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**Case Study**

# A rare case report on Budd-Chiari syndrome: A role of clinical Pharmacist's interventions for achieving better therapeutic outcomes

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## ABSTRACT

This report describes a case of Budd-Chiari syndrome and a role of clinical pharmacist for optimization of therapeutic outcome in it. A 20-year-old female patient presented with complaints of bi-lateral upper and lower limb swelling along with abdominal distension, vomiting and generalized body ache since last 15 days. CT-scan confirmed USG findings and showed complete thrombosis of all three hepatic veins. This confirmed the diagnosis of Budd-Chiari syndrome. Symptomatic relief was provided to the patient through anticoagulant and diuretics therapy. In this case, a patient was prescribed with multiple medications like digoxin, diuretics, propranolol and others. Whenever, these medications are administered together, they may lead to severe drug interactions and further clinical consequences like hyperkalemia and digoxin toxicity. In this case, there is a crucial role of clinical pharmacist to identify the important drug interactions and monitor the patient to prevent any clinical complications. Also, clinical pharmacist may help to optimize the therapeutic outcome by conducting detailed patient counselling.

**Keywords:** Budd-Chiari syndrome, hepatic vein, digoxin, anticoagulant, diuretics

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## INTRODUCTION

Budd-Chiari syndrome is a rare condition induced by thrombotic or non-thrombotic occlusion of major hepatic veins or Inferior Vena Cava (IVC) or both at or near the level of the hepatic vein ostia.[1]Hypercoagulable state is more common in female patients compared to male. In majority of adult patients, primary myeloproliferative diseases are the leading cause forBudd-Chiari Syndrome (BCS). However, BCS is less commonly observed in paediatric patients and usually caused by hypercoagulable state, IVC membranous web, infection, Neoplasia, trauma, total parenteral nutrition, etc.[2]

Clinical features depicting acute form are nausea, vomiting, and severe pain in right hypochondrium, enlargement of right liver, hypotension and often death. Chronic features that gradually lead to liver failure are hepatic cirrhosis, jaundice, ascites, oesophageal varices and infection.[3] Symptoms usually develop gradually over weeks or months. For the confirmation of this syndrome; hepatic vein catheterization, Doppler Ultrasonography and liver biopsy can be performed to detect narrowed or blocked veins.[1,3]

Patients with BCS must be initially treated for ascites and varices. Recommended stepwise therapeutic regimen is medical treatment followed by angioplasty, Transjugluar Intrahepatic Portosystemic Shunt (TIPS)/Direct Intrahepatic PortocavalShunt (DIPS), liver transplant. Correction of precipitating factors such as GIT bleed, infection, and hypokalaemia is necessary. Prophylactic dietary recommendations for vegetarians and vegans include low-sodium intake (<4g) to manage ascites. Anticoagulant therapy, thrombolytic therapy, radiologic Intervention, diuretics therapy are recommended treatment options.

## CASE PRESENTATION

A 20-year-old female patient was admitted to tertiary care hospital with the complaints of ascites, generalised body ache, vomiting, pedal edema and burning micturition since last 10 days. Patient was previously admitted for the past medical history of sickle cell trait, Cerebrovascular accident, dilated cardiomyopathy, portal hypertension and patient had received 1 unit of packed cell volume blood transfusion five months ago.

Physical examination revealed that patient had abdominal distension, everted umbilicus, dilated veins with tender hepatomegaly and splenomegaly. On percussion, patient had horseshoe shaped dullness and shifting dullness s/o ascites. Patient had bilateral crepitation more on the right side s/o pleural effusion.

Laboratory findings showed normal level of AST, ALT and serum creatinine. There was a mild elevation in total bilirubin 1.5mg/dl (0-1.4 mg/dl), direct bilirubin 0.5mg/dl (0-0.3 mg/dl) and indirect bilirubin 0.6 mg/dl (0.2-1.2 mg/dl). Haemoglobin concentration was found to be 10.3 g/dl (11.7-15.5 g/dl) s/o hypochromic microcytic anaemia. Total Leucocytes and platelets were in normal range. International Normalized Ratio (INR) was 1.714 (0.8-1.2) and erythrocyte sedimentation rate was 41mm/hr(0-22 mm/hr).

USG findings showed ascites, congestive hepatomegaly, splenomegaly, dilated portal veins, peri-splenic and peri-portal collaterals. Doppler examination suggested thrombus occluding hepatic vein with increased flow in the portal and splenic veins with few collateral veins. Thus, all the findings were s/o BCS.

Patient was provided symptomatic relief on the day of 17-4-18. Tab. Spironolactone + furosemide (20/50 mg) (1-1-0) for pedal edema, Tab. Leviteracetam (500mg) (1-0-1) as an anti-convulsant, Tab. Acenocoumarol (2mg) (0-0-1) as an anticoagulant, Tab. Vitamin B6 (0-1-0), Tab. Propranolol (10mg) (1-0-1) and Tab. Ramipril (2.5mg) (0-0-1) were given for portal hypertension, tab. Digoxin (0.25 mg) (1-0-0) as an antidysrhythmic agent and syrup Lactulose (18 cc) (HS) for prevention of hepatic encephalopathy; were prescribed. All medications were well tolerated by the patient hence they were continued for the next day, too. On 19-4-18, patient was discharged pertaining to improved condition with discharge medications: Tab. Multivitamin (1-0-1), Tab. Spironolactone + furosemide (20/50mg) (1-0-0), Tab. Leviteracetam (500mg) (1-0-1), Tab. Acenocoumarol (2mg) (0-0-1) and Tab. Digoxin. (0.25mg) (1-0-0)

## DISCUSSION

Different case studies were reviewed and compared with our case report for better understanding and interpretation.[4,5,6] An another case report identified in Afro-Brazilian female patient was studied thoroughly for review purpose.[7]

In that case, 25 year old female patient came with the complaints of ascites, dyspnea after exercise, edema in abdominal region and swelling in legs. Similarly, our patient also had complaints of ascites and pedal edema. Brazilian patient had a 5 year old history of asymmetric recurrent migratory arthritis in her wrist and ankles, recurrent painful ulcers and lesions in oral as well as vagina; painful transient

erythemanodosum of forearm and legs suggestive of Behcet's disease. Patient was also occasional smoker and moderate alcohol drinker. Contrary, our patient had past medical history of sickle cell trait, Cerebrovascular accident (left MCA territory infract), dilated cardiomyopathy, portal hypertension and patient had 1 unit of packed cell volume blood transfusion 5 months ago.

Examination findings indicate that in this patient Budd-Chiari syndrome has occurred as a complication of Behcet's disease. Whereas, physical examination of our patient depicted the cause of Budd-Chiari syndrome in this patient due to reduced blood flow by sickle cells.

Laboratory findings of reviewed patient confirmed hypochromic and microcytic anemia, high hemosedimentation velocity and C-reactive protein, serum ascites albumin gradient greater than 1.1 and positive skin pathology test. Similarly, our patient also had hypochromic microcytic anemia. Other biochemistry tests were normal.

Initial treatment approach was to provide symptomatic relief with added antibiotic therapy and use of angiotensin-converting enzyme inhibitors, diuretics and a methylprednisolone pulse therapy followed by oral corticosteroids, azathioprine, colchicine, anticoagulants and methotrexate (replacing the colchicine at hospital discharge for better convenience of administration). Similarly, our patient was also provided symptomatic relief through spironolactone, anti-convulsant and anticoagulant.

We also found another case report on BCS, which was caused by latent polycythemia vera accompanying Factor V Leiden mutation.[8] The clinical presentation in this case was quite similar to that of our case and mainstay therapeutic approach was anti-thrombotic therapy.

The distinguish feature of our study is the involvement of clinical pharmacist in the identifications of severe drug interactions and monitoring g of the clinical parameters on the basis of that. The significant role of clinical pharmacist in management of BCS, can be understood by following pharmacists' interventions.

## PHARMACIST INTERVENTION

Pertaining to the complaints of patient about generalised weakness, drug synergism of propranolol and digoxin was intended for treating the patient. Both these drugs improve exercise tolerance in-patient with abnormal ventricular function.

Due to polypharmacy; chances of drug interactions were quite high. Hence, patient was monitored closely. Possible drug interactions could be:

1) digoxin + propranolol: it is a serious interaction as either drug can increase toxicity of the other and can increase risk of bradycardia.[9]
2) spironolactone + digoxin: spironolactone increases the half-life of digoxin by activating P-glycoprotein (MDR1) efflux transporter and also decreases its renal clearance resulting into subsequent toxicity and false digoxin assay results. Thus, careful monitoring is required during coadministration.[9]
3) propranolol + digoxin: pharmacodynamic synergism enhances risk of bradycardia and hyperkalemia.[9]
4) Spironolactone shows pharmacodynamics synergism with propranolol, ramipril and digoxin enhancing risk of hyperkalaemia. Hence, adequate electrolyte monitoring is required.[9]

Common drugs used for treating BCS are digoxin, spironolactone, propranolol and Ramipril. All these drugs tend to alter the mechanism of sodium/potassium ATPase pump that can lead to hyperkaliaemic state. Symptoms associated with hyperkalaemia are muscle cramps, generalised weakness, tingling, dyspnoea and chest pain. Thus, it is necessary to monitor potassium levels due to polypharmacy.[10]

As the patient was receiving anticoagulant therapy, frequent monitoring of prothrombin time and activated partial thromboplastin time was important and no significant alterations were observed in these parameters.

## HIGHLIGHTS OF IMPROTANT PATIENT COUNSELLING POINTS

1. About the disease

In the simplest way, patient was explained about the disease that the liver is basically responsible for many vital physiologic processes. Blood flowing through the liver transports toxins for excretion. When blood cannot flow out freely from the liver, blood pressure rises locally, leading to blood clots over there. In addition, there might plasma leakage followed by accumulation in abdomen leading to ascites.

2. Dietary modification[11,12]

- Consumption of low sodium diet that includes no more than 1500-2400 mg of sodium per day.
- Avoid consumption of total and saturated fatty foods such as ghee, butter, dairy

products; sugar, aerated drinks, and refined carbohydrates. Consume higher quantity of fruits, vegetables, whole grains, fish, poultry, and low-fat dairy products.

3. Lifestyle modifications[11]

- *Maintain an appropriate body weight*: The target body mass index is $<25\ kgm^{-2}$, Waist circumference should also be maintained at an appropriate level, abdominal girth should be maintained < 70 cm for women and <80 cm for men (patient's abdominal girth was 67 cm).
- *Exercise*: Periodic exercise of 30 min or longer daily aerobic exercise, at a moderate intensity, should be practiced.
- *Quit smoking*: Smoking (including passive smoking) should be avoided as it is a strong risk factor for cardiovascular disease and also affects blood pressure.
- Manage stress: patient as well as family members were counselled to keep the surrounding stress-free. Practice relaxation by performing moderate yoga, deep breathing and slow walking.

## CONCLUSION

In the therapeutic management of a critical case like BCS, there is vital role clinical pharmacist to optimize the therapeutic effect by identifying the major drug-drug interactions, drug synergism and medication error. By doing proper drug related monitoring, the drug related any possible harm to the patient can be priory prevented. Also, by detailed patient counselling related to disease, diet and life-style modification; a clinical pharmacist can play a significant role in achieving better therapeutic outcome in a severe and rare disease condition like BCS.

## REFERENCE


1. Emedicine.medscape.com. Budd-Chiari Syndrome: Practice Essentials, Background, Pathophysiology.https://emedicine.medscape.com/article/184430-overview(cited 10 January 2019).
2. Misra V. The Budd-Chiari Syndrome in a Child: A Case Report and Review of the Literature. Journal of clinical and diagnostic research 2012.
3. Sriram B. SRB's manual of surgery, 5th ed. Jaypee Brothers Medical Publisher (P) Ltd.; 2016.
4. Everan abut et al. A case of budd-chiari syndrome secondary to multiple thrombogenic condition: a case report and review of literature. Turk J Gastroenterol 2004; 15 (2): 100-103
5. NaofumiChinen et al. A Case of Acute Budd-Chiari Syndrome Complicating Primary Antiphospholipid Syndrome Presenting as Acute Abdomen and Responding to Tight Anticoagulant Therapy.Hindawi Publishing Corporation Case Reports in Rheumatology Volume 2016
6. Ana Catalina Ramírez et al.Budd Chiari Syndrome: Three Case Reports and a Literature Review.Rev Col Gastroenterol 2014; 29 (3).
7. Carvalho D et al. Budd-Chiari syndrome in a 25-year-old woman with Behçet's disease: a case report and review of the literature. Journal of Medical Case Reports 2011;5(1).
8. Abut E et al. A case of Budd-Chiari syndrome secondary to multiple thrombogenic conditions: a case report and review of literature. Turkish Journal of Gastroenterology 2004 Jun;15(2):100-3.
9. Reference.medscape.com. Drug Interactions Checker - Medscape Drug Reference Database. https://reference.medscape.com/drug-interactionchecker (cited 3 january 2019).
10. Ben Salem C et al. Drug-Induced Hyperkalemia. Drug Safety 2014;37(9):677-692.
11. Teichgräber U et al. Effect of respiration, exercise, and food intake on hepatic vein circulation. Journal of Ultrasound in Medicine 1997;16(8):549-554.
12. Cundiff D et al. Diet as prophylaxis and treatment for venous thromboembolism 2010.