MEDICAL SCIENCE

Rare case of Budd-Chiari Syndrome with recurrent thrombosis requiring stenting

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ABSTRACT

Budd-Chiari syndrome is a very rare disorder in paediatric age group, which is characterised by narrowing or obstruction of hepatic venous outflow. The obstruction might be thrombotic or non-thrombotic along the course of hepatic venules to junction of inferior vena cava to the right atrium, which leads to abdominal pain in right hypochondrium, hepatomegaly, ascites and portal hypertension. The cause of venous obstruction in children is mainly due to hypercoagulable state. The mean age of Budd-Chiari syndrome is 20-40 years, but here we can see that it occurs in paediatric age group, so good clinical suspicion is required along with investigation to confirm the diagnosis. Traditional approach to the treatment of Budd-Chiari syndrome is systemic thrombolysis or surgical Porto systemic shunt. Recently, successful treatment of Budd-Chiari syndrome is done by endovascular techniques, including angioplasty and stent placement. Here, in this case it shows a successful percutaneous recanalization of complete hepatic vein occlusion by angioplasty and stent placement.

Keywords: Budd-Chiari syndrome, Abdominal distension, Hepatic vein obstruction, Stent placement.

1. INTRODUCTION

Budd-Chiari syndrome is a heterogenous group of clinical condition, characterised by blockage of the hepatic veins' outflow from the right atrium to the inferior vena cava (Dileep et al., 2021). The mean age of occurrence of Budd-Chiari syndrome is twenty to thirty-nine years. Its occurrence is very rare in children i.e., one in a million individuals. It can be acute, chronic, fulminant or asymptomatic. The primary Budd-Chiari syndrome is predominately due to thrombosis, although secondary Budd-Chiari syndrome is due to compression or invasion of hepatic veins/inferior vena cava (Ferral et al., 2012). Inability of blood to exit the liver due to obstruction of small or major veins causes hepatic congestion, which causes micro vascular ischemia and hepatocellular damage. The clinical manifestations like abdominal pain in right hypochondrium, hepatomegaly, ascites and portal hypertension. The diagnosis will be made by Doppler ultrasonography, CT or Magnetic resonance angiography. The main aim of the treatment is to restoration and adequacy of venous outflow. Liver transplantation is the main

stay treatment for Budd-Chiari syndrome.

2. CASE REPORT

A case of 13-year-old female child resident of Amravati, Hindu by religion, admitted in AVBR Hospital, Sawangi, Wardha, with a 3-month history of abdominal distension, episodes of vomiting's and black coloured stools and increased work of breathing for 6 days. The child was asymptomatic 3 months back, then child noticed that abdominal distension which was slowly progressive associated with increased work of breathing, not associated with any abdominal pain, jaundice, bruising. 10 days later child developed fever which was high grade, continuous in nature, associated with chills and rigors and relived on oral medication and lasted for one day. Child then developed vomiting's, which was non- projectile, non-bilious and non-blood stained, contains food particles. Gradually abdominal distension was increasing that leads to difficulty in breathing, for which the child was admitted to AVBRH.

On examination child is vitally stable with heart rate-86/minute, respiratory rate-30/minute, blood pressure-120/74 mm Hg, with no pallor, icterus, clubbing or lymphadenopathy, Ophthalmic examination was done suggestive of no Kayser-Fleischer ring. On systemic examination: The abdomen was distended, with visible veins and with no scars and sinuses and no visible pulsations, the umbilicus was everted with flanks being full, on percussion fluid thrill was present. As abdomen was tense, the liver and spleen couldn't appreciate. Respiratory system was normal with no added sounds, cardiovascular system-s1s2 heard with no murmur. Abdominal girth was 72 cm.

Routine investigations reveal haemoglobin of 10.4 g/dl, total leucocyte count of 10,700 cells/mm3 and a differential count of 60%-granulocytes, 35%-lymphocytes, 1%-eosinophil. The platelet counts 1.02 lakhs. ESR-4, the liver function test and kidney function test were normal. The serum uric acid level was 3.2 mg/dl. The HIV, Hepatitis B was negative. The ascitic fluid examination shows WBC=2-3 cells/HPF, TLC=approximately 250 cells/cu mm, DLC shows 65%-polymorphs, 35%-lymphocytes, with protein-4 g/dl, LDH-77 IU/L, Sugar-100 mg/dl, ADA-2.750 IU/L. Ascitic fluid suggestive of transudative. Coagulation profile was deranged with INR–1.5, PT-18.4 and APTT–41.3. Viral hepatitis and abdominal tuberculosis were ruled out by doing Truenat and doing Mantoux test. Ceruloplasmin levels are normal that clears the suspicion of Wilson's disease. Thrombophilia factors screening was done which shows Protein C value at 56%, while free protein S at 69%, these two values are normal and antithrombin levels at 67.5% were low. Homocysteine level were sent which suggestive of normal, the value is 9.1 μmol/L. On admission child was kept on o2 by nasal prongs, started on IV fluids, injection ceftriaxone, inj. Pantoprazole and injection metronidazole, as coagulation profile was deranged, injection vitamin K was given and Fresh frozen plasma was transfused. Fibro scan -2. Ultrasonography of the abdomen shows raised echotexture of the liver with mild hepatomegaly with gross ascites. Endoscopy was done to rule out oesophageal varices, the report suggestive of normal. CECT abdomen was done suggestive of gross ascites with splenunculus measuring 10x8 mm in size (Figure 1).

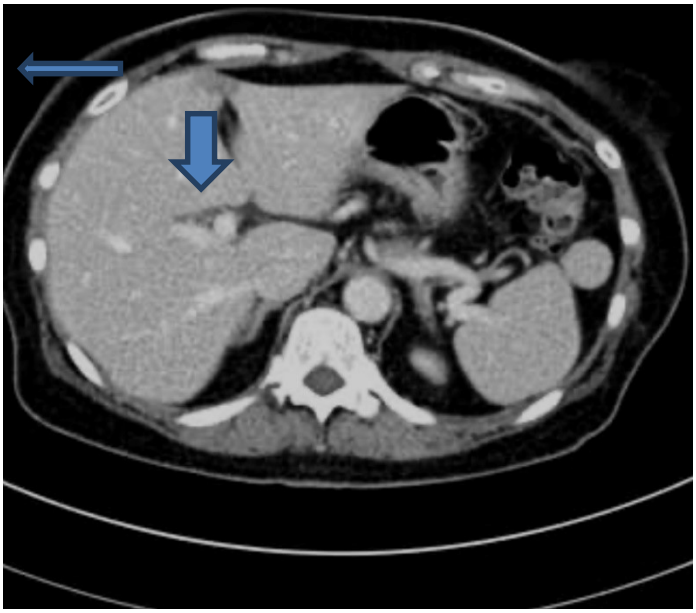

Figure 1 Gross ascites and Accessory spleen can be seen

Venogram was done suggestive of narrowing of inferior vena cava. The inferior vena cava stenting was done under all aseptic precautions and local anaesthesia, stent placement was done. Good forward noted across the stent. Review Doppler was done suggestive of good flow across the stent placement. The child is vitally and haemodynamically stable hence being discharged on anti-coagulant therapy. The abdominal girth on discharge was 67 cm.

After one month the child was admitted in AVBRH with similar complaints like reappearance of abdominal distension and increase work of breathing. As distress increased child was admitted in PICU and kept on o2 by nasal prongs, as distress was not settled child got intubated and kept on mechanical ventilation. On examination abdominal girth was 80 cm. Air entry was decreased (Figure 2, 3).

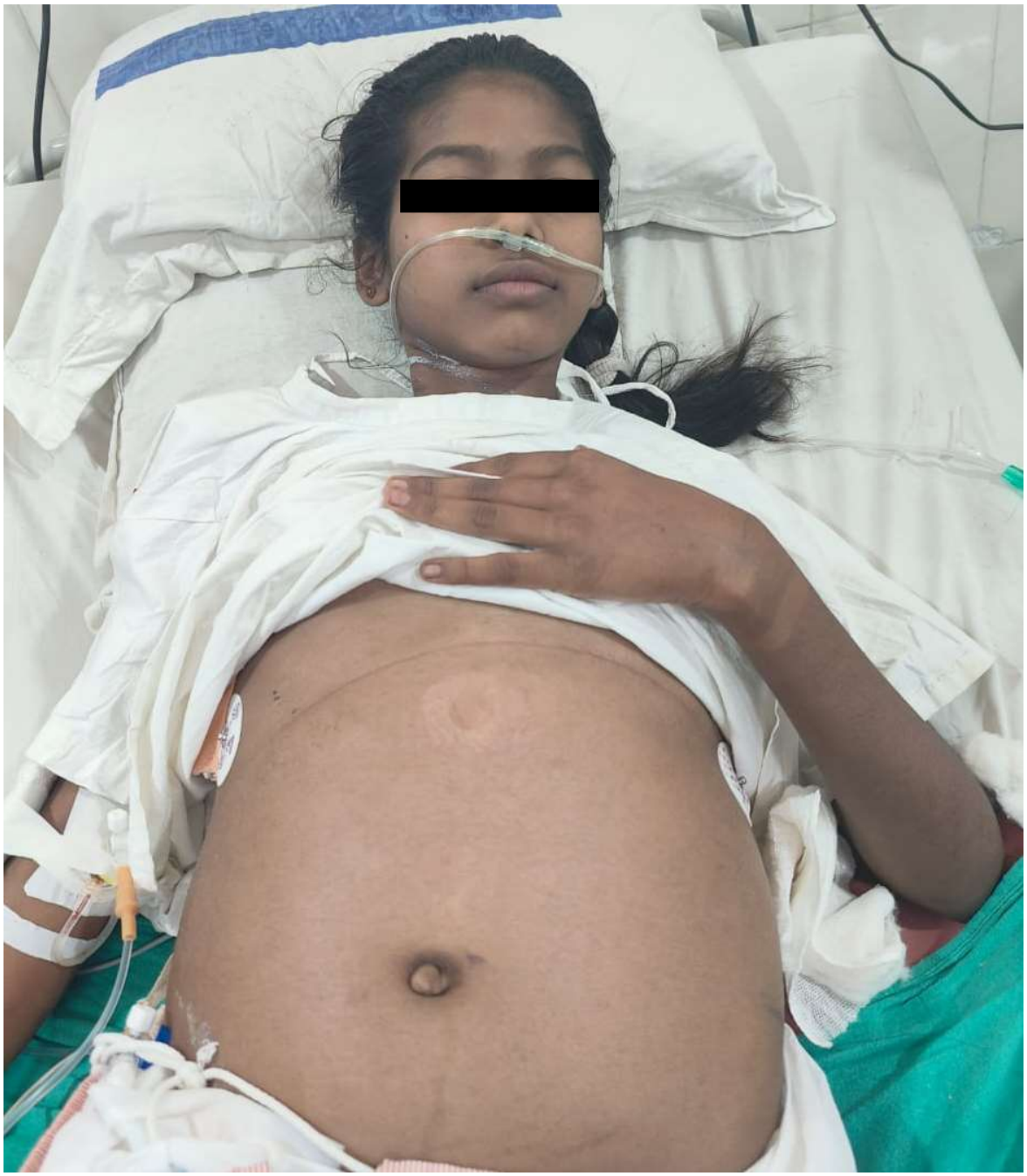

Figure 2 Abdominal distension

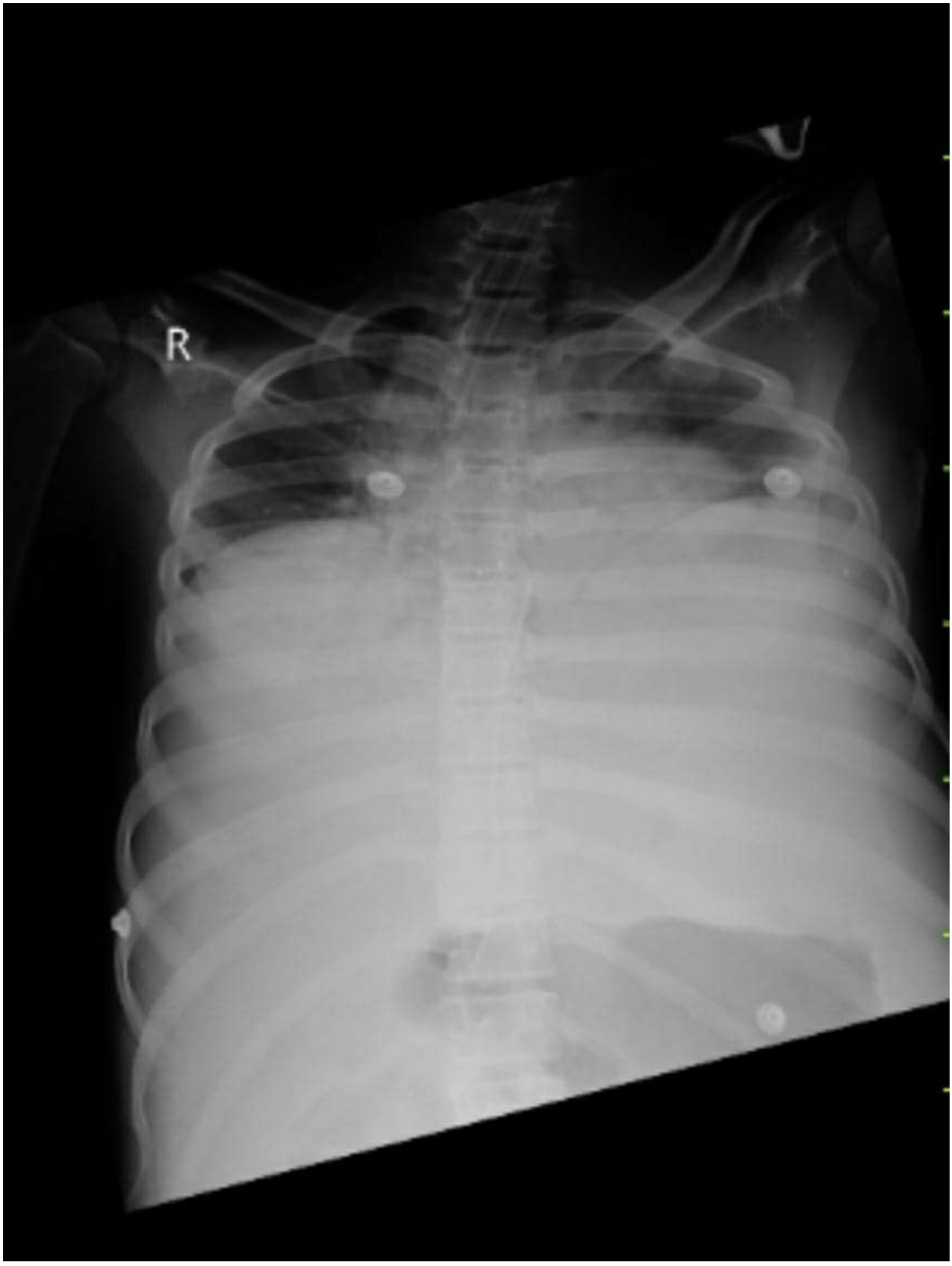

Figure 3 Chest X-ray shows massive pleural effusion

Routine investigations were sent suggestive of normal; liver function test and kidney function test were normal. Coagulation profile was deranged, so fresh frozen plasma was transfused. Chest X-ray was done suggestive of bilateral pleural effusion. Ultrasonography of the abdomen was done suggestive of liver shows enlarge in size with raised echotexture, with gross ascites, portal vein measures 7mm which was oedematous. IVC stent was present. Child was kept NBM and started on dextrose IV fluids and injection ceftriaxone, metronidazole, pantoprazole and injection furosemide was started.

Ascitic tap was done which suggestive of transudate in nature. Venography was done under all aseptic conditions right internal jugular vein puncture done. Access maintained through 6f sheath. Selective cannulation of right hepatic vein done using hi-torque command 18 ST 0.018" 300 cm 10 cm/4 g. Liver parenchyma pierced using chiba needle ultrasound guided right hepatic vein access was taken. Check venogram done and hepatic vein was confirmed. Balloon-plasty was done across the tract along the stent in inferior vena cava. Abbott vascular omnilink elite 9 mm x 29 mm x 135 cm stent was deployed across the tract communicating hepatic vein to inferior vena cava, good forward flow was seen across the stent (Figure 4, 5).

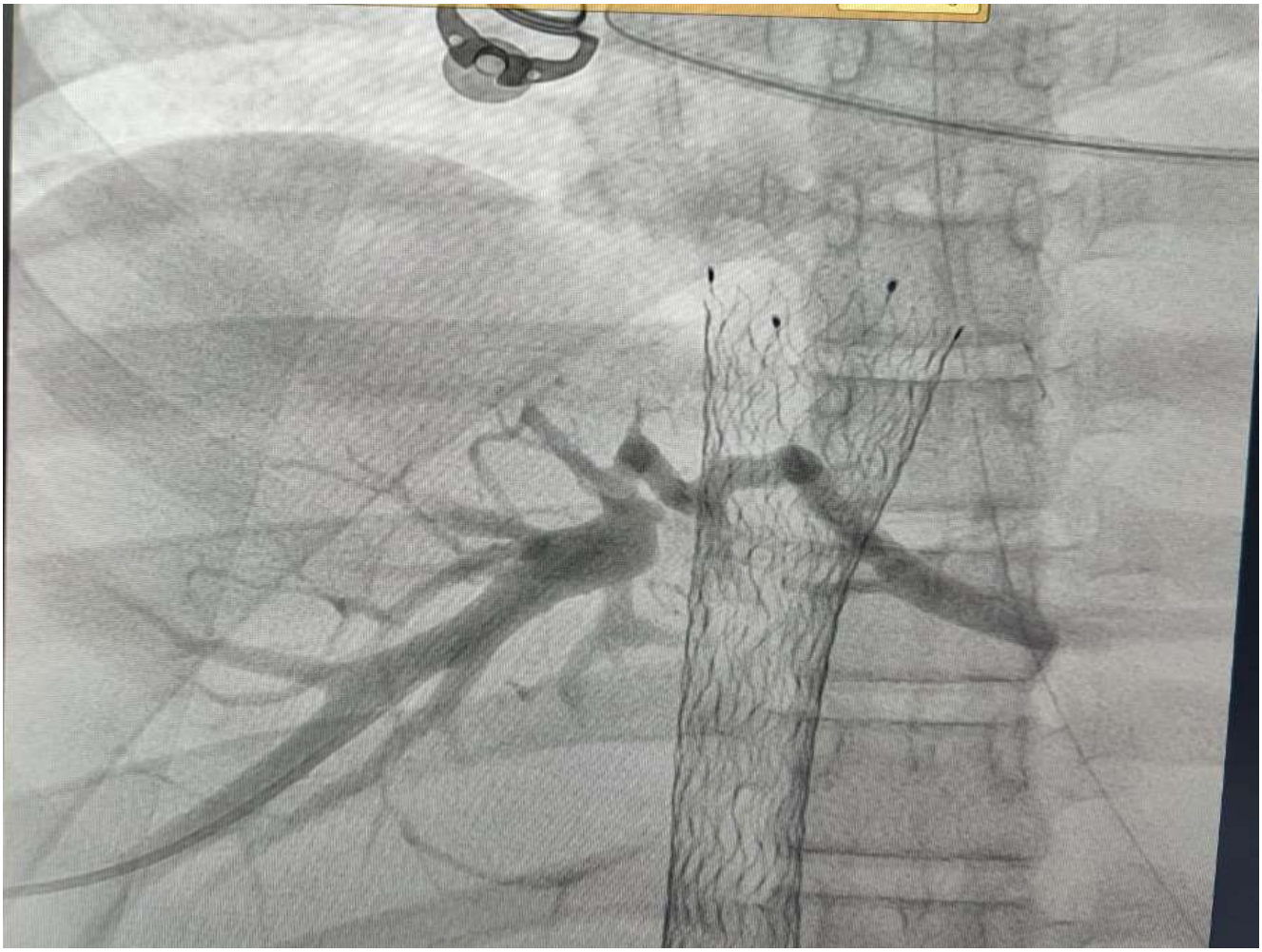

Figure 4 Hepatic vein thrombosis

Gradually the ventilation settings were reduced and child got extubated. The patient was ambulating on the next day, there was no distress was present and there is marked decrease in the abdominal girth. No post procedure anticoagulation was given. Coagulation profile was normal. Subsequent examinations during the hospital stay of 22 days, the child is vitally stable and decrease in the girth of abdomen and hence child got discharged (Figure 6). Abdominal girth was 71 cm which decreased 9 cm from admission.

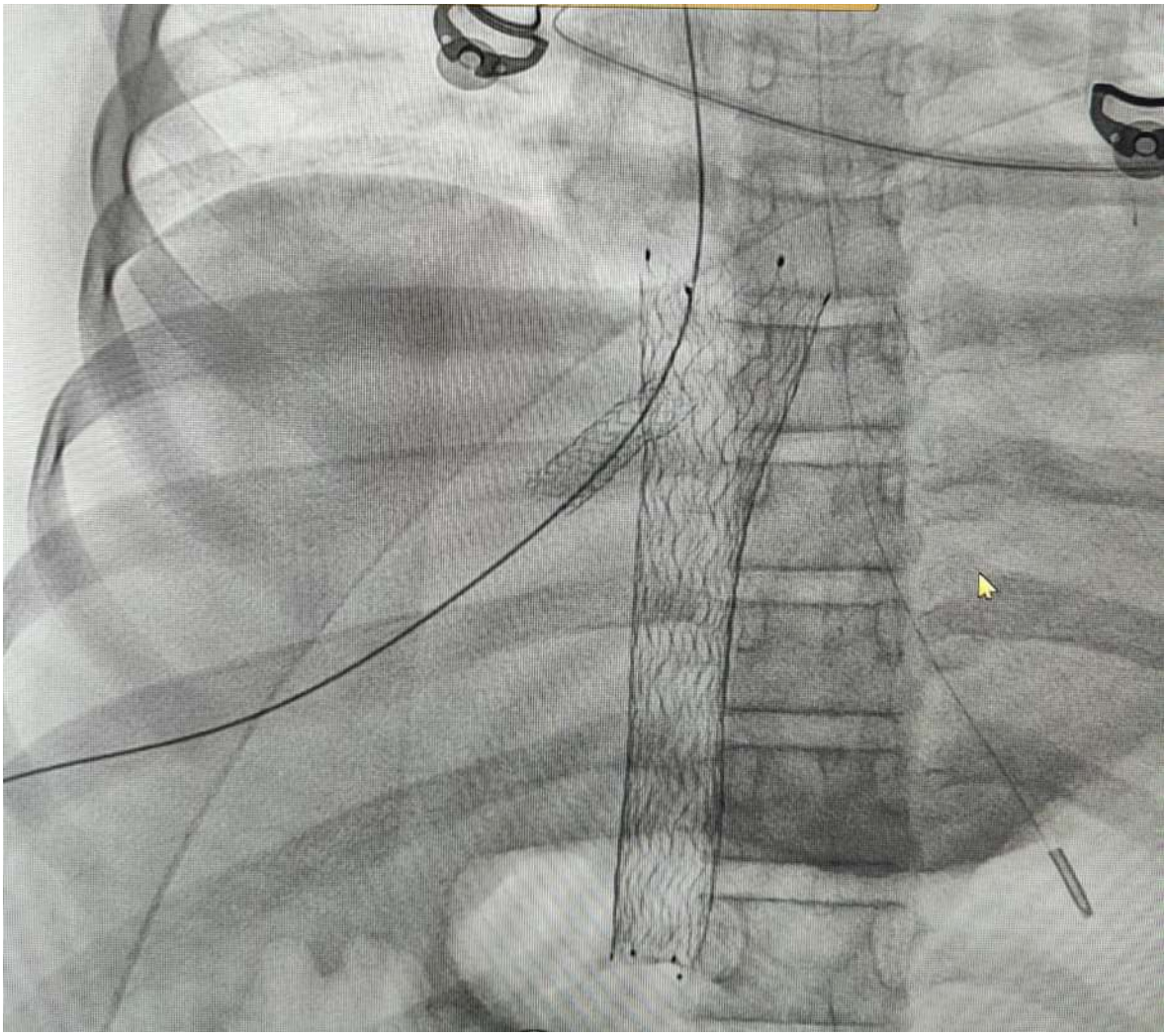

Figure 5 Stent placement

Figure 6 This is the picture after 15 days of discharge

3. DISCUSSION

Budd-Chiari syndrome is a very uncommon condition seen in children, characterised by obstruction of hepatic venous outflow tract. The term Budd-Chiari was coined in the year 1800s after the work of George Budd. He described three cases of hepatic vein thrombosis in 1845 and Hans Chiari, who reported the first pathologic description of obliterating Endo phlebitis of the hepatic veins (Aydinli and Bayraktar, 2007).

The causes of Budd-Chiari syndrome are inherited and acquired state of hypercoagulable. Inherited causes such as protein C, S deficiency, factor V Leiden mutation and anti-thrombin III deficiency are the common causes of hepatic vein thrombosis which results in BCS (Timsaal et al., 2021). Acquired causes such as polycythaemia Vera, paroxysmal nocturnal haemoglobinuria and myelofibrosis, accounts for >50% of BCS cases.

There are three varieties of BCS that is fulminant, acute and sub-acute/chronic type. In 75–80% of instances, patients exhibit encephalopathy, ascites, liver failure, lower limb oedema, fever and abdominal pain and distension. Fulminant BCS child develops severe abdominal pain, liver failure with raised liver enzymes and coagulopathy with encephalopathy and renal failure within a few days (Lin et al., 2017). The more prevalent variety of BCS, chronic instances, appears with a later start and fewer symptoms. It clinically manifests as cirrhosis and portal hypertension. This was evident in our patient by a Metavir score of 2 in fibro scan, which suggestive of fibrosis with periportal expansion but few septa formation (Li et al., 2018). Our case also presented with similar positive clinical findings for abdominal pain and ascites with respiratory distress. The BCS also includes jaundice, lower extremity ulceration and impaired mental status, although none of these symptoms were present in our case.

The diagnosis of Budd-Chiari syndrome is made through imaging techniques. Ultrasonography of the Doppler is the gold standard method, its accuracy is 95% and it demonstrates hepatic vein/IVC obstruction. In our patient it shows gross ascites with IVC stent placement and hepatic vein thrombosis. In our patient it is type III BCS, where both hepatic vein and IVC were involved (Patil et al., 2012). Other diagnostic methods are CT scan and MRI. CT scan is usually done to know the morphology of the liver architecture and the vascular abnormalities.

If the child had severe signs and symptoms like painful hepatomegaly, marked ascites then start with anticoagulant therapy, followed by Tran's jugular intrahepatic Porto systemic shunt. If it is unsuccessful then angioplasty with stent placement. In our patient on first admission, we have done stent placement of IVC, whereas second admission we have done hepatic vein stent placement was done. The best option overall for children is liver transplantation, with lifetime anticoagulant treatment as a backup.

4. CONCLUSION

The most common presentation of Budd-Chiari syndrome is thrombosis of hepatic vein which is its diagnostic feature. Our case was different as it initially presented with IVC thrombosis and then later developed hepatic vein thrombosis. There is no such case report in the literature of Budd-Chiari syndrome presenting initially with IVC thrombosis. Hence any case of IVC thrombosis should raise suspicion of developing BCS in future.

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None

Author contribution

Sushma Myadam collected the information and prepared the case report, which was thoroughly reviewed by Dr Revat Meshram, and Dr Amar Taksande.

Informed consent

Oral and written consent was taken from relatives.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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