improve symptoms, liver transplantation (LT) emerges as a definitive treatment with Improvement in cardiac function. Liver transplant patients' timing and proper selection are crucial and represent a challenge. We report two cases of liver transplantation in HHT with severe hepatic involvement.

Results: Patient 1 (P1) was a 47-year-old male, and patient 2 (P2) was a 37-year-old female with multiple arteriovenous malformations (AVMs). Cardiac index (CI) and cardiac output were 6.8, 9.5 (L/min/m2), and 5.8, 9.3(L/min) in P1 and P2, respectively, associated with dilated cardiomyopathy with mean pulmonary hypertension of 60 mmHg in P1 and 33 mmHg in P2. Additionally, both presented portal hypertension and ischemic biliopathy refractory to medical treatment. They also received bevacizumab one year before LT, showing marked clinical improvement. P1 was anticoagulated due to a mechanical aortic valve. After intensive diuretic therapy, the mean pulmonary pressure was 35 mmHg in P1. Natural MELD was 11 in both patients, and additional MELD was 26 and 28, respectively. Six hours of orthotopic liver transplant with cava preservation and a high-quality donor were performed. Only 1 and 3 units of red cells were transfused, respectively, with non-hemorrhagic perioperative events observed. Post-transplant complications in P1 included vasoplegic shock, splenic steal (solved with artery embolization), and reversible renal failure due to hypotension and tacrolimus, while P2 presented a mild reversible cellular rejection. Tacrolimus was prescribed for both cases due to its antiangiogenic properties. Pre LT clinical characteristics are shown in Table 1. Pre LT and Post LT hemodynamic and echocardiographic parameters are shown in Table 2. After 56 months, P1 is in good clinical conditions CI of 5.6 (L/min/m2), and low doses of diuretic requirements. P2 was discharged ten days post-transplantation and, after 43 months, is in an excellent performance with a CI of 3.31 (L/min/m2).

Conclusions: We provide data about the applicability and timing of liver transplantation in selected patients with HHT with severe hepatic involvement. According to our knowledge, this is the first Latin American report of liver transplantation in HHT. Despite the high risk of bleeding, highlight the low rate of perioperative transfusion requirements in both cases.

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P-130 CONGENITAL PORTOSYSTEMIC SHUNTS: EXPERIENCE IN A THIRD LEVEL CHILDREN`S HOSPITAL

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Introduction and Objectives: Congenital Portosystemic Shunts (CPSS) are rare vascular malformations that involve communication between the portal and the systemic venous system. Patients with this condition may be asymptomatic or present with severe complications such as hepatic encephalopathy (HE), hepatopulmonary syndrome (HPS), pulmonary arterial hypertension (PAH), or liver nodules (LN). This study aimed to share our experience in the diagnosis and treatment of patients with CPSS.

Materials and Methods: This is an observational, retrospective study including patients diagnosed with CPSS between 2011 and 2022 in our hospital.

Results: We present nine children between three months and sixteen years old at the time of diagnosis, which was incidental in four patients and due to CPSS complications in five patients: two presented HE, one HPS and two with LN (one adenoma and one focal nodular hyperplasia). According to the Bicetre classification, four cases were type I, three were type II and two were type IV. Six patients had CPSS-related congenital cardiopathies, one had polysplenia, and another patient had severe scoliosis. Two patients had genetic syndromes: Down Syndrome and Turner Syndrome. We obtained an angioCT or angioMRI in all cases; eight patients also underwent an interventionist study. Four patients underwent shunt closure; one patient was by interventionist radiology and the other three were by conventional surgery, as closure by interventionist radiology was not feasible. Both HPS and HE resolved after closure. Two of the other six patients died of cardiac complications, and none of the other patients have presented CPSS complications to date and are under evaluation for treatment strategies.

Conclusions: As CPSS is a rare condition, it is advisable to consider a high diagnostic suspicion, mainly in patients with cardiovascular malformations and/or hyperammonemia of undetermined cause. Abdominal Doppler ultrasound should be considered as a baseline study. Given the complexity of the condition, a multidisciplinary approach is recommended.

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O-1 ECONOMIC IMPACT OF LONG-TERM ALBUMIN INFUSIONS IN PATIENTS WITH DECOMPENSATED CIRRHOSIS AND UNCOMPLICATED ASCITES FROM THE BRAZILIAN PUBLIC AND PRIVATE HEALTHCARE SYSTEMS PERSPECTIVES

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Introduction and Objectives: Liver cirrhosis is among the most common liver-related causes of death and is associated with severe complications that entail a major burden for patients and healthcare systems. The ANSWER trial showed that long-term human albumin infusions (LTA) (40g twice/week for two weeks followed by 40g/week for up to 18 months) added to standard medical treatment (SMT) managed to significantly reduce mortality and disease-related complications in patients with cirrhosis and uncomplicated ascites. Assess the economic impact of implementing LTA following the ANSWER protocol in patients with cirrhosis and uncomplicated ascites in Brazil from the public (SUS) and private (ANS) healthcare systems perspectives.