

Poster | 成人先天性心疾患

Poster (I-P09)

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Fri. Jul 7, 2017 6:00 PM - 7:00 PM Poster Presentation Area (Exhibition and Event Hall)

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[I-P09-02] フォンタン術後肝腎症候群にアルブミンとピトレシン持続静注使用にて腎機能改善を示唆された1症例

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The case is a 33 year old male with history of mitral atresia and TCPC conversion at 18 years old. He developed signs of failed Fontan circulation with systemic ventricular dysfunction and recurrent atrial tachycardia, leading gradually to progressive protein losing enteropathy and liver cirrhosis. He was admitted to the hospital for bacterial cellulitis, after which he developed acute renal failure which required repeated continuous hemodialysis. Clinical findings were suggestive of hepatorenal syndrome. Cardiac catheterization revealed mean PA pressure of 14mmHg, PA wedge pressure 10mmHg, Cardiac index was 6.6L/min/m², Rp 0.61 Woodunitsxm², and Rs was 4.69 Wood Unitsxm². The decreased systemic vascular resistance was compatible to the pathophysiology of hepatorenal syndrome which involves arterial vasodilatation. We initially started intravenous norepinephrine, which was not tolerated due to worsened atrial tachyarrhythmia. The patient was then started on low dose continuous vasopressin. Urine output was increased, and improvement of creatinine levels was noted during the therapy. Adverse effect was not observed. In liver cirrhosis with primary liver disease, hepatorenal syndrome is known to have high morbidity which may respond to vasoconstrictive therapy. The first recommendation is terlipressin, a vasopressin analogue which is not available in Japan. This case suggests that hepatorenal syndrome maybe an important but underdiagnosed pathophysiology in post Fontan liver dysfunction. Careful administration of vasopressin may be transiently useful in a limited setting.