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Creutzfeldt-Jakob Disease

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History of Present Illness

A male in his 40s presents with six weeks of gradually progressive neurologic complaints. He initially noticed blurred vision and a sensation of ear fullness. He was evaluated multiple times at an outside hospital for those complaints, and his evaluation and treatment included a negative head CT and two courses of antibiotics for presumed otitis media. His symptoms persisted, so he was referred to a neurologist who scheduled an outpatient MRI which had not yet been performed. On presentation to the University of Cincinnati Emergency Department (ED), he reports progressive and new neurologic symptoms including clumsiness, intermittent numbness in his hands, balance problems, and slurred speech. His symptoms have become so severe that he is no longer able to perform his daily activities.

Past Medical History
Bells Palsy (10+ years ago)

Past Surgical History
None

Medications
None

Vitals
T 97.9  HR 78  BP 168/94  RR 13  SpO2 98% on RA

Physical Exam

The patient is a well-appearing male who is awake, alert, and oriented. Respiratory, cardiovascular, and abdominal examinations are normal. Neurologic examination reveals diplopia with left gaze, spastic dysarthria, and dysphonia. The patient also exhibits dysmetria on the left with finger-nose and heel-shin testing as well as truncal ataxia and a wide-based, unsteady gait. Otherwise, cranial nerve testing, motor strength, and sensation are normal.

ED Labs and Imaging

- Glucose 109
- BUN 28, otherwise BMP normal
- WBC 6.4, Hgb 16.0, Plt 169
- AST 42, ALT 67
- CTA head & neck: No flow-limiting stenosis, aneurysm, or evidence of dissection.

Hospital Course

Neurology was consulted from the ED and recommended admission for further workup. The patient underwent an extensive laboratory workup including thyroid studies, B12, HIV, syphilis, hepatitis, vitamin E, ceruloplasmin, antinuclear antibody, tissue transglutaminase, gliadin, and heavy metal testing. These tests were normal. Initial cerebrospinal fluid (CSF) studies included cell count, protein, glucose, gram stain, flow cytometry, paraneoplastic panel, cytomegalovirus, herpes simplex virus, enterovirus, varicella, West Nile, and Lyme disease. These tests were also normal. MRI head was obtained which demonstrated findings consistent with Creutzfeldt-Jakob disease (CJD) (figure 1). Confirmation with CSF RT-QuIC testing was sent for testing at an outside facility.

The patient was discharged on hospital day four and had progressive neurologic deterioration including seizure-like activity and rapidly progressive dementia. Two weeks after his initial pre-
presentation, the RT-QuIC testing returned positive, further suggesting a diagnosis of CJD. The family declined a brain biopsy for definitive diagnosis. Palliative care was consulted, and the patient expired 12 days later. Family declined an autopsy.

Discussion

CJD is a transmissible, rapidly progressive, uniformly fatal neurodegenerative condition caused by the misfolding and pathologic aggregation of human prion protein. The condition is rare, affecting an estimated 1 in 1,000,000 people annually. CJD classically presents as a rapidly progressive dementia with focal neurologic deficits, akinetic mutism, and myoclonus. MRI and CSF studies may be suggestive of CJD, but the diagnosis should only be considered after all potentially treatable causes have been excluded.

Multiple clinical variants exist, including spontaneous (sporadic CJD, 85% of cases), genetic (familial CJD, 10% of cases), and iatrogenic or acquired (variant CJD). There are case reports of CJD transmission from grafts of dura mater, transplanted corneas, implantation of inadequately sterilized electrodes in the brain, and injections of contaminated pituitary growth hormone derived from human pituitary glands taken from cadavers. All human growth hormone used in the United States is now synthesized by recombinant DNA procedures, eliminating the risk of transmitting CJD by this route. All variants are transmissible via blood, CSF, and CNS tissue, and there is overlap among variants in terms of clinical presentation, disease course, and diagnostic testing.

CJD has classically been described as a rapidly progressive dementia with focal neurologic deficits and myoclonus. In practice, there is marked variability in the presenting symptoms of patients with CJD. Sporadic CJD most commonly presents in individuals 60-70 years of age, whereas variant CJD tends to affect younger individuals with a mean age at diagnosis of 28 years. Early symptoms are often non-specific and psychiatric in nature, including depressive personality changes, sleep disorders, behavioral disturbances and other psychiatric complaints. Interestingly, 12% of referrals for suspected CJD are made by psychiatrists. An unrelenting, rapidly progressive dementia typically follows; symptoms include diminished cognition, impaired memory and generalized loss of higher cerebral function. Focal neurologic deficits tend to develop as the disease progresses, but rarely may be the presenting complaint. Cerebellar dysfunction predominates, with gait ataxia present in a majority of cases. Visual disturbances are common and range from subjective blurriness to visual hallucinations and cortical blindness. Myoclonus is often a late finding and may be elicited via the startle response to tactile or auditory stimuli. A hallmark of the late stage of disease is akinetic mutism, a term which describes a patient’s inability to purposefully move or vocalize. Seizures occur in approximately 15% of cases.

Care is supportive and palliative. Opiates, valproate, and benzodiazepines may be considered for symptomatic management. There are no treatments that alter disease progression or mortality. The disease is invariably fatal; death occurs on average five months after the onset of symptoms in patients with sporadic CJD.

The differential diagnosis of rapidly progressive dementia is broad, and importantly contains both treatable and non-treatable causes. Autopsy studies from the US National Prion Disease Pathology Surveillance Center, which analyzes tissue samples and clinical information for all referred cases of suspected CJD in the US, demonstrate that over 30% of cases referred as suspected prion disease were negative for pathologic evidence of CJD. The majority of these cases were diagnosed post-mortem as Alzheimer disease, vascular dementia, or another incurable neurodegenerative condition. More impor-

<table>
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<th>Common Treatable Causes of Rapidly Progressive Dementia</th>
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<tr>
<td>Immune-mediated</td>
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<td>Acute Disseminated Encephalomyelitis</td>
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<th>World Health Organization Criteria for the Diagnosis of Sporadic CJD</th>
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<td>III A</td>
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<td>III B</td>
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</table>

Possible CJD: I and at least 2/4 of II and duration < 2 years
Probable CJD: I and at least 2/4 of II and at least 1/2 of III and duration < 2 years
Definite CJD: Neuropathologically confirmed diagnosis

Table 2: WHO diagnostic criteria for CJD.

Table 2: WHO diagnostic criteria for CJD.
The patient is a female in her late 40s with a past medical history of multiple sclerosis and psychogenic non-epileptic seizures who presents via EMS with altered mental status. Per EMS and family, the patient has been unusually somnolent and lethargic for the past two hours. A similar event occurred approximately two weeks prior after the patient consumed an unknown quantity of laundry detergent pods. The family is now concerned that the patient has ingested laundry detergent again. The patient herself is unable to provide further history.

**Past Medical History**
- Multiple sclerosis
- Psychogenic nonepileptic seizures
- Anxiety
- Endometriosis
- Scleroderma
- Prior suicide attempt via benzodiazepine overdose

**Past Surgical History**
- Caesarean section x2
- Tonsillectomy
- Antrectomy with Roux-en-Y gastrojejunostomy

**Medications**
- Amitriptyline
- Baclofen
- Dextroamphetamine-amine-amphetamine
- Diazepam
- Ergocalciferol
- Fluoxetine
- Furosemide
- Interferon beta-1a
- Lisdexamfetamine
- Omeprazole
- Prochlorperazine
- Quetiapine
- Sumatriptan

**Vitals**
- BP 121/76
- P 87
- T 36.6
- RR 25
- SaO2 95% on room air

**Physical Exam**

Well-nourished, well-developed adult woman in no acute distress. Normocephalic with intact extraocular movements and no nystagmus. Pupils equally round and reactive to light. Cardiac exam reveals a regular rhythm and rate with intact peripheral pulses. Lungs clear to auscultation bilaterally with normal work of breathing and no respiratory distress. Her abdomen is soft, non-tender, with no rebound or guarding. Genitourinary exam shows normal female external genitalia with no hemorrhoids, fissures, or masses; normal rectal tone with guaiac-negative stool. Neurologic exam is notable for a depressed mental status although she is arousable to verbal stimuli. The patient follows commands with notable diffuse weakness in all four extremities. Normal deep tendon reflexes without clonus. No obvious sensory deficits. Speech is slurred. Grossly depressed mood but no obvious suicidal ideation. Patient’s skin is warm and dry.

**Labs & Diagnostics**
- WBC 17.4
- Hgb 9.2
- Platelets 771
- Urinalysis and urine pregnancy negative
- Urine drug screen positive for tricyclic antidepressants
- CT Head: No definite interval changes in patient’s demyelinating disease
- CT abdomen and pelvis: Marked gastric distention with abnormal gastric debris in the setting of prior antrectomy

**Hospital Course**

Given the patient’s significantly depressed mental status in the context of known neurologic disease, neurology was consulted in the emergency department. The neurology team independently evaluated the patient, but had a low concern for neuraxial pathology as the primary cause of the patient’s somnolence. Neurology recommended EEG to rule out atypical seizures causing the patient’s altered mental status in addition to further observation pending improvement in her cognition.

Psychiatry was consulted the following day (hospital day one) to evaluate the patient and provide recommendations. On their examination, the patient’s mental status had improved dramatically. Furthermore, the patient adamantly denied any self-injurious intent or suicidal ideation. As such, psychiatry felt that there were no significant safety concerns at the time and that patient could safely be discharged home with a slight reduction in her quetiapine dose pending medical clearance.

The patient was subsequently discharged from the hospital on hospital day two. As the patient’s mental status had improved spontaneously, more aggressive diagnostic and therapeutic interventions - such as endoscopy - were not pursued. Instead, the patient was counseled on the risks of ingesting caustic agents and was instructed to follow-up with her neurologist as an outpatient for further evaluation and titration of her medications.
Ingestion of detergent agents, whether intentional or accidental, can cause serious and potentially life-threatening toxicologic disease. While ingestion in the adult population often stems from psychiatric disease or self-injurious behaviors, the advent of water-soluble laundry detergent pods has led to a significant increase in the number of pediatric poisonings, due in part to their bright coloration and pleasant smell. This article will discuss the various manifestations of toxic exposure, pathogenesis, and evidence-based management of detergent poisonings in both the pediatric and adult populations.

The exact mechanism by which detergent causes injury has not been fully elucidated but is presumed to be multimodal in nature. Detergents are composed of a complex mixture of various chemical entities, including caustic alkali agents (such as sodium carbonate or sodium silicate), surfactants, anti-redeposition agents, fragrances, opacifiers, and many other non-toxic ingredients. These compositions are specially formulated to loosen, solubilize, and emulsify debris while simultaneously chelating inorganic solutes and prevent redeposition. While the caustic alkaline agents are typically viewed as the culprit pathological entity, the other chemical constituents of detergents likely contribute to or exacerbate these injuries.

In-vitro histological studies and case reports indicate that caustic alkaline substances may rapidly induce liquefactive necrosis of exposed mucosal structures. Esophageal mucosa may develop transmural injuries after only one second of exposure to 30% sodium hypochlorite solution. This is not to be confused with household bleach which is composed of only 5.25% sodium hypochlorite. On initial exposure, mucosal tissues quickly become inflamed and edematous before progressing to frank necrosis within minutes to hours. The hours to days following an ingestion are characterized by progressive inflammation and local superficial thrombosis, with evidence of wound granulation by day 10. During this period, the nascent wound is at increased risk of perforation. By three weeks, however, fibrin deposition and recollagenization mechanisms have progressed, producing a mature, stable wound similar to normal tissue. More severe or extensive injuries may eventually progress to scarring and stricture formation around three weeks to one month.

Pharyngoesophageal insults occur most frequently given their relative position in the aerodigestive axis. Gastric injuries are also common but tend to be less severe due to partial neutralization of the caustic substances in the acidic medium of the stomach. Injuries of the duodenum and more distal enteric structures are less common. This is likely due to dilution of the offending agent. However, more distal injuries are often associated with significant morbidity, such as visceral perforation and peritonitis. Additionally, regurgitation and aspiration may lead to inhalation of the caustic entity into the tracheobronchial tree, potentially leading to respiratory compromise.

Serious mucosal injuries following ingestion of detergents and other caustic household products tends to be rare and correlate with the concentration of the agent as well as the volume imbibed. A grading scale has been developed to categorize the severity of esophageal injury and likelihood of long-term sequelae based on endoscopic appearance (Figure 1). Higher grade injuries (>2B) have been shown to correlate with increased risk of serious pathology, such as gastric or esophageal cicatrization, necessitating surgical or endoscopic intervention, while low grade injuries often require no further diagnostic or therapeutic intervention. The utility of this grading scale is limited in the emergency department, however, by the need for formal fiberoptic endoscopy and the lack of reliable symptomatic predictors. Nonetheless, urgent endoscopic evaluation should at least be considered in all possible cases of caustic ingestion in order to identify the extent of mucosal injury.

Clinical manifestations of caustic ingestions are varied, but often present with severe oropharyngeal, chest and/or abdominal pain, and dysphagia/odynophagia. Hematemesis is suggestive of gastric involvement, while abdominal distention, rigidity, and objective fevers may indicate visceral perforation causing pneumoperitoneum and/or mediastinum. While clinical symptoms have not been shown to correlate with the severity of injury, vital sign abnormalities (such as fever and hypotension) and clinical evidence of peritonitis should warrant urgent endoscopic evaluation. Coughing, drooling, hoarseness, and respiratory distress are suggestive of laryngeal exposure and glottic edema, and should prompt emergency providers to strongly consider establishing a definitive airway early in the patient's course given the potential for progressive disease causing respiratory compromise.

On arrival to the emergency department, providers should assess for evidence of oropharyngeal injuries and respiratory distress, including tachypnea, stridor, pooled secretions, and accessory muscle use, as these may indicate the need for laryngoscopy and endotracheal intubation. Objective fever or hypotension in conjunction with severe chest pain or signs of peritonitis (e.g., distended abdominal, exquisite pain to palpation) should raise concern for a perforated viscus and should prompt emergent surgical consultation. Even well-appearing, hemodynamically stable patients warrant a discussion with an endoscopic specialist urgently, given the aforementioned lack of correlation between symptom severity, degree of injury, and potential need for esophagogastroduodenoscopy. Emergency physicians may consider concomitantly contacting a local poison control center to discuss the case with a medical toxicologist, who may provide further guidance in developing a treatment plan and disposition for the patient.

Fiberoptic endoscopy remains the definitive diagnostic tool, though its use is contraindicated in patients with findings concerning for perforation. While lacking sensitivity, plain chest radiographs provide a cost-effective and quick method of assessing for free sub-diaphragmatic or mediastinal air as well as possible pneumonitis. Esophagography with water-soluble contrast is another viable modality to assess for gross dysmotility and perforation, though it lacks the ability to categorize mucosal changes or the extent of an exposure. Modern high-resolution computed tomography may demonstrate inflammation in exposed mucosal tissue as well as evidence of free air within the mediastinum or peritoneum.

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### Caustic Injury Grading

<table>
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<tr>
<td>0</td>
<td>Normal</td>
</tr>
<tr>
<td>1</td>
<td>Mucosal edema and hyperemia</td>
</tr>
<tr>
<td>2A</td>
<td>Superficial ulcers, bleeding, exudates</td>
</tr>
<tr>
<td>2B</td>
<td>Deep focal or circumferential ulcers</td>
</tr>
<tr>
<td>3A</td>
<td>Focal necrosis</td>
</tr>
<tr>
<td>3B</td>
<td>Extensive necrosis</td>
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Case 1

History of Present Illness

The patient is an elderly female with a history of chronic obstructive pulmonary disease (COPD) and a prior cerebrospinal fluid (CSF) leak status post surgical repair complicated by multiple previous episodes of meningitis who presents to the emergency department (ED) for altered mental status. Her symptoms began upon awakening on the day of presentation, and she was last seen normal before going to bed the night before. She has no history of seizures, but her family reported she had a shaking episode prior to arrival. Glucose in the field was 197.

Past Medical History

- COPD
- Meningitis

Allergies

- Erythromycin, cephalaxin

Past Surgical History

- Multiple previous surgeries for CSF leak repair

Medications

- Acetazolamide
- Pantoprazole
- Simvastatin

Vitals

- T 100.8, HR 140, RR 41, BP 174/104, O2 99%

Physical Exam

Physical exam is notable for an elderly female who is not responsive to verbal or painful stimuli. She is ill-appearing with shallow breathing and tachypnea. No evidence of head trauma. Her eyes are open with a conjugate gaze, but she has a sluggish pupillary response bilaterally and no blink to threat. There is no response to noxious stimuli but she does have a gag and cough. She is tachycardic with a regular rhythm. Pulmonary exam reveals coarse breath sounds in all lung fields. Her abdomen is benign. No obvious rash noted on skin exam.

Labs

- CBC: WBC 32.6 with 20% bands / Hgb 15.9 / Hct 47.9 / Plt 277
- BMP: Na 139 / K 3.1 / Cl 105 / HCO3 18 / BUN 19 / Cr 1.18 / BS 167
- Lactate: 6.7
- VBG: pH 7.35 / pCO2 35 / HCO3 19 / BE -5.8
- Trop: 0.78 (EKG without ischemic changes)
- UA: Negative
- CSF: see Table 1 (page 12)

Imaging

- CXR: multifocal consolidations concerning for pneumonia
- CT head: Focal gyral swelling and hypodensity within the left frontal lobe with adjacent sulcal hyperdensity concerning for focal encephalitis/meningitis along with a small hyperdensity within the insula concerning for subarachnoid hemorrhage (figure 1).
- CTA head and neck: Mild atherosclerotic disease without significant stenosis or aneurysm

Hospital Course

Based on the patient’s presentation, there were initial concerns for an acute intracranial process including meningitis. Blood cultures were drawn and she was fluid resuscitated with intravenous crystalloid. Based on her allergy profile, she was empirically treated for meningitis with vancomycin, piperacillin-tazobactam, acyclovir, and dexamethasone. She was also given two doses of midazolam for concerns for non-convulsive status epilepticus. Given the patient’s respiratory and neurologic status, she was intubated using delayed sequence induction with topical and nebulized lidocaine and ketamine.

Following intubation and imaging, a lumbar puncture was performed showing purulent fluid with gram-positive cocci in pairs concerning for pneumococcal meningitis. She was admitted to the neuroscience intensive care unit (NSICU) for definitive care. While hospitalized, her blood and CSF cultures grew Streptococcus pneumoniae. Her hospital course included anticoagulation for a cortical vein thrombosis resulting from meningitis. The patient ultimately had a tracheostomy for long-term ventilator weaning and was transferred to a long-term acute care rehabilitation facility.
History of Present Illness

A male in his 60s presents to the ED with altered mental status, generalized weakness, dizziness, and gait abnormalities. His symptoms began after a mechanical fall five months prior, but he notes a sharp decline over the past two days. No recent fever, headache, or neck pain. However, review of his records is notable for at least four visits to healthcare facilities over the past four months with similar symptoms, including headache and dizziness. This resulted in various diagnoses including post-traumatic headache, cognitive communication deficit, vertigo, hydrocephalus, and seizure.

Four months prior to this presentation, he was transferred from an outside hospital to the neurology service at a tertiary care facility for headache, altered mental status, and concern for seizure activity. His imaging revealed mild hydrocephalus, and he was diagnosed with meningoencephalitis. He was empirically treated with vancomycin, ceftriaxone, ampicillin, and acyclovir. These medications were quickly discontinued based on CSF cell counts (see table 1), negative herpes simplex virus testing, and a negative electroencephalogram. Nine days after hospital discharge, he was seen in the ED for similar complaints and discharged home with a diagnosis of vertigo after a CTA head and neck came back negative for dissection. Two days prior to his current presentation, he presented to an outside hospital where a repeat head CT showed unchanged hydrocephalus, and he was again discharged home.

Case 2

The patient presented with a low suspicion for infection given the subclinical nature of his decline and lack of fever, headache, or neck pain. Therefore, empiric antibiotics and antivirals were not initiated. After neuro-imaging demonstrated evidence of worsening hydrocephalus, the patient was admitted to neurology for further management. While inpatient, he underwent a lumbar puncture (LP) which resulted in a diagnosis of cryptococcal meningitis and he was started on amphotericin and flucytosine. He developed worsening mentation and respiratory distress requiring intubation and transfer to the NSICU. He ultimately did well, was successfully extubated, and was discharged on high dose fluconazole for eight weeks.

Discussion

Meningitis is a rare but potentially devastating disease with a mortality as high as 40% depending on the causative organism. Predisposing factors, such as otitis media, sinusitis, pneumonia, or immunocompromised states are seen in nearly half of all cases of bacterial meningitis. Pneumococcal meningitis is more likely to spread from a distant focus of infection (such as pneumonia), thus making it imperative that providers maintain a high degree of suspicion for meningitis in patients with possible pneumococcal infections.

Nonetheless, the presentation of meningitis is widely variable depending on the causative organism. As highlighted by the above cases, bacterial meningitis often presents with rapid progression of neurologic symptoms and quick clinical deterioration. Conversely, fungal meningitis more commonly has a prolonged course and is frequently misdiagnosed in the early stages. Therefore, emergency providers should be intimately familiar with the identification and treatment of all causes of meningitis.
Lumbar puncture (LP) is a procedure that is regularly successfully performed in the emergency department using manual palpation of anatomic landmarks. Nonetheless, failure rates as high as 19% have been reported. This is often attributed to difficult anatomy or an inability to identify landmarks due to obesity. In fact, one study found that unsuccessful LPs most frequently occur in patients with a BMI above 35, suggesting that obesity is a strong predictor of difficulty for this procedure. Other factors that portend complexity include vertebral abnormalities, scoliosis, and degenerative changes that create narrow interspinous spaces. Such challenging anatomy may result in multiple procedural attempts which may lead to increased complications, frequent needle redirection, frustrating traumatic taps, and the occasional need for consultant involvement (e.g., interventional radiology). Ultrasound can be a helpful adjunct for proceduralists as it allows for identification of structures and recognition of difficult anatomy.

Introduction

Positioning

Position the patient as one would when performing a routine LP. Upright positioning is most desirable because it more effectively opens the interspinous spaces, but this is not always feasible for certain patients. Right or left lateral decubitus positioning is also effective, making certain that the patient’s hips and shoulders are perpendicular to the bed to prevent twisting of the vertebrae. This position allows for accurate measurement of the opening pressure, a number which impacts management and mortality in certain cases of meningitis. It is important to maintain this positioning throughout landmark identification, needle insertion, and procedural completion.

Identification of Landmarks

Either the linear array or curvilinear probe can be used for this procedure, and probe selection should be based on the patient’s body habitus. For thin patients, the linear array probe is desirable due to its clear imaging capabilities of superficial structures. For larger patients, the curvilinear probe is preferred because it can penetrate deeper into the tissues.

1. Using the selected probe, start in the longitudinal plane with the indicator mark towards the patient’s head. Place the probe over the sacrum at the superior aspect of the gluteal cleft.

2. Identify the sacrum which is a hyperechoic line lying horizontally on the ultrasound screen (figure 2).

3. Slide the probe superiorly until spinous processes are identified. These will appear as small, curved hyperechoic lines with
hypoechoic shadowing posteriorly (figure 3).

4. Identify the interspinous space between the spinous processes (figure 3). Continue sliding the probe superiorly, counting the spinous processes while moving. Center the L4-L5 interspinous space in the ultrasound screen and make a small skin mark perpendicular to and in the center of the probe at this site.

   a. Of note, the ligamentum flavum can be visualized on this view. In morbidly obese patients, it may be useful to measure the distance from the skin to this ligament. This distance will determine the size of spinal needle that will be required to complete this procedure.

5. Slide superiorly and make a similar skin mark at the site of the L3-L4 interspinous space.

6. Rotate the probe 90° with the indicator pointing toward the patient’s left.

7. Start with the probe over the sacrum at the superior aspect of the gluteal cleft, now scanning the spine in the transverse plane.

8. Slide the transducer superiorly to identify the spinous process of each lumbar vertebra. These can be recognized as hypoechoic structures similar to what is shown in figure 4.

9. Make a small skin mark perpendicular to the probe directly over each spinous process to demarcate midline.

10. Connect the lines drawn over the interspinous spaces with those in the midline (figure 5).

11. Perform the LP using standard technique with these intersections demarcating the needle insertion sites that can be used to enter the L4-L5 and L3-L4 interspaces respectively.

Discussion

When body habitus or anatomic variants make palpation of landmarks impractical, ultrasound can be a helpful adjunct when performing an LP. Several studies have shown that utilization of ultrasound to identify landmarks helps to minimize the number of attempts and reduces failure rates, particularly in elderly and obese patients. One meta-analysis demonstrated that ultrasound-assisted LP reduces the number of traumatic taps and needle redirections. Nonetheless, routine ultrasound use in all patients has not been shown to decrease failure rates in patients without predicted difficult anatomy. Therefore, while it should not supplant thorough palpation and traditional landmark identification, ultrasound recognition of lumbar spine anatomy is a useful skill that emergency providers can utilize in challenging cases.

As the use of oral anticoagulants increases, it is essential for the emergency medicine provider to have an understanding of the reversal of these agents due to bleeding complications and the need for emergent reversal. Currently, only warfarin and dabigatran have targeted reversal agents on the market to mediate their effects. Research is being conducted to formulate agents for the reversal of the other direct oral anticoagulants (DOACs): rivaroxaban, edoxaban, and apixaban. The off-label use of prothrombin complex concentrate (PCC; trade name: Kcentra) is often relied on to mediate the anticoagulant effects for many of the oral anticoagulants due to the lack of a specific agent or time to onset of action. PCCs contain the vitamin K dependent clotting factors (II, VII, IX, and X) and are derived from human plasma.

In a patient who presents with clinically significant bleeding or the need for emergent reversal for a procedure, laboratory or point of care (POC) testing should be conducted to assess if the oral anticoagulant is on board. A prothrombin time (PT) and an activated partial thromboplastin time (aPTT) should be ordered in all patients. The interpretation of these tests as well as the need to order additional coagulation tests will depend on the clinical situation and specific oral anticoagulant (Table 1).

Warfarin (trade name: Coumadin) is a vitamin K antagonist and exerts its action by decreasing the production of factors II, VII, IX, and X. Vitamin K may be administered to promote the synthesis of the aforementioned factors. PCC may also be administered. For major bleeding, vitamin K 5-10 mg IV as well as PCC should be administered due to the delayed onset of action of vitamin K (12 to 14 hours for IV vitamin K). PCC has an onset of action within 15 minutes and duration of action of 6-8 hours. PCC dosing is weight-based and dependent on the measured INR. More recent literature suggests that fixed-dosed PCC may be considered with similar results and a reduction in the amount of product and cost. These agents may be repeated in 12 hours if the INR remains elevated.

Dabigatran (trade name: Pradaxa) is an oral direct thrombin inhibitor. The reversal agent idarucizumab (trade name: Praxbind) was developed to directly reverse dabigatran. It is a humanized monoclonal antibody fragment (Fab) that neutralizes the anticoagulant effect of dabigatran and its metabolites by binding to these compounds with a higher affinity than that of dabigatran to thrombin. Idarucizumab was shown to completely reverse the anticoagulant effects of dabigatran in 88 to 98% of patients with a median time to hemostasis of 11 hours. Coagulation parameters (aPTT, TT, dTT, and ECT) were found to drop to within normal limits immediately after infusion of the medication. Dosing recommendations can be found in Table 2.

Apixaban (trade name: Eliquis), edoxaban (trade name: Savaysa), and rivaroxaban (trade name: Xarelto) are oral anti-Xa inhibitors. There are currently no direct reversal agents on the market. Andexanet alpha is a recombinant modified human factor Xa decoy protein that is currently under investigation. PCC can be considered for administration if the appropriate criteria are met (Table 3). The available clinical data for the use of PCC for reversal of the anticoagulant effects of DOACs is inconsistent. Three randomized studies have evaluated the effect of 4-factor PCC versus a comparator (placebo or 3-factor PCC) in human volunteers on coagulation laboratory values. The results demonstrated inconsistent correction of anticoagulant-induced laboratory abnormalities. Unfortunately, until more specific agents are approved in the US, PCCs are the only option for reversal at this time.

In the emergency department, reversal of oral anticoagulants is mandatory in patients with life-threatening bleeding. Emergency providers should rapidly assess for the presence of anticoagulants with a thorough history and laboratory data. Vitamin K and PCCs should be considered in patients on a vitamin-K antagonist or those on anti-Xa inhibitors while idarucizumab may be given to patients on dabigatran. Early administration of these agents is crucial in the management of these critically ill patients.

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<td>↑ Or ↔</td>
<td>↔</td>
<td>↔</td>
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<td>↑ Or ↔</td>
<td>↔</td>
<td>↑</td>
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<tr>
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<td>Variable sensitivity (non-linear)</td>
<td>Minimal effect</td>
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</tr>
<tr>
<td>Apixaban, edoxaban, or rivaroxaban</td>
<td>↑ Or ↔</td>
<td>↑</td>
<td>↑</td>
<td>↔</td>
</tr>
<tr>
<td>Variable sensitivity due to lack of standard reagent</td>
<td>Less sensitive</td>
<td>Dose dependent increase</td>
<td>Minimal effect</td>
<td></td>
</tr>
</tbody>
</table>

*Anti-Xa low molecular weight heparin will result more quickly than anti-Xa Fondaparinux (send out lab) #ECT is also a send out lab and will take several days to result.

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosing</th>
<th>Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitamin K</td>
<td>5-10 mg</td>
<td>12-14 hours</td>
</tr>
<tr>
<td>Idaracizumab</td>
<td>5 g (administered as two separate 2.5 g doses no more than 15 minutes apart)</td>
<td>Immediately</td>
</tr>
<tr>
<td>PCC</td>
<td>25-50 units/kg initially</td>
<td>15 minutes</td>
</tr>
</tbody>
</table>

Table 2: Reversal agent dosing

Table 1: Identification of on-board oral anticoagulants

Table 3: Criteria for PCC administration in patients on oral anti-Xa medications
History & Physical

While the classic triad for meningitis of fever, neck stiffness, and altered mental status has poor positive predictive value (only seen in 44% of cases), it continues to have clinical utility. The absence of all three symptoms in an immunocompetent adult nearly excludes meningitis altogether. When the triad is combined with headache, nearly all patients with meningitis (95%) have at least two of these four symptoms.

For physical exam maneuvers, Kernig's sign, Brudzinski's sign, and nuchal rigidity all have poor sensitivities and positive likelihood ratios less than 1, thus having very little impact on post-test probability of disease presence (see Table 2). Initially described in a 1991 study, the Jolt maneuver is positive if there is accentuation of a pre-existing headache with repetitive side-to-side head movement. It had a reported sensitivity of 100% in the initial publication. However, multiple studies since then have called these results into question due to an inability to replicate the outcomes, likely given that the original patient population inclusion criteria required both a new headache and fever. In summary of the literature, physical exam maneuvers are neither sensitive nor specific enough for meningitis, making the decision to perform a lumbar puncture even more challenging.

### Aseptic Meningitis

Based on lack of symptom constellation and physical exam findings in the second patient presentation, it is not surprising that an infectious etiology was not suspected. However, subacute neurologic decline should raise the suspicion for chronic meningitis caused by tubercle bacilli, syphilis, and fungi, as these agents may cause prolonged, indolent courses of disease progression without the typical signs and symptoms of meningitis. This frequently leads to multiple diagnoses and varying therapies as seen in the presented patient. Consequently, emergency providers should strongly consider performing a lumbar puncture in such cases of progressive neurologic decline.

The differential diagnosis for aseptic meningitis, defined as meningitis with a negative bacterial culture, is broad and requires additional CSF testing beyond the typical labs sent from the ED. While selection of more specific tests may require consultation with neurology, emergency providers are often the ones performing the initial lum-

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<table>
<thead>
<tr>
<th>Patient CSF Results</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Reference</strong></td>
</tr>
<tr>
<td>Opening Pressure</td>
</tr>
<tr>
<td>Total Nucleated Cells</td>
</tr>
<tr>
<td>% PMN</td>
</tr>
<tr>
<td>% Lymph</td>
</tr>
<tr>
<td>% other cells*</td>
</tr>
<tr>
<td>RBC</td>
</tr>
<tr>
<td>Glucose</td>
</tr>
<tr>
<td>Protein</td>
</tr>
<tr>
<td>Gram Stain</td>
</tr>
<tr>
<td>HSV PCR</td>
</tr>
<tr>
<td>Acid Fast stain</td>
</tr>
<tr>
<td>Cryptococcal ag</td>
</tr>
<tr>
<td>Culture - bacterial</td>
</tr>
<tr>
<td>Culture - fungal</td>
</tr>
<tr>
<td>Culture - AFB</td>
</tr>
</tbody>
</table>

*Mixed acute and chronic inflammation to include reactive lymphs and macrophages

**If it is a traumatic tap, decrease the amount of WBCs by 1 for every 1,000 RBCs.

***If it is a traumatic tap, decrease the amount of protein by 1.1 for every 1,000 RBCs.

Table 1: CSF results of the two patients.
bar puncture and should be mindful to collect as much fluid as possible for subsequent testing, as each unique test takes approximately 1 mL of fluid. In adults, up to 40 mL can be removed safely at one time even if for diagnostic rather than therapeutic purposes. Theoretically, the volume of fluid removed does not correlate with incidence of post-lumbar puncture headache, though the data behind this claim is not robust.

While lumbar puncture is the definitive test to identify meningitis, emergency providers must carefully consider the risks and benefits of performing this procedure in each case. A long-held belief in emergency medicine was that a head CT must be performed before every LP in order to assess a patient’s risk for herniation based on the presence of an intracranial mass lesion. However, the data behind this mantra is not conclusive. A 2017 meta-analysis reported an overall incidence of post-lumbar puncture herniation to be only 1.5%. Nonetheless, the majority of studies reporting these cases were published prior to CT availability, making it difficult to extrapolate that advanced imaging could have predicted neurologic decline in such patients. In addition, one study found that 20% of patients with pneumococcal meningitis who developed brainstem herniation had normal head CTs prior to the herniation. This suggests that head CT may not routinely predict patients who are at risk of herniation. Given this, providers should carefully consider the timing of LP and head imaging based on each patient’s clinical presentation. In the end, neither test should lead to a delay in antibiotic administration if clinical suspicion is high for bacterial meningitis.

Ultrasound as an Adjunct
A major barrier to performing a lumbar puncture in the emergency department is time. Lumbar punctures are “blind” procedures with variable success rates. The concern for failed attempts, particularly in patients with obesity, scoliosis or arthritis, can decrease the desire to proceed with the procedure. Fortunately, ultrasound to identify landmarks, particularly when palpation is challenging, is an excellent adjunct to increase the initial success rate and decrease the time needed to perform the procedure. In one study, landmarks were identified with ultrasound by 88% of providers in less than one minute and by 100% of providers in less than five minutes.

Treatment
For any patient with suspected meningitis, time to treatment has both prognostic and medicolegal implications. Thus, initiation of age-appropriate antibiotics and acyclovir when indicated should be done before necessary imaging and lumbar puncture if either of these is expected to cause a delay. Although there is no specific time to therapy goal, such as emphasized with sepsis, early initiation of empiric treatment is crucial. Nonetheless, there is a 20% reduction in organism identification with pretreatment of bacterial meningitis, so emphasis should be placed on timely lumbar puncture prior to treatment if at all possible.

Empiric antibiotic coverage should target the most common meningitis organisms for the patient’s age. In adults, typical regimens should include an anti-pneumococcal and anti-meningococcal medication with strong central nervous system penetration such as cefotaxime or ceftriaxone. Due to the rising prevalence of penicillin-resistant *strept pneumonieae* species, vancomycin should also be added. In patients over the age of 50, *listeria monocytogenes* is a frequent cause of meningitis and is not targeted by these medications. Therefore, ampicillin should be added to the above regimen. Consideration for *pseudomonas* coverage with cefepime or meropenem is indicated in immunosuppressed or recently hospitalized patients. Routine coverage with acyclovir is not mandated, but its empiric use should strongly be considered in immunosuppressed patients, those with reported seizure activity, or patients with stigmata of herpetic disease. Finally, regular use of anti-fungals is also not recommended unless the patient has risk factors causing immunosuppression. In summary, empiric treatment is best tailored to the specific patient and their risk factors with a focus on early administration of broad-spectrum treatment.

More controversial is the role of corticosteroids in the treatment of bacterial meningitis, as the body of evidence is conflicting. In children, there is some evidence of earlier resolution and decreased risk of long term hearing loss with administration of steroids before or at the time of antibiotics. However, the benefit is most prominent with *Haemophilus influenzae* infection, which is becoming increasingly rare with vaccination. In adults with pneumococcal meningitis, there is a 6% absolute mortality reduction with corticosteroid administration in high-income countries and no apparent adverse effects, although there are a number of confounders to the data analysis. It is hypothesized that high-dose dexamethasone (10 mg IV) should be replaced with low-dose hydrocortisone (50 mg IV) in patients with septic shock due to increased potential risk of adverse events, though this has yet to be studied. Other than pneumococcal meningitis, there is no clear mortality benefit to steroid administration. However, if emergency providers are starting empiric treatment without knowing the causative organism, steroids should be at least considered in suspected bacterial meningitis based on the most recent Cochrane Review.

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**Table 2: Characteristics of physical exam maneuvers in diagnosing meningitis.**

<table>
<thead>
<tr>
<th>Sign</th>
<th>Description</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>LR+</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kernig</td>
<td>Unable to fully extend knee when lying flat with hip flexed at a right angle</td>
<td>2.14%</td>
<td>97%</td>
<td>0.97</td>
</tr>
<tr>
<td>Brudzinski</td>
<td>Passive neck flexion leads to patient flexing at the hip</td>
<td>2.11%</td>
<td>97%</td>
<td>0.97</td>
</tr>
<tr>
<td>Nuchal rigidity</td>
<td>Discomfort or pain with neck flexion</td>
<td>13.30%</td>
<td>68-80%</td>
<td>0.94</td>
</tr>
<tr>
<td>Jolt</td>
<td>Accentuation of pre-existing headache with repetitive lateral head movement</td>
<td>21.64%</td>
<td>43%-88%</td>
<td></td>
</tr>
</tbody>
</table>
Caustic Ingestion
Continued from page 5

AnnALS of B Pod

all suspected cases of caustic ingestion may benefit from cross-section imaging in the emergency department.

The management of most caustic substance exposures in the emergency department is largely supportive. Providers should consider administering intravenous opioid agents for analgesia and crystalloid fluids for resuscitation. Current guidelines recommend against the use of emetics to expel the caustic substance or neutralizing xenobiotics due to the risk of potentially exacerbating the injury or causing an aspiration event. The use of prophylactic antibiotics is common practice, though there is not robust evidence for their use. In patients with symptoms concerning for vисcus perforation and consequent mediastinitis or peritonitis, however, empiric treatment with a third generation cephalosporin is recommended. Corticosteroid use following ingestion remains controversial, with some studies showing a decreased incidence of strictures and others showing no significant benefit.1,12 Expert opinion suggests that the empiric use of systemic glucocorticoids (e.g., dexamethasone 10 mg IV) is justified in patients with evidence of airway involvement to reduce edema and the risk of cardiopulmonary collapse.9 Proton pump inhibitors are sometimes used to de-acidify the gastric contents and are thought to reduce the likelihood of gastric ulceration or reflux, though their efficacy in reducing morbidity has not been demonstrated.13

Toxicologists recommend against the routine use of activated charcoal as it does not readily absorb high pH substances and itself carries the risk of aspiration and chemical pneumonitis. Similarly, the placement of blind nasogastric tubes is cautioned against due to the potential of perforating through injured, friable mucosa.

Ultimately, emergent surgical resection remains the definitive treatment of severe transmural, circumferential, or perforating injuries. Patients with severe injuries and damage to the swallowing apparatus may require prolonged parenteral nutrition; in these cases, naso-gastric feeding tubes should be placed intraoperatively under direct visualization to reduce the risk of perforation.

While long-term sequelae are uncommon for mild injuries (grade 1), more serious injuries are associated with various late complications. The most common delayed complication is stricture formation. Up to one-third of patients with esophageal burns (and virtually all patients with injuries of grade 2B or worse) will develop strictures.4 While not necessarily life-threatening, strictures may be associated with esophageal dysmotility, dysphagia, and gastric outlet obstruction depending on the location of the injury. Severely symptomatic patients may require endoscopic dilation. Strictures are prone to recurrence, however, and symptomatic relief from dilation is often temporary. More alarmingly, patients with severe injuries have increased risk of esophageal malignancy, with the incidence of esophageal neoplasms roughly 1000-3000 times higher than that of the general population.13

Though the ultimate cause of the above patient’s altered mental status was never fully elucidated, the concern for possible caustic ingestion highlights a unique pathology with which emergency providers must be familiar. Recognizing the hazards of and knowing the management of caustic ingestions are both essential parts of an emergency physician’s repertoire.

11. Anderson, K.D., Rouse, T.M., & Randolph, J.G. (1990). A controlled trial of corticosteroids in patients with severe injuries and damage to the swallowing apparatus may require prolonged parenteral nutrition; in these cases, nasogastric feeding tubes should be placed intraoperatively under direct visualization to reduce the risk of perforation.

CJD
Continued from page 3

tantly, 23% of prion-negative autopsies demonstrated evidence of a potentially treatable disease.9 These treatable conditions included immune-mediated conditions, neoplasms, infections, and toxic and metabolic encephalopathies. It is of paramount importance that patients presenting to the ED with dementia receive a thorough evaluation for treatable causes if such a workup has not yet been completed, as the treatable and non-treatable causes cannot reliably be distinguished on history and physical examination alone. One mnemonic to recall common treatable causes of dementia is “IMITT”; as these conditions can imitate the more common, incurable causes of dementia (Table 1).

All patients with suspected CJD should undergo CSF studies, MRI, and EEG. CSF studies are typically normal. Pleocytosis should raise suspicion for an etiology other than CJD. 14-3-3 proteins, a family of regulatory proteins that function in gene regulation, have a reported sensitivity of 85-95% in cases of autopsy-proven CJD.9 Nonetheless, elevated 14-3-3 is not specific for CJD (specificity 71%). Elevation of these proteins may also be present with neoplasia and paraneoplastic syndromes, subarachnoid hemorrhage, stroke, encephalitis, and metabolic encephalopathy.6

MRI brain with and without contrast has a reported sensitivity of 63-96% and a specificity of 92-93%.12 In general, MRI is abnormal in 80.9% of cases.12 MRI findings may be suggestive of a particular variant (e.g., pulvinar sign in variant CJD refers to bilateral FLAIR

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of CJD may be made through a combination of symptoms and testing, as highlighted by the WHO diagnostic criteria (Table 2, page 7). Additionally, there is a real-time quaking-induced conversion (RT-QuIC) CSF assay with a reported sensitivity of 77-97% and a specificity of 99-100% for the diagnosis of sporadic CJD. This is a send-out test at most institutions, but it does provide a near-definitive diagnosis if positive, as it was in the case highlighted above.

Neurologic consultation should be obtained with any case of suspected CJD. As emergency physicians, it is our responsibly to maintain a broad differential and thorough evaluation in patients with altered mental status, new-onset dementia, and unexplained neurologic deficits. A diagnosis of CJD or dementia should only be considered when all potentially treatable diagnoses have been sufficiently investigated and reasonably excluded.

Disposition

<table>
<thead>
<tr>
<th>Meningitis</th>
<th>Continued from page 13</th>
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</table>
| With bacterial meningitis, mortality is high even with appropriate antibiotic therapy - up to 20% with Streptococcus pneumoniae and 40% with Listeria monocytogenes or Neisseria meningitidis. Potential complications—including septic shock, adrenal insufficiency, and hydrocephalus—cause mortality to approach 70%. Thus, there is good argument for all patients with bacterial meningitis and patients already experiencing complications from meningitis of any form to be placed in the intensive care unit. The admitting team will vary based on institutional practice, but neurology and internal medicine are common admitting services. Ultimately, mental status, airway protection, and potential for acute clinical deterioration are all factors to keep in mind when deciding where in the hospital to admit these patients.

Summary

Emergency providers should maintain a high clinical suspicion for meningitis in patients with any combination of fever, altered mental status, headache, and neck stiffness, particularly given the poor sensitivity of history and physical examination findings. In patients with neurologic symptoms, providers should avoid anchoring on a single diagnosis such as pneumonia given the high percentage of patients with pneumococcal meningitis who present with a distinct infectious source. Similarly, in patients with a subacute neurologic decline, aseptic meningitis such as tuberculosis, syphilis, and fungal infections should be considered. Finally, while routine steroid administration is still up for debate, timely empiric antibiotic administration should be prioritized and can be life-saving.


Figure 1: Patient’s MRI showing signal abnormality involving the bilateral caudate nuclei (yellow arrow), bilateral frontal and temporal cortices (R>L) (red arrow).

Hyperintensities involving the pulvinar thalamic nuclei.

Periodic sharp wave discharges (PSWC) on EEG are highly suggestive of sporadic CJD. However, this characteristic pattern is found in only 37.5% of cases. EEG interpretation may be difficult as CJD occasionally presents as seizures and even status epilepticus. Neural tissue, obtained by brain biopsy or post-mortem, is the only means of definitive diagnosis of CJD. However, a probable diagnosis
History of Present Illness

A teenage male is brought into the emergency department after losing consciousness in the middle of a sporting event. The patient regained consciousness shortly thereafter, and denies chest pain or shortness of breath preceding the event. A thorough history reveals no prior past medical history or family history of early or unexplained death. Vital signs and serum troponin are within normal limits. The patient's EKG, however, demonstrates epsilon waves (Figure 1).

Epsilon waves are considered pathognomonic for arrhythmogenic right ventricular cardiomyopathy (ARVC). ARVC is a condition in which the muscular wall of the right ventricle is replaced with fibroadipose tissue. This tissue is predisposed to conduction abnormalities, leading to potentially fatal ventricular arrhythmias. These arrhythmias most commonly occur in the context of strenuous activity, and it is thought that the catecholaminergic surge associated with exercise or exertion potentiate the occurrence and propagation of tachyarrhythmias through the fibrofatty tissue by decreasing the refractory period and depolarization threshold of cardiomyocytes.

**EKG Features of ARVC**

- The epsilon wave (arrows) appears as a positive, low-amplitude impulse following the QRS complex. Localized QRS prolongation in V1 may be found in 24-70% of patients with ARVC.
- T wave inversions in the right-sided leads (V1-V3) may be identified in roughly 50% of cases, and constitute a major criterion for the diagnosis of ARVC in the absence of RBBB.

The epsilon wave is highly specific for ARVC, though it has poor sensitivity. It is estimated that only 30-33% of patients with ARVC are found to have an epsilon wave.1 In fact, the most common presenting rhythm of ARVC is ventricular tachycardia in the context of sudden cardiac death. Nonetheless, the epsilon is considered a major criterion for the diagnosis of ARVC.


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**List of Submitted B Pod Cases**

**Case**

- Bronchial stump dehiscence
- Fournier gangrene
- Hemolytic uremic syndrome
- Ruptured thoracic aortic graft
- Perinephric abscess
- Hypothermia
- Pneumoperitoneum/pneumomediastinum
- Endocarditis

**Case Physicians**

- Gottula
- Iparraguirre
- Miller
- O’Brien
- Shewakramani
- Spigner
- Lagasse
- Modi