



# ORIGINAL RESEARCH PAPER

# Paediatric Medicine

## UNRAVELING ABDOMINAL PAIN: A CASE OF BUDD – CHIARI SYNDROME IN A PEDIATRIC PATIENT

**KEY WORDS:** Budd chiari syndrome, Thrombocytopenia , IVC stenting

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### ABSTRACT

Budd-Chiari Syndrome (BCS) is a rare disorder resulting from obstruction of the hepatic veins or inferior vena cava, leading to liver congestion and potential failure. This case report highlights a 12-year-old male who presented with intermittent abdominal pain for one month, progressively worsening in intensity. Clinical examination revealed hepatomegaly, and further diagnostic workup, including Doppler ultrasound and CECT imaging, confirmed the presence of Budd-Chiari Syndrome caused by inferior vena cava stenosis. Endovascular intervention with IVC stenting was performed, leading to symptomatic improvement. This case underscores the importance of considering BCS in pediatric patients presenting with unexplained abdominal pain and hepatomegaly. Early diagnosis and intervention are crucial for improving outcomes in this rare but potentially fatal condition.

### INTRODUCTION

Budd Chiari Syndrome (BCS) is a rare condition, brought on by an obstruction of the inferior vena cava or hepatic vein. Depending on where the obstruction occurs, it can take two different forms: obstruction of the hepatic section of the inferior vena cava (IVCO) and primary hepatic vein obstruction (classical BCS). While the IVCO type is rarely documented in Western nations, it is ubiquitous in Asia and Africa(1). Clinical manifestations include fulminant acute forms, subacute forms, and chronic (decompensated) forms. Abdominal discomfort, hepatomegaly, ascites, and pedal edema are typical symptoms.

We report a 12 year old child who presented with abdominal pain on and off since 1 month with hepatomegaly on palpation, with further workup diagnosed to have Budd chiari syndrome.

### Case Report

A 12-year-old male, born to a non-consanguineous marriage, presented with complaints of intermittent abdominal pain for the past 1 month, which had increased in intensity over the last four days. The pain was not associated with vomiting or diarrhea. He did not have any comorbidities in the past. His birth and developmental history were insignificant.

On general examination, the patient was afebrile, with a pulse rate of 90/minute, respiratory rate of 24/minute, and blood pressure of 100/70 mm Hg. There were no signs of anemia, jaundice, clubbing or pedal edema. Anthropometric measurements were normal. On abdominal examination, mild distention was noted, and the liver was palpable 5 to 6 cm from the right costal margin with a firm consistency and smooth surface with mild tenderness over right hypochondrium region. Viral hepatitis, hepatitis due to tropical fever and autoimmune hepatitis were considered.

Investigation showed the following: Hemoglobin-12.7gm/dl; Total count-6867 cells/cubic mm Platelets- 0.84lakhs/cubic mm; Peripheral smear showed thrombocytopenia. Total Bilirubin- 0.5mg/dl; Transaminases: normal; ALP- 180 U/L; S. Albumin 3.9gm/dl; C Reactive Protein- 0.7 mg/dl; Creatinine- 0.3 mg/dl. Infective work up such as malaria antigen, Scrub typhus IgM and Dengue serology IgM were negative. As per Paediatric haematologist advice, work up for thrombocytopenia done. Reticulocyte count was normal. Serum Ferritin and serum Vitamin B12 levels were normal. Prothrombotic work up (Protein C, protein S, Antithrombin III, factor V Leiden mutation, homocysteine levels) done were normal. Hence possibility of spurious thrombocytopenia secondary to peripheral destruction was considered and folic

acid was started.



**Fig 1-** Contrast CT abdomen showing features of Budd chiari syndrome

USG abdomen showed hepatomegaly, splenomegaly with edematous gallbladder and minimal ascites. Portal venous doppler showed features of Inferior vena cava stenosis or web (Intrahepatic stenosed IVC measuring 2mm with intrahepatic venous collaterals). CECT abdomen showed features of Budd Chiari malformation as shown in Fig1. Fibro scan done showed F4 fibrosis with no steatosis (4). IVC stenting with angioplasty procedure was done by intervention radiologist (5).

### DISCUSSION

In the pediatric age group, Budd chiari syndrome(BCS) is comparatively rare; the majority of cases are recorded from the Indian subcontinent. The variation in clinical and pathological symptoms across BCS patients is mostly dependent on the location of blockage. Furthermore, the quantity and rate of blockage of hepatic veins have a substantial impact on the pathophysiological process (1).

Prothrombotic and nonthrombotic variables can be used to classify the etiology of BCS. Hereditary thrombophilic diseases, including protein C and S, factor V Leiden, and antithrombin III (ATIII) deficiency, are among the most prevalent etiologies. Myeloproliferative diseases, antiphospholipid syndrome, and paroxysmal nocturnal hemoglobinuria (PNH) are examples of acquired disorders that are thought to be comparatively uncommon. Sarcoidosis, immunoallergic vasculitis, granulomatosis, Behcet's disease,

and celiac disease are possible systemic prothrombotic causes of BCS. It can also happen as a pathological result of hydatid illness, benign or malignant tumors, or pyogenic infections that clog the hepatic vessels (2).

The use of imaging tools is the most important conclusion made for the diagnosis of pediatric BCS. Because it can show hepatic vein or IVC obstruction and has a 95% diagnosis accuracy, Doppler US is regarded as the gold standard investigation to diagnose BCS(4).

### CONCLUSION

In children, BCS is an uncommon cause of liver cirrhosis. Diagnosis is often delayed due to varied presentation. The symptoms may have an acute or chronic history. When a child exhibits the aforementioned symptoms, pediatrician should consider BCS as a differential diagnosis because it could be mistakenly identified as an illness. Higher death rates in youngsters are a result of delayed diagnosis. This case study demonstrates the clinical significance that prompt diagnosis and treatment improving the prognosis.

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