

JCK Poster

JCK Poster 4 (III-JCKP4)

Cardiac Surgery

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1:00 PM - 2:00 PM

[III-JCKP4-07]A case of successful resolution of protein-losing enteropathy after conversion to pulsatile bidirectional cavopulmonary shunt from Fontan procedure.

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We report a case of a 14-year-old boy who had resolution of medically uncontrolled protein-losing enteropathy (PLE) as a complication of Fontan circulation, after Fontan take-down and pulsatile bidirectional cavopulmonary shunt (BCPS). On 30th April 2014, an 11-year-old boy was admitted to our hospital due to swelling of both legs and scrotum. He underwent Fontan procedure on 27th October 2004 because of functional single ventricle with left isomerism, Fallot-type double outlets from right ventricle and complete atrioventricular septal defect. His laboratory finding at admission showed hypoalbuminemia and elevation of stool alpha-1-antitrypsin. Thus, he was diagnosed as PLE. After then, he had been treated with sildenafil, steroid and unfractionated heparin, but these treatments were not effective. Follow-up echocardiography showed that there was moderate atrioventricular valve regurgitation. After careful discussion, we decided to perform pulsatile BCPS after Fontan take-down to control PLE. He underwent this operation on 2nd February 2016. After the operation, he showed lower arterial oxygen saturation, but symptoms and laboratory findings were improved. He is now on steroid, sildenafil and diuretics. This case suggest that surgical correction of high central venous pressure such as pulsatile BCPS might be an considerable option to control symptoms and laboratory findings of PLE as a complication of Fontan circulation. However, Fontan take-down and pulsatile BCPS result in lower arterial oxygen saturation. Thus, long term follow-up for this patient would be necessary.