ejpmr, 2024, 11(5), 128-131

SJIF Impact Factor 7.065

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Review Article

ISSN 2394-3211

EJPMR

BUDD CHIARI SYNDROME – AN OVERVIEW

[1]Monica K., [2]Manisha M., [3]Dr. Aswathi Elisabeth Philip and [4]*Dr. M. Vani

[1,2]3rd Year Pharm. D, K.K. College of Pharmacy, Chennai.
[3]Assistant Professor, Department of Pharmacy Practice, K.K. College of Pharmacy, Chennai.
[4]Professor, Department of Pharmaceutics, K.K. College of Pharmacy, Chennai.

***Corresponding Author: Dr. M. Vani**

Professor, Department of Pharmaceutics, K.K. College of Pharmacy, Chennai.

Email Id: mandrummoorthy@gmail.com

Article Received on 01/03/2024 **Article Revised on 21/03/2024** **Article Accepted on 11/04/2024**

ABSTRACT

To study an overview of Budd Chiari syndrome among different population. Relevant articles on Budd Chiari syndrome for clinical manifestations, pathophysiology, treatment oregimens from various journals were studied to gather complete information to meet the objectives. Meta-analyses of the current literature provided evidence that BCS is a rare vascular liver disease Hepatic vein obstruction is the most typical cause.

KEYWORDS: Obstruction, Etiology, Pathophysiology, Cirrhosis, Hepatomegaly, Treatment.

INTRODUCTION

An obstruction or narrowing of the hepatic veins is known as Budd-Chiari syndrome. The liver enlarges as a result of this obstruction, which causes blood to flow back into the liver. Additionally, the spleen could enlarge.

A rare condition known as Budd-Chiari syndrome is characterized by obstruction of the liver's blood-flowing veins. Hepatomegaly is brought on by an obstruction in the blood flow out of the liver, which results in blood building up and collecting there. Splenomegaly can also result from the spleen being enlarged. The outcome of this blood backup is dilated, twisted veins in the oesophagus and an increase in blood pressure in the portal vein, which conducts blood from the intestines to the liver. Ascites, a fluid buildup in the belly, is caused by portal hypertension. The inferior vena cava may potentially be affected by the clot. The abdomen may develop and show signs of varicose veins. The liver can scar under several circumstances. Fatigue, stomach ache, nausea, jaundice, and oesophageal hemorrhage are some more symptoms. Depending on the location and number of afflicted veins, this illness can range in severity from person to person.

Patients with coagulation disorders, such as those who are pregnant, have tumors, chronic inflammatory diseases, clotting disorders, infections, or any myeloproliferative disorders, are more likely to experience it. The cause of Budd-Chiari syndrome (abnormal, enlarged blood vessels in the belly and/or rectum) remains unknown in around one-third of cases.

Budd-Chiari syndrome may cause the following

Increased pressure in the portal vein, which conducts blood from the intestine to the liver, is known as portal hypertension.

Oesophageal varices are twisted veins in the oesophagus (the "food tube").

Ascites, a fluid accumulation in the abdomen.

- liver scarring, or cirrhosis.
- Varicose vein

DEFINITION

Hepatic venous outflow tract obstruction, which is an uncommon condition known as Budd-Chiari syndrome, can occur in any level or manner.[1] A venous process (such as thrombosis or phlebitis) blockage is referred to as primary Budd-Chiari syndrome. Secondary Budd-Chiari syndrome, on the other hand, is characterized by compression of the inferior vena cava and/or hepatic veins by a lesion that originates outside of the vein.

ETIOLOGY

Budd Chiari syndrome is the result of an underlying condition in 80% of cases, most of which are linked to a hyper-coagulable state. When diagnosing and treating Budd Chiari syndrome, this element must be taken into account. The following are the primary contributing factors of Budd Chiari Syndrome.

Myeloproliferative disorders

Polycythaemia Vera and thrombocythemia are two examples of myelo-proliferative disorders that are linked

to Budd Chiari syndrome because they are usually always associated by some form of hypercoagulability.

Malignancy
Malignancy accounts for 10% of cases of Budd-Chiari syndrome. It can directly compress or invade the veins, create hypercoagulability, and result in venous thrombosis and blockage. Hepatocellular carcinoma, renal cell carcinoma, leiomyosarcoma, right atrial myxoma, and Wilms tumour are the most frequent cancers connected to the Budd-Chiari syndrome.

Lesions of the liver
Vasculature compression can occasionally be caused by liver illness or lesions. Common lesions include hepatic cysts, adenomas, cystadenomas, invasive aspergillosis, and aortic aneurysm are common lesions that occurs in Budd-Chiari syndrome.

Pregnancy and oral contraceptives
Oral contraceptives and pregnancy lead to hypercoagulable state and are responsible for about 20 percent of cases of Budd-Chiari syndrome.

- Congenital
- Hepatic vein or inferior vena cava web
- Interruption of the diaphragm
- Venous thrombosis secondary to
- Dehydration
- Sepsis
- Polycythemia rubra vera
- Antiphospholipd syndrome
- Pregnancy and the postpartum state
- Oral contraceptive pill use
- Sickle cell disease
- Thrombocytosis
- Paroxysmal nocturnal hemoglobinuria (PNH)
- Injury and/or inflammation secondary to
- Phlebis: bone marrow transplant and chemoradiotherapy
- Autoimmune disease
- Behcet's disease
- Tumor invasion, e.g. renal cell carcinoma, hepatocellular carcinoma, adrenal carcinoma
- Leiomyosarcoma of inferior vena cava

It may be associated with concurrent portal vein thrombosis.

Idiopathic
20 percent of cases are idiopathic

The following hypercoagulable diseases also contribute to Budd-Chiari syndrome

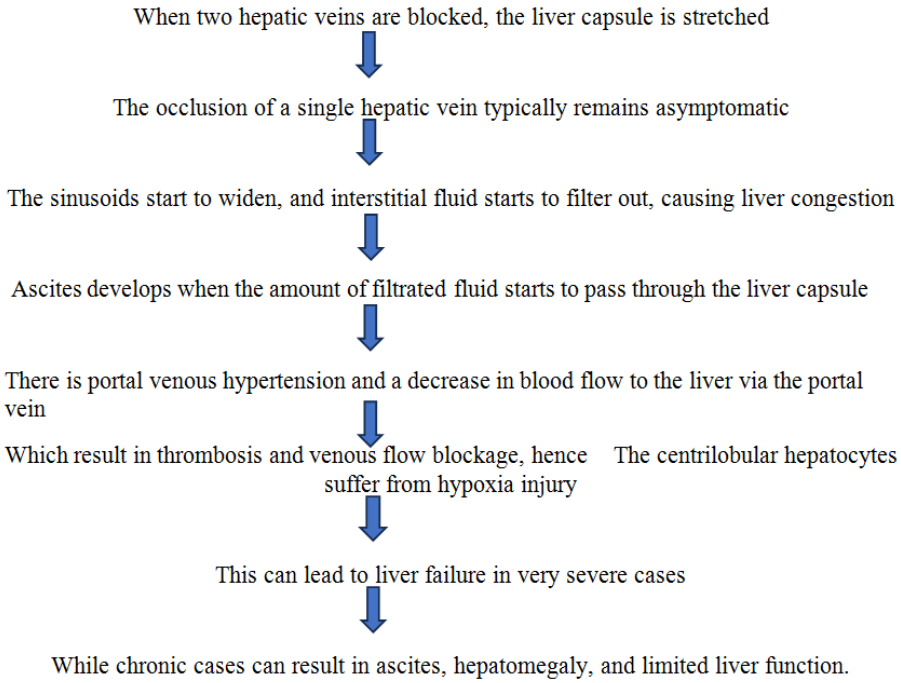
- A mutation in factor V (Leiden) causes protein C resistance antibodies to phospholipids syndrome a lack of antithrombin protein C shortage Haemoglobinuria at night that is recurrent
- Sickle cell
- Inflammatory bowel disease, a class of conditions that irritate and inflame the gastrointestinal tract.
- Pregnancy.

EPIDEMIOLOGY
Four studies—in Denmark, Sweden, Italy, and France—and two in Asia (Japan and South Korea) were carried out. They were all of quite high caliber. BCS had an annual incidence of 0.168 to 4.09 per million. BCS affected 2.40 to 33.10 people out of every million. According to meta analyses, the combined yearly incidence and prevalence of BCS were 1 per million and 11 per million, respectively.

Studies' differences from one another were statistically significant. According to subgroup meta-analyses, BCS had a combined yearly incidence of 0.469 per million in Asia and 2 per million in Europe, and a combined prevalence of 5 per million in Asia.

Conclusion: Meta-analyses of the current literature provided evidence that BCS is a rare vascular liver disease. The prevalence of BCS may not be higher in Asia and Europe. More epidemiological data from non-Asian nations should be necessary since Budd-Chairi syndrome is typically identified in the third or fourth decade of life and primarily affects women. Hepatic vein obstruction is the most typical cause. In Asian nations, males are more likely to experience it, and inferior vena cava occlusion or a combination of inferior vena cava and hepatic vein blockage is the most common cause.

PATHOPHYSIOLOGY



DIAGNOSIS

Physical examination that reveals abdominal swelling as the cause of the diagnosis. In order to assess your liver's health and determine whether you have a higher risk of blood clots, your doctor will also set up blood tests.

- If your veins are obstructed by clots, imaging tests will reveal this. One of these tests is ultrasound, which sends high-frequency sound waves through the body's tissues. The echoes are captured and used to create videos or pictures of the inside organs of the body.
- Computed tomography [CT] scan, which creates images of a cross-section of the body using X-rays and computers.
- Magnetic resonance imaging (MRI), a test that creates extremely detailed images of the body using a big magnet, radio waves, and a computer.
- In order to determine whether you have experienced cirrhosis (liver scarring), your doctor may prescribe a liver biopsy (removal of cells or tissue for microscopic inspection).

COMPLICATIONS

Complications are mostly related to the degree of liver failure.

In general, untreated Budd-Chiari syndrome can lead to the following

- Hepatic encephalopathy
- Variceal haemorrhage
- Hepatorenal syndrome
- Portal hypertension
- Bacterial peritonitis in the presence of ascites
- Hepatocellular carcinoma

TREATMENT

Blood clots in the liver are intended to be broken up as part of treatment for Budd-Chairi syndrome. Surgery, non-surgical techniques, and pharmacological therapy are all used as forms of treatment.

Drug therapy: Typically, medications are recommended to dissolve blood clots. Additionally, warfarin (Coumadin), a blood thinner, is frequently administered as a clot preventative.

- Giving prescriptions for blood thinners such heparin and warfarin; • putting in a stent to aid the blood vessel's drainage
- Using thrombolysis, a minimally invasive technique to dissolve or condense blood clots.
- Undergoing surgery to open up veins or remove blood clots.
- Procedures: Two non-surgical methods are employed to treat Budd-Chiari syndrome.
- Percutaneous transluminal angioplasty and the transjugular intrahepatic portosystemic shunt
- A stent is inserted at the center of the liver during the transjugular intrahepatic portosystemic shunt (TIPS), a radiologic operation that modifies blood flow. A radiologist connects the portal vein by puncturing the liver during the surgery. To keep the track open, a metal stent—a brief, hollow tube—is inserted into the opening. The TIPS surgery changes the liver's blood flow and lowers pressure in all the aberrant veins, including those that supply the colon and the liver.
- During a percutaneous transluminal angioplasty operation, a catheter—a skinny, hollow tube with a balloon at one end—is inserted through the patient's skin and into the blood vessel. Once the clot is found, the catheter is directed toward the clotted

area. To enlarge the vein, the balloon is inflated when the catheter reaches the clot. The vein may be opened by inserting a stent there.
- Putting in a stent to assist the blood vessel's drainage.

Surgery: Patients with liver failure, a condition in which the liver no longer functions effectively, are usually treated with a liver transplant.

CONCLUSION

Review of the current literatures provided evidence that BCS is a rare vascular liver disease and Hepatic vein obstruction is the most typical cause. Most common treatment or interventions are surgery.

REFERENCES

1. DeLeve LD, Valla DC, Garcia-Tsao G., American Association for the Study Liver Diseases. Vascular disorders of the liver. Hepatology, 2009 May; 49(5): 1729-64. [PMC free article] [PubMed]
2. Plessier A, Valla DC. Budd-Chiari syndrome. Semin Liver Dis., 2008 Aug; 28(3): 259-69. [PubMed]
3. Akiyoshi H, Terada T. Centrilobular and perisinusoidal fibrosis in experimental congestive liver in the rat. J Hepatol, 1999 Mar; 30(3): 433-9. [PubMed]
4. Witte CL, Witte MH, Dumont AE. Lymph imbalance in the genesis and perpetuation of the ascites syndrome in hepatic cirrhosis. Gastroenterology. 1980 May; 78(5 Pt 1): 1059-68. [PubMed]
5. Tanaka M, Wanless IR. Pathology of the liver in Budd-Chiari syndrome: portal vein thrombosis and the histogenesis of veno-centric cirrhosis, veno-portal cirrhosis, and large regenerative nodules. Hepatology, 1998 Feb; 27(2): 488-96. [PubMed]
6. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. World J Gastroenterol, 2007 May 21; 13(19): 2693-6. [PMC free article] [PubMed]
7. Merck Manual Consumer Version. Budd-Chiari Syndrome. *(https://www.merckmanuals.com/home/liver-and-gallbladder-disorders/blood-vessel-disorders-of-the-liver/budd-chiari-syndrome)* Accessed, 10/15/2019.
8. National Organization for Rare Disorders. Budd Chiari Syndrome. *(https://rarediseases.org/rare-diseases/budd-chiari-syndrome/)* Accessed 10/15/2019.
9. National Institutes of Health/Genetic and Rare Diseases Information Center. Budd-Chiari syndrome. *(https://rarediseases.info.nih.gov/diseases/5968/budd-chiari-syndrome)* Accessed 10/15/2019.
10. Ferral H, Behrens G, Lopera J. Budd-Chiari Syndrome. American Journal of Roentgenology. 2012; 199: 737-745. Accessed 10/15/201.