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Single-center experience of hemodialysis in patients after Fontan palliation

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1. Introduction

The Fontan procedure is associated with increased venous pressures leading to long-term sequelaex including cirrhosis, protein-losing enteropathy, impaired bone development, plastic bronchitis, and renal dysfunction [1,2]. Fontan patients may require hemodialysis due to acute kidney injury (AKI) or chronic kidney disease (CKD). Fontan circulation is thought to not tolerate intermittent hemodialysis (IHD) due to its reliance on preload and large fluid shifts during IHD. At our institution, three Fontan patients have had insults to their renal function requiring IHD.

2. Cases

2.1. Patient 1

A 33-year-old male with double inlet left ventricle with atriopulmonary Fontan palliation presented with bilateral pulmonary emboli and atrial flutter with complete heart block. Transesophageal echocardiogram revealed depressed systolic function (EF 25–29%). Atrial flutter was treated with amiodarone; however, this was discontinued due to bradycardia; heart block was treated with isoproterenol and placement of single ventricular lead epicardial pacemaker. Right heart catheterization with catheter-directed thrombolysis was performed. He subsequently developed AKI with a rise in creatinine from 1.36 on admission to 3.55, thought to be due to a combination of arrhythmia, heart block, and decreased cardiac function. He was placed on continuous renal replacement therapy (CRRT) on hospital days 2–16. Ventricular function improved (EF 50–54%) with improved hemodynamics, enabling transition to IHD on day 17. This was hemodynamically well-tolerated and he was discharged on scheduled dialysis. He returned two weeks after discharge with sustained ventricular tachycardia following dialysis with hemodynamic compromise requiring brief cardiopulmonary resuscitation. He was found to have a serum potassium of 2.8, which was repleted; beta blocker therapy was initiated with eventual discharge home. He required dialysis for three more weeks, when renal function improved and patient was weaned off of hemodialysis. One year following presentation, renal function had normalized (creatinine 1.06).

2.2. Patient 2

A 35-year-old male with tetralogy of Fallot, severe tricuspid stenosis, and hypoplastic right ventricle with extracardiac Fontan palliation and complete heart block requiring permanent pacemaker presented with cardiac arrest. He required defibrillation and ten minutes of cardiopulmonary resuscitation. He subsequently developed AKI with a rise in creatinine from 0.83 on admission to 2.81. He was placed on CRRT on hospital day 2–7, at which point CRRT was converted to sustained low efficiency dialysis (SLED). SLED was continued through hospital day 11; he remained oliguric and thus IHD was started on day 14. The patient initially had a 20 mmHg decrease in systolic blood pressure during dialysis, which resolved with reduction in ultrafiltration rate. He again had hemodialysis on hospital day 16, during which he had non-sustained ventricular tachycardia, which resolved spontaneously and did not require treatment. Following this, his urine output and renal function improved; no further hemodialysis was required and he was discharged on oral furosemide. Renal function had normalized two years post-discharge (creatinine 0.88).

2.3. Patient 3

A 36-year-old female with Fontan palliation for double outlet right ventricle and subsequent failing Fontan physiology as demonstrated by hepatic cirrhosis (MELD 19) and stage III CKD (baseline creatinine 1.6) presented with AKI (creatinine 2.11). Echocardiogram demonstrated diastolic dysfunction. Despite milrinone therapy, renal function worsened with creatinine rising to 3.52. She was started on CRRT on hospital day 9. On hospital day 21 cardiac catheterization demonstrated elevated Fontan pressure of 32 mmHg and ventricular end-diastolic pressure of 30 mmHg. On hospital day 22, an Impella device was placed

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to improve hemodynamics, but she continued to have worsening oliguric kidney failure. Conventional IHD was attempted on hospital day 25 with limited ultrafiltration due to hypotension; thus she was deemed too unstable for outpatient dialysis. She was evaluated for heart/liver/kidney transplant and was determined to not be a candidate due to multi-organ dysfunction. She was discharged to palliative care and continued to receive low-volume IHD. However, she was unable to tolerate high enough ultrafiltration to maintain euvolemia and expired one month after discharge.

3. Discussion

Renal dysfunction is a common complication of Fontan palliation, with reduction in glomerular filtration rate (GFR) observed in 14% and microalbuminuria in 34% of Fontan survivors [3–6]. The primary mechanism for renal dysfunction after Fontan palliation is increased central venous pressure, leading to stasis within nephrons [5]; swine models have demonstrated that increased renal venous pressure alone causes decreased GFR and proteinuria [7]. In patients with Fontan palliation, a strong correlation has been demonstrated between superior vena cava pressure and microalbumin to creatinine ratios [5]. Other factors contributing to renal dysfunction include decreased cardiac output with renal hypoperfusion, and Fontan associated liver disease that progresses to hepatorenal syndrome [1,5,8].

We report a series of three patients with Fontan palliation with renal dysfunction requiring hemodialysis. One patient expired after hospitalization due to fluid overload and an inability to tolerate IHD, while two others have done well after discharge. Both Patient 1 and Patient 2 were relatively healthy without failing Fontan physiology prior to presentation, and both of these patients tolerated IHD with minimal difficulty. Patient 3 had extensive extra-cardiac disease, with Fontan-associated liver disease and CKD, and was unable to tolerate IHD. Notably, one further patient at our institution, a 54-year-old female with Fontan palliation, required CRRT post-operatively after conversion from atriopulmonary to extracardiac Fontan. She, too, had little extracardiac disease and normal ventricular function. She tolerated nine days of CRRT until renal function improved and she was weaned off dialysis. While it has historically been thought that patients with Fontan palliation cannot tolerate the rapid fluid shifts in hemodialysis, we present the cases of two patients who tolerated IHD.

The success of Patients 1 and 2 must be presented with a caveat. Both patients had episodes of ventricular arrhythmias during dialysis. Given their propensity for arrhythmia, it may be prudent to observe patients with Fontan palliation in an inpatient setting when initiating hemodialysis. Given Patient 1's hypokalemia, it may be wise to consider a higher potassium dialysate content in these patients as well.

Our study was limited by its small size and its retrospective nature. However, IHD may be tolerated in patients with Fontan palliation, particularly in those without chronic Fontan failure and significant extracardiac organ involvement. Further experience is necessary in order to determine what parameters predict successful IHD in patients with Fontan palliation, and whether long-term IHD is viable for these patients.

References

[1] M. Mori, et al., Beyond a broken heart: circulatory dysfunction in the failing Fontan, Pediatr. Cardiol. 35 (4) (2014) 569–579.
[2] G. Grutter, D.C., D., F. Gandolfo, R. Adorisio, S. Alfieri, G. Michielon, A. Carotti, G. Pongiglione, Plastic bronchitis after extracardiac Fontan operation, Ann. Thorac. Surg. 94 (2012) 860–864.
[3] S.R. Sharma, R., S. Furth, K. Dodds, J. Rynchik, D. Goldberg, Assessment of kidney function in survivors following Fontan palliation, Congenit. Heart Dis. 11 (6) (2016) 630–636.
[4] D.G. Lee, J, Prevalence of CKD in adult Fontan, Can. J. Cardiol. 31 (2015).
[5] P. Anne, et al., Nephropathy in patients after Fontan palliation, Int. J. Cardiol. 132 (2) (2009) 244–247.
[6] R. Alexander, F.B. Opotowsky, Michael Landzberg, Fred Wu, Finnian McCausland, Justin Owumi, Brittani Loukas, Venkata Sabbisetti, Elizabeth Landzberg, Sushrut Waikar, Kidney dysfunction in patients with single ventricle Fontan, Am. Coll. Cardiol. 67 (13) (2016) 898.
[7] J.M.S. Doty, H. Bob, Harvey J. Sugerman, Charles R. Blocher, Richard Pin, Itaf Fakhry, Todd W.B. Gehr, Domenic A. Sica, Effect of increased renal venous pressure on renal function, J. Trauma 47 (6) (1999) 1000.
[8] F. Sammour, et al., Renal function of patients with a failing Fontan circuit undergoing total cavopulmonary revision surgery, Pediatr. Cardiol. 30 (3) (2009) 282–288.