International Journal of Science and Research (IJSR) ISSN: 2319-7064

Impact Factor 2024: 7.101

Budd Chiari Syndrome: A Case Study

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Abstract: Budd- chiari syndrome is a rare but serious vascular disorder characterized by obstruction of hepatic venous outflow at the level of hepatic veins, inferior vena cava, or both. This obstruction leads to increased hepatic sinusoidal pressure, hepatomegaly, ascites and progressive hepatic liver dysfunction. The condition may be acute, subacute or chronic and its etiology involves prothrombic states, inherited thrombophilias etc. Diagnosis relies on clinical suspicion supported by doppler ultrasonography, computed tomography or magnetic resonance imaging showing impaired venous drainage. Management includes anticoagulation therapy, endovascular interventions, transjugular intra hepatic portosystemic shunt and in advanced cases, liver transplantation.

Keywords: hepatic vein, ascites, cirrhosis, TIPS (Transjugular intra hepatic portosystemic shunt)

1. Introduction

BUDD CHIARI SYNDROME is defined as the obstruction of hepatic venous outflow at the level of hepatic veins inferior vena cava, or the both, resulting in hepatic congestion, impaired sinusoidal drainage and progressive liver dysfunction.

2. Classification

By Cause:

- PRIMARY BCS: Venous anomalies such as thrombosis, web, endophlebitis.
- SECONDARY BCS: Initial lesions or external compression of the vein from nearby structure (tumor, abscess, cyst)

By Clinical Course:

- ACUTE: Symptoms develop rapidly over a few weeks, often with severe pain, ascites and hepatomegaly
- SUBACUTE: Gradual onset, developing over up to three months with collaterals.
- CHRONIC: Symptoms develop slowly, leading to complications of cirrhosis.
- FULMINANT: A rare, severe presentation with rapid hepatic failure.

By Location of Obstruction:

- HEPATIC VEIN OBSTRUCTION
- INFERIOR VENACAVA OBSTRUCTION
- COMBINED OBSTRUCTION

3. Etiology

Inherited

- Antithrombin III deficiency
- Prothrombin c deficiency
- Protein S deficiency
- Factor v leiden mutation
- Prothrombin mutation

Acquired

- Myelo proliferative neoplasms
- Polycythemia rubra vera
- Essential thrombocytosis
- Paroxysmal nocturnal hemoglobinuria

Infections

Aspergillosis

· Antiphospholipid syndrome

- Filariasis
- Hydatid cyst
- Liver abscess
- Pelvic cellulitis
- Schistosomiasis
- Syphillis
- Tuberculosis

Malignancies:

- Adrenal carcinoma
- Hepatocellular carcinoma
- Leiomyosarcoma
- Lung cancer
- Myxoma
- Renal carcinoma
- Rhabdomyosarcoma

Miscellaneous:

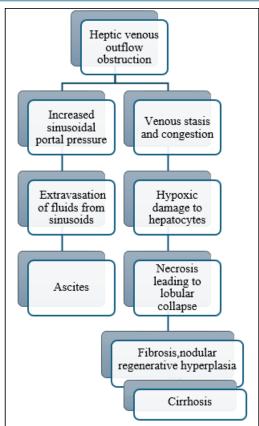
- · Behcets disease
- Celiac disease
- IBD
- Sarcoidosis
- Trauma to abdomen or thorax

4. Pathophysiology

Volume 14 Issue 12, December 2025
Fully Refereed | Open Access | Double Blind Peer Reviewed Journal
www.ijsr.net

International Journal of Science and Research (IJSR)

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5. Case Study of Mr. Raj

Mr. Raj, 42 years old admitted to the hospital with the complaints of right upper quadrant abdominal pain, vomiting and nausea and lower limb edema. Following a comprehensive investigation and examination he was diagnosed as budd chiari syndrome (hepatic vein obstruction). He is conscious and oriented. His vitals are as follows:

Temperature: 99.8 F Pulse :82 beats/min

Respiration: 24 breaths / min Blood pressure: 110/70 mm of Hg

The ultrasonography reveals collaterals and narrowing of right hepatic vein with grade 1 esophageal varices.

6. Signs and Symptoms

Book Picture	Patient Picture
*Abdominal pain and distention	Present
*Anorexia	Present
*Jaundice	Present
*Ascites	Present
*Collaterals	Present
*Pedal edema	Present
*Hepatomegaly	Present
*Anorexia and vomiting	Present
*Mild fever	Present

7. Diagnosis

- a) History collection
- b) Physical examination
- c) Liver function test
 - Bilirubin increases

- Transaminase increases
- Serum albumin decreases
- High SAAG (serum albumin ascites gradient)
- d) CECT (contrast enhanced computed tomography) abdomen
- e) MRI
- f) Venography

8. Management

- 1) Systemic thrombolytic therapy
- 2) Trans luminal angioplasty
- 3) Endovascular stenting
- 4) TIPS (Transjugular intrahepatic portosystemic shunt)
- 5) Liver transplant

9. Complications

Hepatic and Gastrointestinal Complications:

- · Portal hypertension
- Ascites
- Variceal haemorrhage
- · Hepatic encephalopathy
- Cirrhosis

Renal and Systemic Complications:

Hepatorenal syndrome

- Peritonitis
- Pulmonary embolism
- · Hypercoagulable state

Prognosis:

- Untreated:80 % mortality
- With intervention: 5 year survival upto 87 %
- Liver transplant: 70% 5 year survival

10. Nursing Management

Assessment:

Complete health history

Abdominal assessment includes

- Abdominal distention, ascites, splenomegaly, hepatomegaly
- Skin and mucous membrane-jaundice, peripheral edema
- Respiratory rate

Nursing Diagnosis:

- 1) Excess fluid volume related to hepatic venous obstruction as evidenced by abdominal distention.
- 2) Acute pain related to hepatic congestion as evidenced by verbal reports of pain.
- 3) Imbalanced nutrition less than body requirement related to anorexia as manifested by poor intake.
- Ineffective peripheral tissue perfusion related to altered blood flow due to thrombosis as evidenced by delayed capillary refill.
- 5) Risk for bleeding related to impaired hepatic synthesis of clotting factors and portal hypertension.

Planning:

 Assess the abdominal girth and reduced or stabilize abdominal girth within 24-48 hours.

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Fully Refereed | Open Access | Double Blind Peer Reviewed Journal

www.ijsr.net

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- Monitor the progression of pain level.
- Assess heart rate and respiratory pattern frequently to detect early hypoxia due to diaphragmatic pressure.
- Monitor liver function test including ALT, AST, ALP and Bilirubin to prevent further complications.
- Provide small, frequent meals and meet atleast 50% of daily caloric needs to prevent further malnutrition and supports liver recovery.
- Monitor capillary refill.
- Encourage in light activities to reduce venous stasis
- Monitor the coagulation status.

Implementation:

- Measure abdominal girth and monitor intake and output.
 Restrict sodium intake, administer diuretics.
- Administer diuretics and prescribed analgesics
- Elevate the head of the bed to ease breathing when ascites present
- Monitor daily food intake, measure weight, monitor the lab values of electrolytes.
- Educate about signs that need immediate reporting
- Encourage family involvement and use relaxation technique
- Encourage in follow up imaging studies

Evaluation:

- The patient shows decreased abdominal girths as compared to previous measurements.
- The patient reports a decreased intensity in pain
- The patient reports improved apetite and decreased nausea.
- The patient maintains a stable PT/INR and platelets level.

11. Conclusion

Budd chiari syndrome is a rare but serious disorder caused by obstruction of hepatic veins, leading to impaired hepatic blood flow, hepatomegaly, ascites, and progressive liver dysfunction. Early recognition of symptoms such as abdominal pain, ascites and jaundice is essential to prevent complications.

Nursing care plays a critical role in monitoring fluid balance, managing pain, optimizing nutrition, preventing bleeding, and promoting tissue perfusion.

Though timely intervention and interdisciplinary collaboration helps to minimize complications, improve quality of life and support better clinical outcomes.

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