**Submission template for Capstone Project Case Report**

<table>
<thead>
<tr>
<th>TITLE OF CASE</th>
<th>Budd-Chiari Syndrome</th>
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<tr>
<td>AUTHORS OF CASE</td>
<td>Leighanne Larson PAS2</td>
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<tr>
<td>SUMMARY</td>
<td>This patient is a 55 y.o. WM with a history of hypertension presented to the ED with fatigue, dry mouth, decreased appetite and confusion for 1 week. His wife stated that over the past few days he has become jaundice. On physical exam he appeared to be in mild distress, icteric, and irritable. His vitals were stable. Labs were drawn in the ED, which showed leukocytosis, hyponatremia, hypokalemia, elevated BUN/CR, elevated liver function tests, and possible portal venous thrombosis via abdominal ultrasound. The patient was admitted to the hospital with cholecystitis and elevated liver enzymes by the surgeon I was doing my rotation with. During the hospital stay a CT and ERCP was performed, which showed choledocholithiasis and a stent was placed in the common bile duct. The gallstone was not extracted due to being anticoagulated. The CT showed portal venous thrombosis, and hepatosplenomegaly. Repeat labs were drawn which showed improving leukocytosis, improving hypokalemia and worsening elevated liver enzymes. The patient was diagnosed with Budd-Chiari syndrome, which is a thrombus in the hepatic portal vein. The patient was anticoagulated with Heparin 2500U IV during his hospital stay at Memorial. He was later transfer to Beaumont Hospital in Detroit for evaluation by a specialist. He will likely be having a portosystemic shunt in the near future. He will continue to follow up with his specialist for further treatment.</td>
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<td>BACKGROUND</td>
<td>Budd Chiari syndrome is a rare disease (1-100,000) with similar symptoms to other medical problems. The condition presents similarly to cholecystitis, hepatic necrosis, hepatitis, etc. Although this condition is not common, skilled radiologists usually identify this on ultrasound or CT scan. These patients need prompt anticoagulation and transfer if necessary. Surgical and GI consults should be obtained for evaluation. It is common for these patients to receive a cholecystectomy during their hospitalization due to initial misdiagnosis. Budd Chiari should be included in the differential for patients presenting with elevated liver enzymes, abdominal pain, jaundice, and/or ascities. This diagnosis was not very well understood by the surgeons and physicians that were on this case. Our service was consulted for an emergency cholecystectomy; however, before the surgery we decided to discuss the case with radiology. After some discussion and education on hepatic venous thrombosis, we obtained a GI consult for an ERCP. The GI doctor made the final diagnosis of Budd-Chiari syndrome and the patient was transferred to Beaumont Hospital. Though the causes and treatment for Budd Chiari are similar to both pulmonary emboli and DVTs, the long-term effects of Budd-Chiari syndrome can be much more devastating. It is important to correctly diagnose these patients for several reasons: negative long term complications occur after misdiagnosis, cholecystectomy will not improve the condition, and early intervention can save lives.</td>
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<td>CASE PRESENTATION</td>
<td>M.G., a 55 y/o WM with a history of HTN presented to the ED with decreased appetite, dry mouth, fatigue and confusion. He states he has just been feeling “poorly” over the past week. He was able to work all week until Thursday when his fatigue would not allow him to work anymore (&quot;I just didn’t have the strength anymore&quot;). His wife states that over the past few days he had become jaundice.</td>
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**Past Medical Hx:**
Surgical Hx: Denies any surgical procedures

Meds:
1. Coreg 25 mg BID *
2. Lisinopril 2.5 mg BID *

*He states he has not been taking either of his medications recently. He is noncompliant.

Family Hx:
-Mother ↓, HTN, MI
-Father ↑, Alzheimer’s Dz

Social Hx:
-He is a janitor and lives at home with his wife.

ETOH/Drugs:
-Denies tobacco abuse.

-Admits to alcohol use and estimates his beer consumption to be a quart/week.

ROS:
-Gen:
  -Denies weight gain/loss
  -Denies recent fever, night sweats, chills, illness
  -⊕ Dry skin
  -⊕ Jaundice
  -⊕ Fatigue and weakness x 2 weeks
-Cardiac:
  -Denies chest pain, palpitations, light-headedness
  -⊕ bilateral pedal edema
-Resp:
  -Denies SOB
-GI:
  -Denies abdominal pain
  -Denies N/V/D
  -Denies hematochezia/melena/mucous in stools
  -⊕ decrease appetite x 2 weeks
-Genito/Urinary:
  -Denies dysuria, Hematuria, urgency, frequency
-Psych:
  -⊕ confusion
  -↑ irritability x 2 weeks

PE:
Vitals: Ht 5’8”, 228 lbs, BP 90/50, P 86, R 16, T 98.7 F, PaO2 98% RA
Gen: Well developed, well nourished male in mild distress, mild agitation, AAOx3
Skin: + icteric skin, good turgor, cap refill <2 sec
HEENT: PEERLA, EMOI, + scleral icterus, TM’s WNL, pharynx clear, no tonsilar hypertrophy, no lymphadenopathy, no sinus tenderness, neck supple, no JVD.
Cardiac: RRR w/o M/R/G
Resp: Lungs CTAB w/o R/R/W
Extremities: + ¼ bilateral pedal nonpitting edema. + bilateral posterior tibial and dorsalis pedius pulses noted.
Neuro: Flat affect. Speech is normal w/o slurring of words. Strength 5/5 throughout. Gait is stable. No tremors. CN II-XII intact.

INVESTIGATIONS

Labs:
**BMP** 125↓187↓177↑
3.1 ↓ | 26 | 1.7↑

Glucose: 127↑
Calcium: 8.7

**CBC**
- WBC: 18.3↑
- RBC: 3.55↑
- HGB: 10.4↓
- HCT: 31.2↓
- MCV: 88.0
- **Diff:**
  - Poly 79↑ (57-75%)
  - Band 10 (0-10%)
  - ANC 16.2↑ (1.5-6.6x 10-3)

**HFP**
- Albumin: 2.2↓
- Total Bil: 4.8 ↑(0.0-1.0)
- Direct Bil: 2.9 ↑(0.0-0.2)
- Indirect Bil: 1.9 ↑(0.0-1.0)
- AST: 84 ↑(0-50)
- ALT: 93 ↑(0-57)
- Alk Phos: 268 ↑(53-128)

**Abdominal US:**
- Fatty metamorphosis of the liver w/ abnormal branching soft tissue in the central portion of the liver, suggesting portal venous thrombosis or neoplastic invasion. Further evaluation w/ spiral CT scan of the liver is recommended.
- There is splenomegaly raising, possible portal hypertension.
- There is a contracted gallbladder w/ gallstones, mild gallbladder wall thickening, and a small amount of fluid. Common bile duct is 6 mm.
- Pancreas, abdominal aorta, and IVC are not visualized due to bowel.
- No renal mass or hydronephrosis.

**Consult for GI** was received. **Upon admission further labs were drawn:**

**CT of the abdomen w/o contrast:**
- Cholelithiasis within a thickened gallbladder wall.
- Hepatosplenomegaly
- Dilated right and left portal veins with intraluminal filling defects, which are nonenhancing and could suggest bland thrombus.
- Fatty masses in the right adrenal gland, malignant process is not ruled out.
- Minimal perihepatic and perisplenic and left pericolic gutter free fluid.
- Recommend MRI for further evaluation.

**ERCP:**
- Stent was placed in the common bile duct to provide biliary drainage. Sphincterotomy was not performed due to the need for anticoagulation.

**Assessment:** Choledocholithiasis
**Plan:** TURF to tertiary center due to the Budd-Chiari.

**DIFFERENTIAL DIAGNOSIS**

1. Acute cholecystitis
2. Choledocholithiasis
3. Portal Hypertension
4. Alcoholic Cirrhosis
5. Cytomegalovirus Infection
6. Hepatic Malignancy
7. Toxoplasmosis
8. Hepatitis
9. Coagulopathy (antithrombin III, protein S & C deficiency)
**TREATMENT**

M.G. was admitted to the hospital after his initial ER visit. His hospital stay lasted 5 days. Surgery and GI were consulted. After US, CT, and ERCP he was diagnosed with Budd-Chiari syndrome. Our treatment plan at Memorial was to anticoagulant him with Heparin 2500U IV, and to continue Coreg 25mg BID for HTN.

**OUTCOME AND FOLLOW-UP**

M.G was transferred to Beaumont Hospital in Detroit for further evaluation and treatment. Anticoagulation was continued with Heparin and Coumadin orally. He will follow up with his specialist at Beaumont Hospital as needed. I was unable to follow up with this patient after he left the hospital.

**DISCUSSION**

1. What is Budd-Chiari syndrome (Pathophysiology, causes)?
2. Are there risk factors for developing Budd-Chiari syndrome? Is there a genetic component?
3. Common signs/symptoms of Budd-Chiari syndrome.
4. How is Budd-Chiari syndrome diagnosed?
5. What are the treatment options for Budd-Chiari syndrome?

1. What is Budd-Chiari syndrome?

Budd-Chiari syndrome implies a thrombosis of the hepatic veins and/or the inferior vena cava. The pathophysiology of Budd-Chiari is an obstruction of the hepatic venous outflow tract results in increased hepatic pressure and portal hypertension. This venous stasis and congestion leads to hypoxic damage to adjacent hepatic parenchymal cells. The ischemic injury to the lining results in the release of free radicals, and oxidative injury to the hepatocyte occurs. Budd-Chiari syndrome probably represents a spectrum of disease caused primarily by a hypercoagulable state and having a varied presentation depending on the balance between rate of formation and the extent of the thrombus and the body's own rate of thrombolysis.

2. Are there risk factors for developing Budd-Chiari syndrome? Is there a genetic component?

Factors for predisposition to the development of Budd-Chiari syndrome:

1. Hypercoagulable states (75% of the patients):
   - Myeloproliferative disorders are the most common causes.
   - Polycythemia vera accounts for 10-40% of cases of the syndrome.
   - Antiphospholipid syndrome
   - Protein C and protein S deficiency
   - Antithrombin III
   - Factor V Leiden mutation.
   - Oral Contraception Use*
   - Cancer
   - Pregnancy

2. Uncommon causes (25%):
   - Tumor (Hepatocellular Carcinoma, Renal-cell Carcinoma, Adrenal Carcinoma)
   - Aspergillosis
   - Behcet syndrome
   - Trauma
   - Irritable Bowel Syndrome
   - Idiopathic

*The relative risk of hepatic-vein thrombosis among women who use OCP is 2.37%. Which is similar to their relative risk of stroke, MI and venous thromboembolism.

There has not been definitive data proving that there is a genetic predisposition to developing Budd-Chiari syndrome. However, there was recent study published by the International Journal of...
Gastroenterology and Hepatology showing evidence there may in fact be a genetic link.

- This study investigates expression of a set of genes involved in the course of chronic liver disease. Thirty-five genes were selected to be analyzed. Thirteen of the cases were Budd-Chiari syndrome, ten were normal, and thirteen were cirrhosis.
- Results: Fourteen genes were increased in Budd-Chiari syndrome vs. a normal liver.
- Conclusion: Budd-Chiari syndrome displays a specific gene expression profile that is different from that of a normal liver.

3. Common signs/symptoms of Budd-Chiari syndrome.

**Clinical Manifestations:** 4 different subtypes

1. **Fulminant:** presents with hepatic encephalopathy within 8 weeks after development of jaundice (uncommon) 8.

2. **Acute:** short duration, intractable ascites, and hepatic necrosis without formation of venous collaterals. Thrombosis is only seen in 1/3 of patients in acute Budd-Chiari syndrome 9. The acute syndrome is associated with extensive blockage of the major hepatic veins, resulting in congestive liver cell necrosis 2.

3. **Subacute** (most common): insidious onset. Ascities and hepatic necrosis may be low at this time.

4. **Chronic:** manifested by complications of cirrhosis. The chronic syndrome is characterized by portal hypertension and is associated with a variable abnormal vascular anatomy. The causes of the chronic syndrome are not clear 1.

**Signs/Symptoms of Budd-Chiari syndrome:**

The classic triad of Budd-Chiari syndrome is abdominal pain, hepatomegaly and ascities 8. Nausea, vomiting and jaundice are more frequent in fulminant and acute forms. Splenomegaly and esophageal varices are seen with chronic forms 9. These patients will usually present to the emergency department or to their primary care provider with signs/symptoms similar to acute cholecystitis. “Most often, patients receive a diagnosis of cholecystitis because of the combination of abdominal pain and an US examination that shows gallbladder wall thickening” 8. “It is essential to consider a diagnosis of the Budd-Chiari syndrome in all patients with thrombophilia who have ascities, upper abdominal pain, or abnormalities on liver function tests” 8.

4. How is Budd-Chiari syndrome diagnosed?

**Laboratory analysis:**

- Complete metabolic profile and basic metabolic profile can be drawn as a baseline.
- AST and ALT may be 5x the upper limits of normal in acute Budd-Chiari syndrome 8.
- Elevation in alkaline phosphatase and bilirubin levels are also increased 8.

**Diagnostic evaluation:**

- Doppler US of the liver (85% sensitivity and specificity) is the technique of choice for the initial diagnosis 8.
- CT with contrast can also be diagnostic.
- If TIPS procedure is considered at initial diagnosis of Budd-Chiari syndrome is diagnosed, do a contrast CT of the abdomen to map the venous system.
- If paracentesis is performed, the ascites fluid albumin gradient will be elevated, with the total protein level in the ascitic fluid greater than 2.5 g per dL 8.

The diagnosis of Budd-Chiari syndrome is confirmed by a “spiderweb” pattern on hepatic venography 8.

5. What are the treatment options for Budd-Chiari syndrome?

The main goal of treatment is to alleviate hepatic congestion, improving hepatocyte function and
resolution of the portal hypertension \(^2\). Percutaneous and endovascular procedures, when performed in properly selected patients, may be more effective than medical treatment methods for preserving liver function and stopping the disease process \(^2\). Selection of treatment is based on the degree of hepatic injury liver biopsy results, potential for parenchymal recovery, and preserved hepatic function \(^5\). There are two different types of treatment options for Budd-Chiari syndrome: medical and/or surgical management.

**Options for medical management**: 

Goal of medical management is to control further development of ascites, the use of anticoagulation therapy to prevent further extension of thrombus, and the treatment of the underlying cause. Nonsurgical alternatives have limited long-term outcome \(^6\).

- Managing the ascites: restricted sodium intake to 90mmol per day and administration of spironolactone and durosemide to achieve a negative sodium balance. Also, the use of Paracentesis and IV albumin is used when refractory to diuretic therapy.
- Anticoagulation: heparin inpatient and oral coumadin for long term anticoagulation (INR between 2.0-2.5 is indicated).
- Thrombolytics (TPA and urokinase) can be considered for patients with acute Budd-Chiari syndrome, especially if it is found to be a fresh thrombus via ultrasound or CT scan.

**Surgical Management** \(^4, 6, 8\)

Side-to-side portacaval shunt (SSPCS) remains the gold standard for achieving good long-term results \(^4\). Shunting is recommended in cases of preserved hepatic function and architecture. In cases of established cirrhosis or fibrosis, or for patients with defined hepatic metabolic defects liver transplant is the treatment of choice \(^6\). Stents can also be used to maintain the patency of surgical shunts that have been made to bypass the obstruction to the blood flow from the splanchic circulation to the IVC via the liver \(^4\). The 5 year survival rate after surgical shunting ranges between 75-94% and is higher if the vena cava is not occluded \(^8\). Portosystemic shunt relieves sinusoidal hypertension, can reverse hepatic necrosis and prevents cirrhosis \(^8\).

Liver transplant is indicated in patients with chronic Budd-Chiari syndrome who have cirrhosis and patients with a failed portosystemic shunt procedure \(^4\). The 5 year survival rate among patients who undergo liver transplant for Budd-Chiari syndrome is 95%. Indications for transplant include: hepatic failure, cirrhosis, and the failure of the portosystemic shunt \(^8\). Liver transplant theoretically corrects the underlying thrombophilia (if there is one), so not all patients may not require long-term anticoagulation \(^8\).

Angioplasty has a high success rate in Budd-Chiari syndrome due to congenital obstruction. For restenosis, balloon dilation can be performed. TIPS (Transjugular Intrahepatic Portosystemic Shunt) procedure can be performed in Budd-Chiari syndrome patients to improve the clinical condition while waiting for liver transplant \(^4\). Recommended in patients with the acute form of Budd-Chiari syndrome if thrombolytic therapy has failed. TIPS serves as a bridge to a liver transplant \(^8\).

**Recent study about the surgical treatment of Budd-Chiari syndrome** \(^7\):  
The aim of this study was to assess the factors which affect survival in adults with Budd-Chiari syndrome. One hundred and twenty patients admitted from 1970-1992 of whom eighty-two were treated with surgical portosystemic shunts and thirty-eight received only medical therapy. Four factors were found to be inversely and independently related to survival: age, response of ascities to diuretics, Pugh score, and serum creatinine. These four factors allowed differentiating patients from a good outcome (5-year survival 95%) from those with a poor outcome (5-year survival of 62%). **Outcome** \(^7\): there was no statistically significant and independent influence of surgical portosystemic shunts on survival. Increased survival in recent years is consistent with improved management of hypercoagulable states as well as improved general care. Therefore, they recommend surgical shunting should be restricted to management of refractory ascites or variceal bleeding in patients with otherwise good prognostic factors.
LEARNING POINTS/TAKE HOME MESSAGES 3 to 5 bullet points

- Budd-Chiari syndrome is a rare complication of a portal venous thrombosis, which leads to liver failure.
- Signs and symptoms of Budd-Chiari syndrome are similar to acute cholecystitis or hepatitis.
- Treatment is with prompt anticoagulation and surgery if indicated.
- The definitive treatment of Budd-Chiari syndrome is a liver transplant.

REFERENCES


Date: Leighanne Larson PA-S

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Author's last name and date of submission, eg,

Smith_June_2008.doc