Peritoneal Dialysis for Refractory Heart Failure from a Congenitally Corrected Transposition of the Great Arteries who has not Undergone Definitive Surgery: A Case Report

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ABSTRACT

Heart failure (HF) is a major cause of significant morbidity, mortality, and hospitalization worldwide including the Philippines. Congenitally corrected transposition of the great arteries (C-TGA) occurs when the right atrium enters the morphological left ventricle which gives rise to the pulmonary artery and the left atrium communicates with the right ventricle which gives rise to the aorta. Heart failure can occur in C-TGA especially if associated with other heart defects. Ideal management is anatomic correction via surgery to prevent or address heart failure. Peritoneal dialysis has been used as a therapeutic intervention for patients with refractory heart failure and kidney injury with or without kidney failure due to its gentler fluid removal compared to conventional ultrafiltration resulting in less myocardial stunning and neurohormonal activation. We present the case of a patient with heart failure who started on peritoneal dialysis (PD) as an adjunct therapy for fluid management after failing to satisfactorily achieve volume control with diuretics.

The patient is a 56-year-old man with C-TGA admitted for decompensated heart failure. He was initially treated with intravenous diuretics on the first admission but was readmitted after 3 months for decompensation this time with borderline low blood pressure making diuresis difficult. The patient was given loop diuretics, tolvaptan, and angiotensin receptor neprilysin inhibitor (ARNI) but still with decreasing trends in urine output and inadequate symptom control. PD was initiated before discharge with subsequent improvement in heart failure symptoms. The patient was on regular follow-up for PD maintenance and titration of heart failure medication.

In this case report, we have shown how PD can be an effective adjunct to guideline-directed medical therapy in patients with severely symptomatic heart failure who have an unstable hemodynamic status and for which volume management cannot be satisfactorily achieved with diuretics.

Keywords: peritoneal dialysis, heart failure, congenital heart disease, congenitally corrected transposition of the great arteries, diuresis, ultrafiltration

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BACKGROUND

Heart failure (HF) is a major cause of significant morbidity, mortality, and hospitalization worldwide with an estimated prevalence of 26 million people affected.[1] In the Philippines, the prevalence is 1–2% accounting for 9% of total hospitalizations and 7% of inpatient mortalities.[2] Congenitally corrected transposition of the great arteries (C-TGA), also called L-transposition, is a rare disease comprising < 1% of congenital heart diseases although some cases are only identified in adulthood since it may not present with heart failure symptoms until then.[3] It is characterized when the right atrium enters the morphological left ventricle which gives rise to the pulmonary artery whereas the left atrium communicates with the right ventricle which gives rise to the aorta. This is called atrioventricular and ventriculoarterial discordance. C-TGA can be associated with a ventricular septal defect (VSD), pulmonary stenosis, tricuspid (systemic) valve abnormalities, and conduction system abnormalities all of which affect a patient's clinical presentation and need for surgery.[3] Symptoms and declining cardiac function require surgical management most notably an anatomic correction surgery wherein the morphologic left ventricle and mitral valve are restored into their systemic roles.[4] Repair of associated defects (i.e. tricuspid valve replacement in severe tricuspid regurgitation with dilation or mild dysfunction of the systemic ventricle) and palliative procedures can also be done.[5] Orthotopic heart transplantation is done in cases with failure of the systemic ventricle (the ventricle leading to the systemic circulation) even with prior surgical repair.[5] There are no recommendations for specific medical therapy in patients with a systemic right ventricle exhibiting systolic dysfunction. Patients with adult congenital heart disease often receive pharmacologic therapy specific to their associated conditions (i.e., beta-blockade in patients with arrhythmias).[6] In cases of acute decompensation of an existing cardiac condition causing volume overload and kidney injury (cardiorenal syndrome type 1), extracorporeal therapies such as ultrafiltration with or without hemodialysis can be offered when diuretics fail but not without complications to renal function, vascular access, and cardiovascular outcomes.[7] Peritoneal dialysis has been used as a therapeutic intervention for volume control in refractory heart failure since the 1940s. Single daily peritoneal exchanges have been used to achieve adequate fluid removal in patients with heart failure but with no overt need for renal replacement therapy.[8] Peritoneal dialysis (PD) offers numerous advantages compared to usual extracorporeal renal replacement therapies (i.e., ultrafiltration and/or hemodialysis): gentler ultrafiltration avoids myocardial stunning, has minimal impact on hemodynamics resulting in lesser neurohormonal activation, demonstrates a slower decline in residual kidney function, provides continuous solute clearance with better titration of heart failure medications, and can be conveniently done at home.[9] In this case, report, we present a case of a patient with heart failure who has been started on peritoneal dialysis as an adjunct therapy for fluid management after failing to satisfactorily achieve volume control with diuretics.

CASE PRESENTATION

The patient is a 56-year-old man diagnosed with congenitally corrected transposition of the great arteries when he was 41 years old. He presents with heart failure symptoms (bipedal edema, shortness of breath, and easy fatigability) for which he was taking diuretics to relieve symptoms. He is pre-diabetic but non-hypertensive with no other comorbidities. His pertinent surgical history was an appendectomy in 1976. He was working as a marketing and engineering executive and was a former heavy alcohol drinker and smoker. On August 3, 2019, he presented with a two-month history of worsening heart failure symptoms now including orthopnea unrelieved with oral diuretics. There was no associated chest pain or palpitations. He was started on dobutamine and intravenous diuretics. Symptoms gradually improved and diuretics were titrated before he was discharged.

The patient was re-admitted on November 22, 2019, after another bout of worsening heart failure symptoms now

Table 1. Laboratory and Diagnostic Test Results

	Admission	2 months post-discharge	5 months post-discharge	Normal values
BUN *(mg/dL)*	34.4	39.09	39.93	8.96-19.88
Crea *(mg/dL)*	1.5	1.28	1.56	0.66-1.24
Na *(mEq/L)*	138	136	138	137-145
K *(mEq/L)*	3.3	3.8	3.8	3.5-5.1
Ionized Ca *(mg/dL)*	3.85	-	-	4.8-5.3
Mg *(mg/dL)*	2.19	-	-	1.7-2.43
Alb *(g/L)*	31	-	46.8	35-50
Uric acid *(mg/dL)*	-	10.4	11.72	3.5-7.2
FBS *(mg/dL)*	88.2	112	102	80-100
Total cholesterol *(mg/dL)*	-	203.1	-	<200
Triglycerides *(mg/dL)*	-	97.5	-	<150
Low density lipoprotein *(mg/dL)*	-	128.6	-	100-127
High density lipoprotein *(mg/dL)*	-	55	-	-
Hemoglobin *(g/L)*	146	147	159	130-150
Hematocrit	0.44	0.46	0.46	0.4-0.54
WBC *($x10^9/L$)*	5.9	4.2	5.23	4.5-11.0
Lymphocytes *(%)*	8	49	16	20-50
Neutrophils *(%)*	78	41	69	50-70
Platelets *($x10^9/L$)*	277	249	236	150-450
NT pro-BNP *(pg/mL)*	-	13827.96	2650.84	0-125

associated with ascites, nonproductive cough, and dyspnea at rest. Pertinent laboratory and diagnostic tests throughout the patient's course are seen in Table 1. Chest radiograph (Figure 1) showed biventricular enlargement which was confirmed via 2D echocardiography (Figures 2 and 3). Other findings in the 2D-ECHO showed *situs solitus* with atrioventricular discordance and ventriculoarterial discordance, eccentric left ventricular hypertrophy with generalized hypokinesia and depressed systolic function, ejection fraction (M-mode) = 30%, eccentric right ventricular hypertrophy with adequate contractility, biatrial dilatation with increased volume indices mild aortic regurgitation, tricuspid valve vegetation versus torn chordae, dilated tricuspid valve annulus with severe tricuspid regurgitation, mitral valve sclerosis, dilated mitral valve annulus with moderate mitral regurgitation, and severe pulmonary hypertension.

Figure 1. Upright PA chest radiograph showed biventricular cardiomegaly. There are no other significant chest findings.

He was started on inotropic support, intravenous furosemide drip, bumetanide, and tolvaptan. He was dependent for almost two weeks on a furosemide drip to achieve adequate diuresis and symptom relief (Figure 4). By the start of the 2nd hospital week, loop diuretics were discontinued, tolvaptan continued, and sacubitril + valsartan was started. Nearing his third hospital week, diuresis was decreasing on top of blood pressures remaining low or borderline low-normal. The patient was still symptomatic at that time. Because of the unstable blood pressure despite the down titration of diuretics, peritoneal dialysis was initiated three days before discharge. He was placed on a customized peritoneal dialysis prescription which consisted of the following: 6 exchanges per day using 4.25% solution at 1.5 liters per exchange with 30 minutes dwell time. The patient was sent home improved on sacubitril + valsartan and peritoneal dialysis.

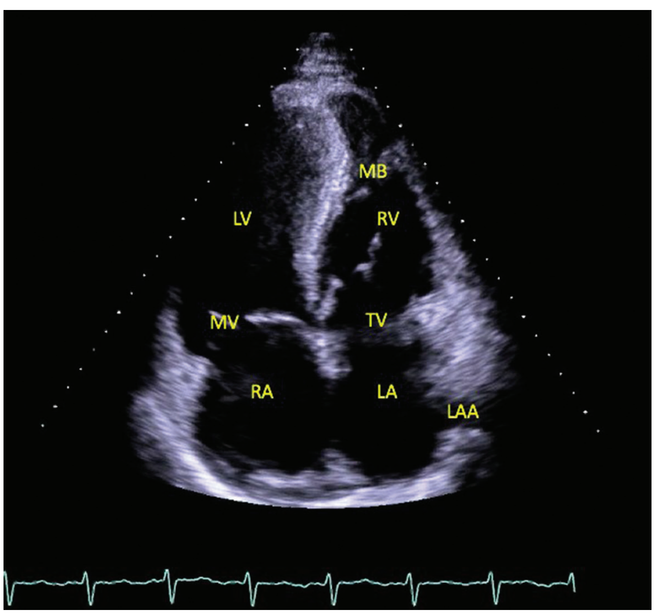
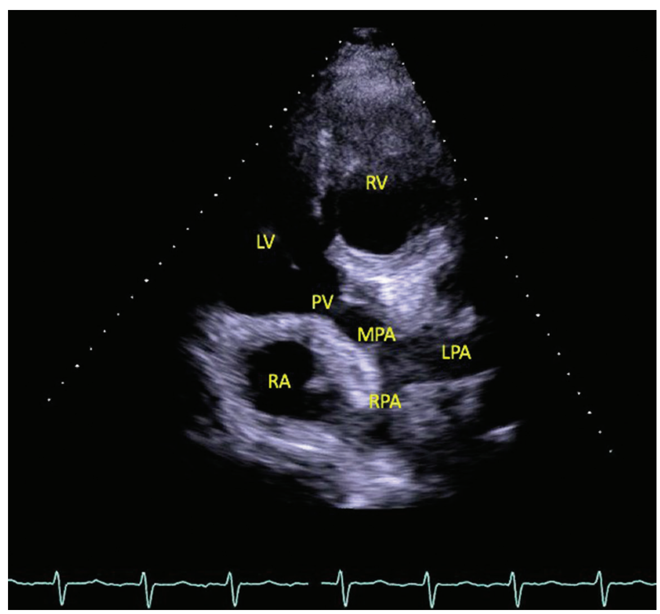


Figure 2. Modified apical four-chamber view to assess the right outflow tract showing continuity of the right-sided morphologic left ventricle to the pulmonic valve.

LA, left atrium; LAA, left atrial appendage; LV, left ventricle; MB, moderator band; MV, mitral valve; RA, right atrium; RV, right ventricle; TV, tricuspid valve.



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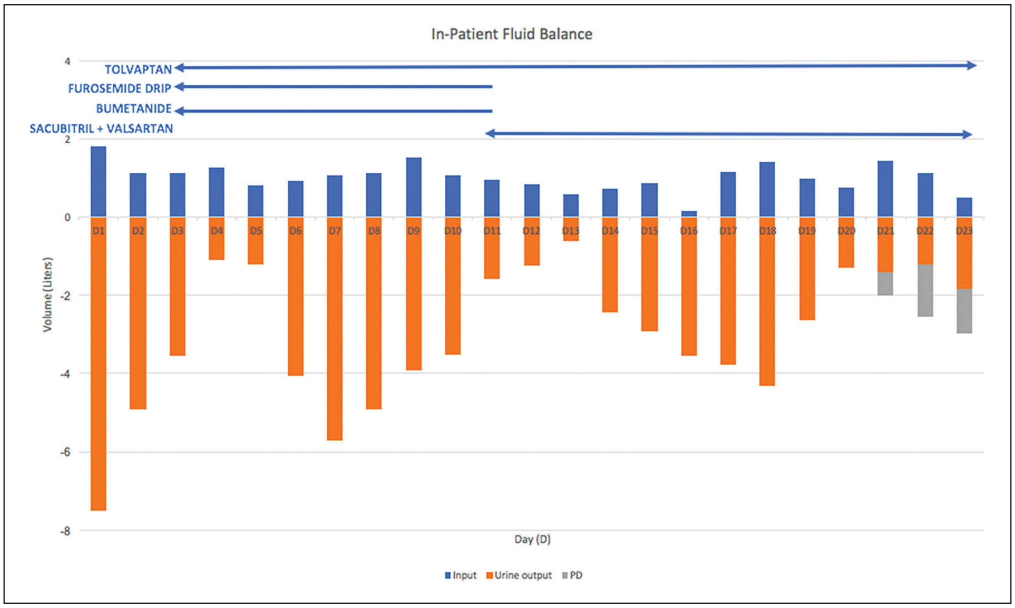

Figure 4. ***In-patient fluid balance.*** There is a note of a decrease in urine output after discontinuation of intravenous loop diuretics. Peritoneal dialysis was started three days before discharge.

Outcomes and Follow-up

The patient was on regular follow-up with both nephrology and cardiology services and was being maintained on sacubitril + valsartan 100 mg per tablet 1 tablet twice a day. No diuretics were being currently given. Laboratory test results on follow-up are shown in Table 1. His follow-up 2D-ECHO showed a decrease in the degree of pulmonary hypertension (mild-moderate) and a slight increase in EF (M-mode) from 30% to 40%.

He was maintained on peritoneal dialysis using 4.25% concentration solution bags, 2.2 L per bag, three exchanges per day at 30- to 60-minute dwells per exchange. The number of exchanges was adjusted to achieve his dry weight of 56 kilograms. The current average fluid balance was maintained at -1048.5 mL since the patient was started on peritoneal dialysis. Since starting peritoneal dialysis, the patient has been able to achieve a comfortable dry weight with the above prescription (Figure 5).

DISCUSSION

In this case report, we have shown how peritoneal dialysis can be an effective adjunctive therapy in patients with severely symptomatic heart failure who have an unstable hemodynamic status and for which volume management cannot be satisfactorily achieved with diuretics. Surgery is the ideal management for C-TGA but was not done for this patient. Medical management, despite a lack of definite recommendations from the latest available guidelines, to address his specific conditions (fluid overload), were done instead and this included volume control with diuretics and PD. He was discharged with improvement in symptoms. Also noted was an improvement in his ejection fraction from the time of his first admission to three months post-discharge. On out-patient follow-up, he was noted to have less symptoms of heart failure (dyspnea, easy fatigability). Peritoneal dialysis has been used for refractory heart failure patients with a wide range of kidney functions.[10] Outcomes from these observational studies included improved ejection fraction,[10] improved New York Heart Association (NYHA) functional class,[11-16] lesser hospitalizations, and shorter hospital length of stay.[17-19] In a recent prospective trial, it has been shown that PD patients with acute decompensated heart failure had better renal function, improvement in cardiac function, and net fluid loss versus patients who underwent ultrafiltration.[20] Locally, the use of PD instead of UF in patients with ADHF with or without renal failure is uncommon and there is no published study, to date, reporting such therapeutic strategy.

The prohormone brain natriuretic peptide (BNP) is produced by ventricular cells in response to high ventricular filling pressures with the overall effect of natriuresis and a decrease in blood pressure.[21] Studies suggest that it has a role in regulating blood pressure and volume balance as well as a marker of left ventricular function.[21,22] Its cleavage product (NT-pro-BNP) has been used to predict mortality and hospitalization in heart failure patients.[22] In patients with end-stage kidney disease on hemodialysis, BNP levels correlate with the presence of left ventricular hypertrophy (LVH) with or without congestive heart failure.[21] No study has yet measured NT-pro-BNP levels in patients with HF on PD for volume management. The decrease in the NT-pro-BNP levels in our patients can be due to better volume homeostasis afforded by peritoneal dialysis. Whether this

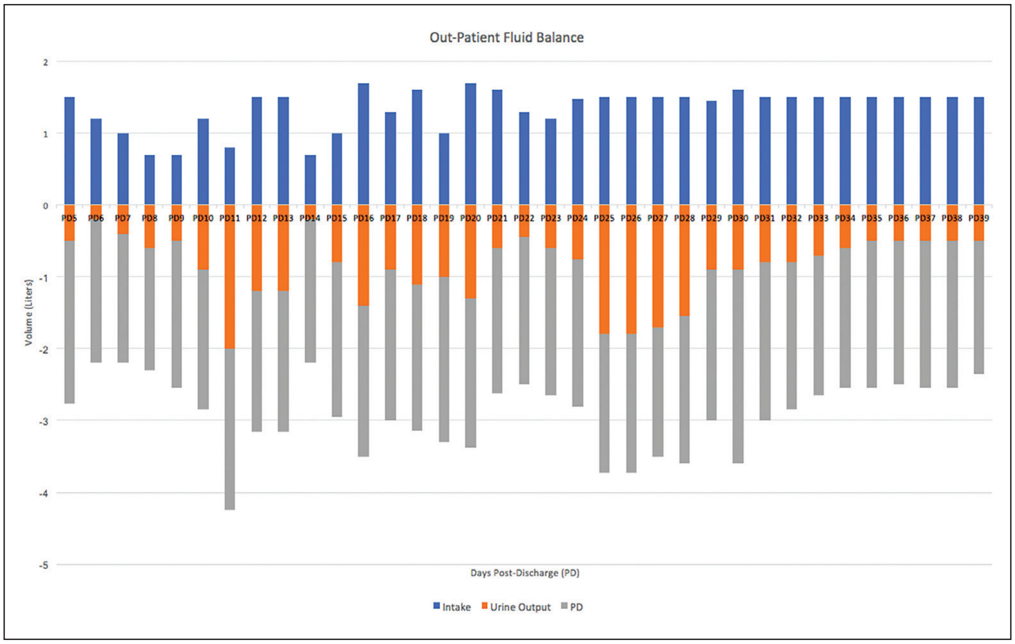

Figure 5. ***Out-patient fluid balance.*** Peritoneal dialysis fluid balance *(grey bars)* has been shown to significantly improve output with eventual decongestion and relief of failure symptoms, augmenting his heart failure medications.

translates to more useful clinical outcomes such as mortality and symptom relief can be the subject of future studies.

Decreased cardiac output subsequently causes decreased intravascular volume and renal perfusion and traditionally explains the pathophysiologic process of worsening renal failure in cardiorenal dysfunction.[23] It has been found that in recent studies venous congestion from "backward failure" and increased intra-abdominal pressures may also play roles in the process.[24] Venous congestion, measured via central venous pressures (CVP) on admission and during hospitalization, was the greatest determinant for the development of renal insufficiency more than the cardiac index (CI) in a study by Mullens and colleagues.[24] Increases in CVP have been linked to increased renal vein pressure leading to the phenomenon of "congestive kidney failure" in the setting of ADHF.[25] Addressing venous congestion and increased intra-abdominal pressure will subsequently improve renal perfusion both of which can be theoretically addressed by PD.[26]

Peritoneal dialysis is a feasible therapeutic option in HF patients who are diuretic-resistant or in those who have labile hemodynamic profiles such that usual guideline-directed medical therapy (GDMT) cannot be maximized. Due to the sustained and gentler decongestion afforded by PD, this therapy can be used to maintain euvolemia in HF patients and may even help in restoring diuretic responsiveness.[26] Peritoneal dialysis can be a useful bridge towards more definitive plans for HF management (i.e., surgery) or it can be a means to palliate symptoms and improve functional status.[26] Various prescriptions can be employed in patients who do not require full dialytic support. Among the commonly used ones are nocturnal automated PD wherein peritoneal dialysis is done with the help of a machine called a cycler to maximize fluid removal while the patient sleeps[27,28] or a single night time exchange using icodextrin, a kind of PD solution that can, in most cases, remove more fluid from a patient compared to using other types of PD solution in the same setting (single night time exchange).[29] Both techniques are relatively simple and can be easily carried out at home in healthcare settings where PD is readily accessible.

CONCLUSION

We have presented a case of heart failure in an adult man in whom PD was initiated to supplement HF management after presenting with unstable hemodynamics and unsatisfactory response to diuretics. Despite the lack of randomized controlled trials exploring PD use in HF, there are already numerous observational studies on the use and benefits of PD in the treatment of HF patients with varying degrees of renal function. Cardiologists and nephrologists should not hesitate to offer this to their patients wherein alleviation of symptoms, reduction in hospital re-admissions, and subsequent improvement of quality of life are meaningful goals.

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Statement of Authorship

All authors contributed in the conceptualization of work, acquisition and analysis of data, drafting and revising and approved the final version submitted.

Author Disclosure

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