Mini commentary on BJOG-21-0823: Pregnancy outcomes in women with Budd-Chiari syndrome or portal vein thrombosis - A multicentre retrospective cohort study.

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Budd-Chiari syndrome (BCS) and portal vein thrombosis (PVT) are rare thrombotic disorders which can affect females of reproductive age. Physiological changes in pregnancy may result in or exacerbate pre-existing known portal hypertension related issues associated with these conditions. These conditions may also present de-novo in pregnancy with acute onset ascites or variceal haemorrhage. Both in pregnancy and in the non-pregnant state, in those with established disease, in general, the overall balance of risk, favours continued anticoagulation.

The study by Wiegers et al. has shown favourable maternal and foetal outcomes once greater than 20 weeks gestation is reached in patients with BCS and/or PVT. However, the risk of preterm birth and early pregnancy loss remains. These results are in keeping with the recent study by Andrade et al. (Journal of Hepatology 2018; 69: 1242-1249) looking at pregnancy outcomes from 24 pregnancies in 16 women with idiopathic non-cirrhotic portal hypertension (INCPH). Rautou et al. (Journal of Hepatology 2009; 51: 47-54) reviewed 24 pregnancies in 16 women with BCS and also reported similar findings. Taken as a whole, these results support the concept that patients with vascular liver disease can achieve favourable pregnancy outcomes but warrant careful consideration in pregnancy.

Preconception counselling is a crucial opportunity to optimise patients with vascular liver disease who are considering pregnancy. This can be achieved in a multidisciplinary forum with input from obstetricians, haematologists and hepatologists. It is useful to identify those women with significant portal hypertension and varices before pregnancy so that appropriate surveillance and eradication with endoscopic band ligation and/or prophylaxis with beta blockers is undertaken. If pregnancy is achieved before surveillance, then a second trimester endoscopy for those with significant portal hypertension should be performed. In patients with portal and mesenteric vein thrombosis, magnetic resonance imaging of the pelvis may be needed to assess for the presence of abdominal wall/pelvic varices. This stratifies the risk of a variceal bleed and allows planning of the mode of delivery (caesarean, vaginal or assisted vaginal delivery). Wiegers et al. reported 2 variceal bleeds in pregnancy but did not find a significant association with adverse maternal outcomes, though this may be related to the low number of patients. Andrade et al. also reported 2 variceal bleeds including 1 patient with PVT without adequate endoscopic prophylaxis who required a portosystemic shunt. This outlines the need for appropriate screening and portal hypertension management according to findings.

The majority of patients with BCS and PVT have an underlying pro-thrombotic tendency and the intra-partum and the post-partum periods are associated with thrombotic events. Vitamin K antagonists are historically the commonest anticoagulation used in BCS and PVT which should be switched to low molecular

heparin ideally before conception. The use of anticoagulation is more common in patients with BCS and PVT than in INCPH (38/45 women in BCS and/or PVT compared to 4/16 in INCPH). 4 out of 6 women with BCS and/or PVT who experienced post-partum haemorrhage (PPH) were on anticoagulation. 2 patients with INCPH had PPH whilst on anticoagulation which may be confounded by the thrombocytopenia and type-2 error. In the study by Rautou et al., 17/24 pregnancies received anticoagulation and the 4 women who experienced post-partum bleeding (vaginal or intrauterine/parietal haematoma) were on anticoagulation which includes one woman with an ectopic pregnancy. No maternal deaths were reported in the three studies and the continued use of anticoagulation when indicated is safe and appropriate.

The mode of delivery did not affect the risk of PPH in the three studies. The mode of delivery should be decided based on the individual risk profile taking into account the severity of portal hypertension, distribution of venous thrombosis, presence of coagulopathy and thrombocytopenia, obstetric indications and the presence of oesophageal or abdominal wall/pelvic varices.

The live birth rates may be lower in patients with BCS compared to PVT (75% versus 82% after excluding first trimester pregnancy loss) but due to the low number of patients it remains difficult to interpret the results in this study. In the study by Andrade et al., all 18 pregnancies reaching 20 weeks gestation were delivered with 2 infant deaths (both preterm births). Rautou et al. reported 16/17 live births in pregnancies reaching 20 weeks gestation.

To conclude, patients with BCS and PVT after 20 weeks gestation and appropriate planning can have a reasonable expectation for delivery and successful outcomes. Preconception counselling and antenatal care with multidisciplinary input is key to achieving this goal.