, although this benefit is achieved at the cost of increased bleeding. <sup>13,14</sup> Double-dose clopidogrel for either a week or longer after stent implantation in order to prevent ST is largely empirically based rather than supported by clinical data. <sup>15</sup> An attempt should be made to determine platelet reactivity to confirm the presence of aspirin and clopidogrel resistance which will alert the physician to adopt alternative drugs and doses to achieve ideal platelet aggregation inhibition. <sup>16</sup> Newer stents e.g. bio-absorbable stents and 2<sup>nd</sup> or 3<sup>rd</sup> generation stents, are associated with reduced incidence of ST. Polymer of bio-absorbable stents get absorbed in 6-9 weeks after implantation.

At least 12 months of dual antiplatelet therapy is recommended after DES implantation. Hazards of premature discontinuation of dual antiplatelet therapy must be clearly understood by the patient and physician. DES should be avoided in patients who are unlikely to comply with 12 months of dual antiplatelet therapy. Encoding the CYP2C19 allele as well as high on clopidogrel platelet reactivity have been associated with adverse clinical events in patients

undergoing PCI. Genetic testing is not wide-spread, its utility has not been prospectively validated, and what to do if a poor metabolizer is identified is uncertain. At present, no prospective randomized studies have demonstrated benefits of using a more potent alternative antiplatelet regimen (such as higher-dose clopidogrel, prasugrel, or ticagrelor) for stable PCI patients identified at increased risk for events on clopidogrel by either a polymorphism in CYP2C19 or high on-treatment residual platelet reactivity. The first such randomized trial to examine this hypothesis, Gauging Responsiveness With a Verify Now Assay: Impact on Thrombosis and Safety (GRAVITAS) recently reported no benefit of a strategy of doubling the standard daily dose of clopidogrel (from 75 to 150mg per day) after PCI in patients with high on-treatment platelet reactivity. <sup>17</sup> Role of such testing is recommended case-by-case and not universally to all the patients with stent implantation.

## Prevention of ST

Proper selection of stents depends upon the anatomy of the coronary lesion and compliance of the patient. Optimization of stent implantation is a super-specialty. Expertise in proper placement and apposition of stent comes by proper training and practice. Routine use of intravascular ultrasound may help in optimal deployment and apposition of DES and reduces the need of repeat revascularization and ST. Elective surgery should be postponed for 12 months or as long as patients remain on dual antiplatelet therapy. For urgent and life-saving surgery, discontinue clopidogrel for 5 days prior to surgery but continue aspirin.

## CONCLUSIONS

ST is an infrequent but devastating complication of stent implantation. The diagnosis of ST should be suspected in a case of MI in the territory of the coronary artery where a stent had been implanted earlier. Definitive diagnosis of ST can only be made by angiography during life. The clinical profile of ST is MI often complicated with cardiogenic shock, hypotension, arrhythmias, poor LVEF and early death. Intensive life support measures, mechanical help to support ventilation and perfusion, vasopressors, antithrombotic drugs, and glycoprotein IIb/IIIa inhibitors are often urgently required. Revascularization (CABG/PCI) is preferred over fibrinolytic procedures with decision making sometimes in joint consultation with cardiac surgeon and invasive cardiologists. If PCI is preferred, the use of thrombectomy devices and intravascular ultrasound guided implantation of stents are advisable. Novel stents are emerging with the potential to inherently lower the risk of ST. Elective surgery within the first year after DES placement should be avoided or performed without discontinuation of either aspirin or clopidogrel if possible. The majority of hospital deaths occur within the first 48 hours of ST, emphasizing the importance of early intervention which can result in myocardial salvage, preservation of LV function and fewer in-hospital complications. Prolonged or life-long dual antiplatelet therapy, liberal use of statins, and judicious administration of other supportive drugs (like ACE inhibitors, beta blockers) are usually required following successful revascularization. Finally, data on the use of antiplatelet agents more potent than clopidogrel for high-risk patients are limited to those with acute coronary syndromes, for whom prasugrel and ticagrelor can be beneficial, with potentially greater risk of bleeding complications. It is thus essential to carefully consider the individual patient's risk of ST compared to bleeding before using these agents.

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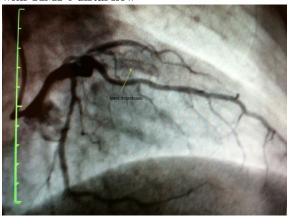
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Figure 1. Patent proximal LAD stent with TIMI-3 distal flow



Figure 2. Stent thrombosis in proximal LAD with TIMI-0 distal flow



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# **Review Article: Toxicology of Clonidine**

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# CASE REPORT

A 14 year-old female with a past medical history of motor tic disorder presented to the hospital after intentional overdose of her prescribed medication. She had a 4 year history of tics. She had various tics, starting with facial grimacing and neck movements. Her latest tic was especially bothersome to her and involves pushing both hands into her abdomen. She states that she was able to suppress the tics during the day. When she was alone at night she had the urge to tic and the tic was sometimes relatively strong and uncomfortable. A pediatric neurologist had prescribed her a medication for the motor tics 3 months prior.

The children at school were constantly making fun of her tics and it made her very upset. The day prior to presenting to the hospital she took 10 of her prescribed pills in an attempt to end her life. She woke up 2 hours later and was dizzy, reporting that the room was spinning. She tried to lie back down but was too uncomfortable. Due to the discomfort and remorse for ingesting the pills, she told her parents what she had done. Her parents immediately took her to the Emergency Department.

On presentation to the ED, her vital signs were T 98.4F, P 49, R16, BP 79/29, saturating 96% on room air. She was dizzy, disoriented to place and time, and her face appeared red. She had dry mucous membranes and her pupils were

2mm and reactive to light. She did not void in the ED. She was given IV fluids, a medication, and a foley catheter was placed. The patient was then transported to the Pediatric Intensive Care Unit for further evaluation and management.

On arrival at the Pediatric Intensive Care Unit, her vital signs were: T98.3, P66 R21 BP123/56, saturating 99% on room air. Physical examination: General – Alert, talking to herself, not oriented to place or time. She was able to answer questions regarding symptomatology if re-directed. Responded to painful stimuli. HEENT- pupils 2mm, reactive to light. Dry mucous membranes. LUNGS- normal, CVS- normal, ABD- normal, SKIN- reddened, especially over the face. The patient had limited neurological examination findings due to uncooperativeness but was found to be hypotonic. Glasgow coma scale =12.

### DISCUSSION

The patient had symptoms from taking excessive amounts of Clonidine (2mg taken; max 0.3 mg/day), her prescription medication that was given to her for her motor tics. Naloxone was the antidote medication given to the patient due to the altered mental status and hypotonia. Atropine was used for the patient's bradycardia.

Clonidine, an alpha-2 agonist, was initially used as a topical nasal decongestant over 40 years ago. In present day, it has been primarily utilized for its potent antihypertensive effect. Clonidine is also used in the treatment of behavioral disorders, migraine headaches, and narcotic withdrawal. In children it is used in the treatment of attention deficit disorder with hyperactivity, refractory conduct disorder, and Tourette's syndrome. Clonidine inhibits neurons in the vasoregulatory center of the medulla. This results in decreased central sympathetic outflow. Clonidine seems to also affect nitric oxide and gamma-aminobutyric acid activity. Clonidine is available in parenteral, oral and transdermal forms. Clonidine is supplied as 0.1, 0.2, and 0.3 mg tablets and in transdermal patch delivery systems. Clonidine is also available in a combination product containing the diuretic agent chlorthalidone. \( \)

### Clonidine Overdose

In the United States, about 6000 calls regarding clonidine exposure are made annually to regional poison control centers, and serious clinical findings are common.<sup>2</sup> Up to 60% of clonidine exposures that are reported to regional poison control centers result in symptoms. In a retrospective study looking at clonidine-only exposures reported to American Association of Poison Control centers from 1993 to 1999, 10,600 children under 19 years of age were analyzed. Unintentional overdose was most common in children younger than 6 years of age. Therapeutic errors were most common in 6-12 year olds. Suicide attempts were most common in adolescents. Of the 10,600 children, 6042 were symptomatic (60%). Most common symptoms included lethargy (80%), bradycardia (17%), hypotension (15%), and respiratory depression (5%). Out of the entire study, there was one fatality.<sup>3</sup>

The absorption, distribution, and elimination of clonidine varies depending upon the formulation of clonidine and route of exposure. Clonidine is 20-40% protein bound in the bloodstream. Bioavailability is approximately 75-100%. Elimination half-life in therapeutic use ranges from 6-24 hours with more than half excreted unchanged in the urine. In children, symptoms develop within 1 hour of ingestion. Peak plasma clonidine concentrations occur at approximately 2 hours. New findings rarely appear more than 4 hours after exposure. Maximal blood pressure reduction occurs between 3-8 hours after a routine daily dose. Severe symptoms can occur in children with as little as 0.1mg of clonidine, especially in children less than 4 years of age. No globally applicable minimum toxic dose of clonidine has been established.

### Management

As with any evaluation in a patient with suspected drug ingestion, all assessment begins with a thorough history and physical examination with careful inspection of the skin for any transdermal patches. The classic toxidrome associated with clonidine consists of central nervous system depression, bradycardia, hypotension, respiratory depression, and small pupil size. This clinical presentation makes the differentiation of clonidine poisoning from opioid toxicity difficult. Transient responsiveness to painful stimuli is more frequently seen in patients poisoned with clonidine than with opioids.

Respiratory depression and apnea that accompany clonidine poisoning may require immediate airway support. Children will often resume breathing upon tactile stimulation. Bradycardia and hypotension are prominent signs of clonidine toxicity. Atrioventricular (AV) block and sinus arrest have been described in clonidine toxicity. Second-degree AV block or complete AV dissociation are uncommon and should prompt consideration of toxicity from

other cardioactive agents, such as digoxin, beta blockers, or calcium channel blockers. In many patients with clonidine overdose, hypertension may occur initially before hypotension, particularly after a large ingestion. In addition, hypertension has been reported in patients with clonidine poisoning who receive naloxone. Hypertensive emergency requiring treatment is very rare. In the unlikely event that hypertension must be treated, a short-acting, easily-titratable agent such as nitroprusside is advised.

Testing for clonidine levels in the body is available but cannot be obtained in time to be useful for effective patient management. Specific measurement of clonidine in the urine or other fluids is typically performed for forensic or other nonclinical purposes.

There are various testing regimens that are recommended in adolescent patients with altered mental function and clonidine overdose. Bedside glucose determination is done to exclude hypoglycemia as the cause of reduced consciousness. Arterial blood gas evaluation or venous blood gas measurement are combined with pulse oximetry to assess adequacy of ventilation in patients with serious and persistent respiratory depression. Serum acetaminophen level is drawn in patients who ingest clonidine with suicidal intent. Serum ethanol level identifies patients that are at a higher risk for coma and respiratory depression due to co-ingestion of clonidine and ethanol. Serum electrolyte determination assesses for electrolyte disturbance. The anion gap can be calculated in the setting of likely co-ingestants or evidence of metabolic acidosis. In order to assess for renal insufficiency, serum blood urea nitrogen and creatinine are checked. Patients with renal insufficiency may have delayed elimination of clonidine after overdose. An electrocardiogram (ECG) evaluates for drug-induced cardiac electrical dysfunction. Chest radiography is done if the history or exam suggests the possibility of pulmonary aspiration.

Management begins with assessment and stabilization of the airway, breathing, and circulation. Clonidine binds well to activated charcoal, the primary means of decontamination in overdose. Patients who present within 1-2 hours of a known or suspected ingestion of clonidine tablets receive activated charcoal (1 gm/kg, maximum dose 50 grams) by mouth or nasogastric tube. Patients with respiratory depression or coma should first have their airway secured prior to activated charcoal administration. In patients who present for care more than 2 hours after ingestion of clonidine tablets, activated charcoal should not be given because the likelihood of preventing medication absorption is low and is outweighed by the risk of pulmonary aspiration.

If the patient ingested a clonidine transdermal patch, nasogastric administration of whole bowel irrigation (500 mL to 1 L of polyethylene glycol per hour) after oral administration of activated charcoal (1 gm/kg, maximum dose 50 grams) should be undertaken.<sup>3</sup>

No true antidote for clonidine intoxication exists. Naloxone and alpha adrenergic antagonists have been utilized to treat seriously poisoned patients with inconsistent results. Many patients who do respond to the initial dose of naloxone do not require additional doses. Clonidine poisoned patients with marked CNS depression and apnea receive intravenous naloxone (0.1 mg/kg, maximum dose: 2 mg; may be repeated every 1 to 2 minutes up to 10 mg total dose). Patients who improve after naloxone administration still warrant hospital admission and intensive monitoring for recurrence of symptoms. Pressor agents, such as dopamine, may be useful in the minority of patients whose bradycardia or hypotension does not respond to intravenous atropine administration and rapid intravenous infusion of isotonic fluids.

# CONCLUSION

Clonidine is a medication used in many pediatric conditions. Characterization and supportive management of overdose needs to be recognized before life threatening sequelae occur.

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