

CURRENT KNOWLEDGE IN MANAGEMENT OF BUDD-CHIARI SYNDROME(BCS) IN PREGNANCY: A CASE SERIES ANALYSIS

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INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare disorder characterized by thrombotic or non-thrombotic obstruction of hepatic venous flow anywhere along the venous course from the hepatic venules to junction of the inferior vena cava (IVC) to the right atrium with an incidence of 1 in 100000 to 500000, and a preponderance in females. However management is a challenge in pregnancy due to the inherent prothrombotic state thus exerting a detrimental effect on the pregnnant mother. With the therapeutic advances in medicine , BCS in pregnancy is on the rise. However, limited evidence exists for management of pregnancy in such women. Here we present a retrospective case-series analysis of pregnant women with BCS in our tertiary-care centre and aim to formulate an evidence-based management plan for an optimal outcome.

CASE REPORTS

CASE 1

38-year-old G4A3, diagnosed BCS with Inferior vena cava (IVC) thrombosis post IVC stenting, Anti-phospholipid Antibody (APLA) positive with history of Catastrophic Pulmonary Embolism, Pulmonary Hypertension, Chronic Hypertension, Diabetes Mellitus, Immune Thrombocytopenic Purpura was referred to us at 14 weeks. As she was keen to continue the pregnancy ,a multidisciplinary team including obstetrician, maternal and fetal medicine specialist, cardiologist, hematologist, rheumatologist, hepatobiliary specialist, endocrinologist, critical care specialist was involved in her care. At 18 weeks the fetus had severe fetal growth restriction with oligohydramnios and cardiac defect. Genetic counselling and testing were offered. She underwent suction evacuation with cardiac monitoring and intensivists care. Pscyhological support and contraceptive advise was also given. She was discharged on long term anticoagulants, steroids for platelet improvement, antihypertensives and oral hypoglycemics.

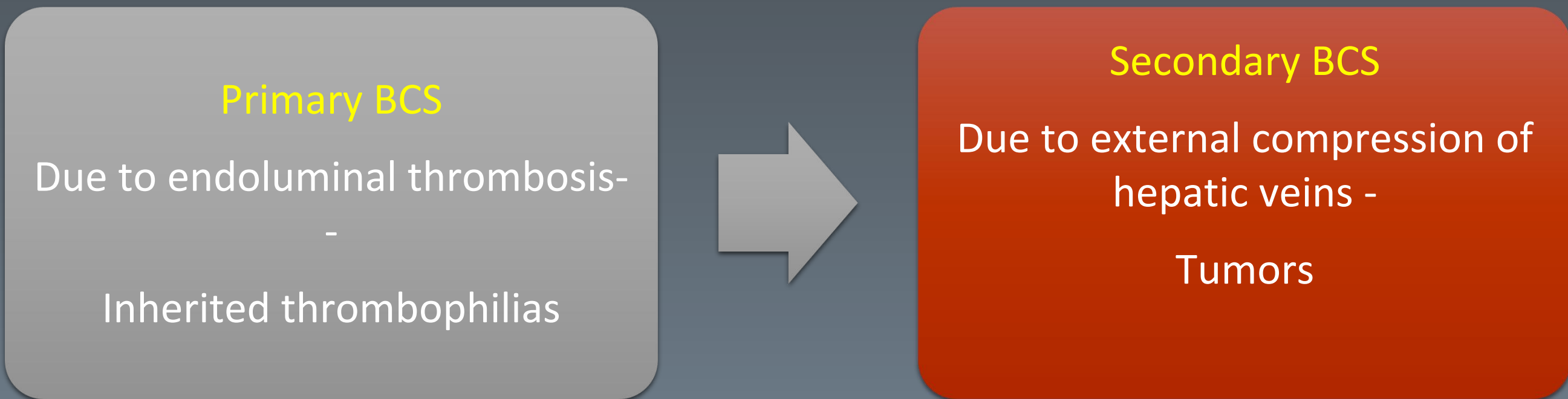
CASE 2

31 year old G3A2 diagnosed with chronic liver disease , hepatic vein thrombosis and BCS with etiology secondary to factor V Leiden mutation. She was on long term anticoagulant therapy with Warfarin. After pregnancy risks being explained, she opted to continue pregnancy. During early pregnancy, she was switched over from Warfarin to LMWH and was also started on LDA 150mg. Her First Trimester Screening was low risk, Targetted Anomaly Scan showed no anomalies, serial growth scans were all normal. At 33 weeks, she went into preterm labor, was given tocolysis and antenatal steroids. Heparin injection was stopped at the onset of labor pains. A live preterm 1.79 kg baby was delivered by Emergency LSCS. Postoperatively, she was given bridging anticoagulation and switched over to Warfarin with daily INR monitoring. A multidisciplinary team including the primary obstetrician, maternal and fetal medicine specialist, cardiologist, hematologist, rheumatologist, hepatobiliary specialist ensured that she had a safe pregnancy and puerperium.

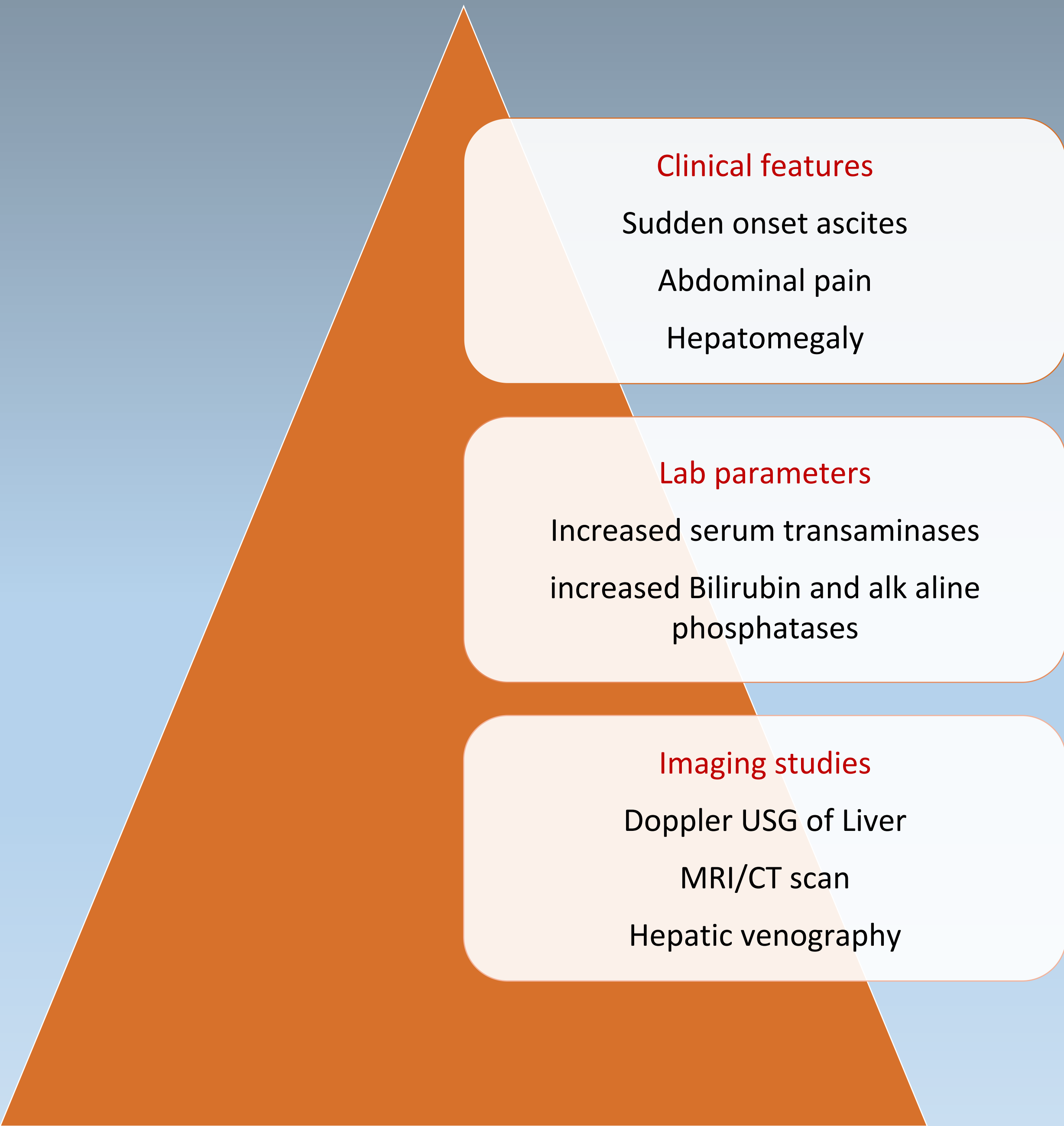
CASE 3

36 year old G2 L1, with previous normal delivery, a known case of BCS, chronic liver disease with portal hypertension , autoimmune hypothyroidism , immune mediated progressive ascending myeloneuropathy and with h/o main hepatic vein angioplasty and stenting 1 year back and was on long term anticoiagulants. After pregnnacy risks being explained, she opted to continue pregnancy. She was switched from Warfarin to LMWH and LDA 150mg was started from early pregnancy. Her First Trimester Screening(FTS) was low risk, Targetted Anomaly Scan showed no anomalies. She developed Gestational Diabetes Mellitus and was controlled on insulin. Her blood pressure remained normal through out pregnancy. All her growth scans were normal and she underwent full term vaginal delivery and delivered baby of 2.8kg. She underwent puerperal sterilisation, bridging anticoagulation done postpartum and was discharged on Warfarin. However, 6 months later, she developed an acute short febrile illness with severe anemia, hepatosplenomegaly, hepato-encephalopathy and pre-renal acute kidney injury and succumbed while being investigated for the cause.

Classification



Investigations



Management

• Preconceptional counselling

Risks of BCS in pregnancy, chance of flare-ups,,worsening portal hypertension, liver function deterioration, hepatic encephalopathy, increased thrombotic risks and development of pulmonary artery hypertension to be explained and

• Multidisciplinary team approach

Which includes Maternal fetal medicine specialists, hepatobiliary specialists, hematologists, internal medicine specialists, anaethetists and neonatologists

• Revisit of Medications

Early pregnancy switch over from Warfarin to LMWH, Therapeutic dose of LMWH as and when indicated
In those at very high risk of thrombosis, Warfarin can be continued after the period of organogenesis.

• Lab parameters

Close monitoring of hepatic function and high index of suspicion for development of complications

• Obstetric management

First trimester screening/Targetted anomaly scan
Serial growth monitoring every 3 weeks from 24 weeks
Vaginal delivery is the preferred mode of delivery unless there are severe esophageal varices.
Stop LMWH therapeutic dose 24hrs prior to delivery.
Consider switching to unfractionated heparin(UH) at 36wks (as UH has short t1/2, rapidly reversible action) especially in patients at high risk of thrombosis. If using UH therapeutic dose, stop 12hrs prior to delivery.

• Postnatal management

Adequate hydration and mobilisation is encouraged.
Restart LMWH 6 hrs after normal delivery and 12hrs following Cesarean Section
Do bridging anticoagulation from LMWH to Warfarin with daily INR monitoring.
Avoid estrogen containing hormonal contraceptives as they are at high risk of thrombosis.
Specialist followup- Gastroenterologist/ hematologist prior to discharge