SUCCESSFUL LIVER TRANSPLANTATION IN HEPATOPULMONARY SYNDROME – CASE REPORT

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Introduction

Hepatopulmonary syndrome (HPS) is characterized by the triad of liver disease, pulmonary vascular dilatation and arterial oxygenation abnormality. It occurs in 4% to 47% in patient with liver cirrhosis [1]. We describe 2 cases of HPS with recovery from hypoxaemia after liver transplantation.

Case 1

TZP, 10-year-old boy, diagnosed biliary atresia at 3 weeks old with Kasai procedure done at 3.5 months old. Eight years later he presented with cyanosis and reduced effort tolerance (ET). Clinically he was not jaundiced, but found to be cyanosed and had finger clubbing, with saturation of 84% and PaO_2 48.9 mmHg under room air. His saturation improved to 95% with 100% supplemental oxygen. Echocardiographic microbubble test was positive. Cardiac catheterization showed aortic saturation of 89% and pulmonary artery angiogram revealed multiple small arteriovenous malformations (AVM) throughout both lungs. He was diagnosed HPS type 1 and underwent liver transplantation. Saturation normalised and finger clubbing completely resolved post-operation (Figure 1).

Case 2

GCP, 13-year-old boy, previously well, presented with cyanosis and reduced ET. He was clinically cyanosed with saturation of 82%, PaO_2 56 mmHg (both room air); with finger clubbing and hepatosplenomegaly. His saturation improved to 99% with 100% supplemental oxygen. He also demonstrated platypnoea where his saturation was worse when he was in upright position (Figure 2). There were leucopenia, thrombocytopenia with normal liver function. Echocardiography showed structurally normal heart with positive microbubble test. Aortic saturation was 86%. Pulmonary arteriogram demonstrated diffuse bilateral pulmonary AVM. Liver biopsy showed bridging fibrosis, portal inflammation with mild steatosis (underlying cause uncertain). He was diagnosed portal hypertension with hypersplenism and HPS type 1. After liver transplant, saturation normalized and finger clubbing resolved.

Figure 1. Central cyanosis and digital clubbing before liver transplant. After liver transplant, cyanosis and digital clubbing completely resolved.

Before liver transplant | **After liver transplant**

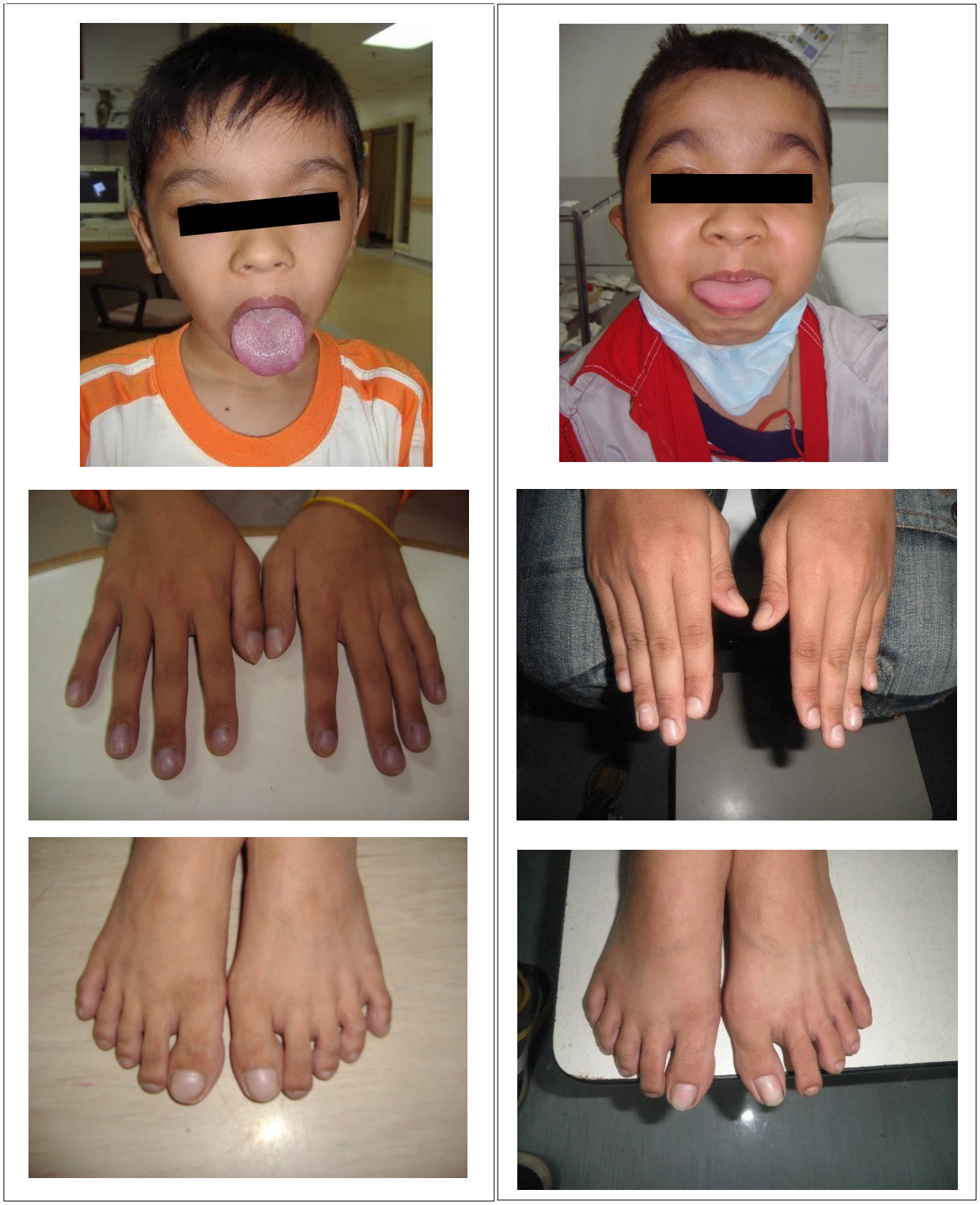

Figure 2. Oxygenation test

FiO_2	Sitting	Supine
21%	81-82%	85-86%
30%	82%	88%
40%	85%	88%
60%	87-88%	94%
100%	95%	99 %

Discussion

HPS is an important cause of respiratory symptoms in the setting of liver disease. Dyspnoea is the most common complaint, presents in 90% of all cases with mean duration of presentation 4.8 ± 2.5 years [2]. Platypnoea accompanied by orthodeoxia is highly specific for HPS. The presence of clubbing has the highest positive predictive value (75%) whilst absence of dyspnoea has the highest negative predictive value (100%) for HPS [1]. It is important to rule out other causes of dypsnoea before diagnosing HPS. HPS can develop without the advanced liver disease, and the disease may worsen irrespective of hepatic function. The precise pathophysiology of HPS remains unclear. There is intrapulmonary vasodilatation and angiogenesis, results in ventilation-perfusion mismatching, diffusion limitation to oxygen exchange, and arteriovenous shunting [3]. It is believed that nitric oxide, a potent vasodilator plays a major role in causing vasodilation. Researchers find that exhaled nitric oxide levels increase in cirrhotic patients with HPS and normalize after liver transplantation [1]. Intrapulmonary vasodilatation can occur in 2 patterns. Type 1 HPS is characterized by diffuse vasodilatation at pre-capillary level close to the normal exchange gas units whereas type 2 HPS lesions are more discretely localized and are large arteriovenous communications distant from the gas exchange units. Thus, type 1 HPS saturation improves with supplemental oxygen but type 2 HPS has poor response to oxygen. Type 2 HPS patients maybe at highest risk for complication after liver transplant [1]. Investigations required include arterial blood gas to show hypoxemia (PaO_2 <70 mmHg) and echocardiography microbubble test using intravenous injections of agitated saline to produced bubbles (in HPS, the bubbles are seen in the left heart after the third heart beat). Pulmonary angiography is performed to demonstrate the dilatation of the pulmonary vasculature and enable the identification of type 1 or type 2 HPS. Liver transplantation is the main treatment of symptomatic HPS. Some patients may benefit from other surgical or radiological interventions. Eighty- five percent of all cases showed significant improvement or complete resolution in hypoxemia after liver transplant [1]. Unfortunately, the mortality after liver transplantation is significantly increased in HPS patients with a 1-year survival rate of 71% [1].

Conclusion

HPS is one of the important cause to rule out when encounter with patient with cyanosis without apparent cardiac and respiratory disease. Liver transplantation remain the

mainstay of treatment for HPS with good respiratory outcome.

References

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[3] Hepatopulmonary syndrome: Update in recent advances in pathophysiology, investigations and treatment. Grace JA, Angus PW. J Gastroenterol Hepatol. 2013 Feb;28(2):213-9.