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Case Report

Management of Terminal Stage CHF Due to Shones Syndrome

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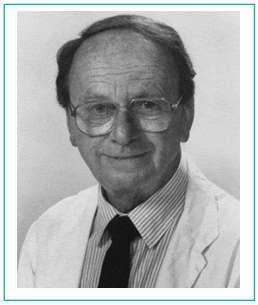

Abstract

Background: we report a case of a 23-year-old male with CHF due to Shone's syndrome s.p multiple open-heart surgeries, aggressive medical therapy, and implanted CRT-D device. He has been managed by pediatric and adult cardiologists, cardiovascular surgeons, nephrologists, and general internists for many years. He came to us with acute respiratory failure, acute kidney injury and fragility due to advanced stage of CHF with reduced ejection fraction. After decent attempts trying to treat him, eventually the decision of transition to hospice care was made.

Case Report: He was started furosemide 120 mg IV; the plan was to continue him on diuretics with temporal improvement recorded. As per his Pediatric cardiologist he is an advanced case of CHF with too frequent exacerbations and hospital admissions, and not a candidate of Heart transplant because of guarded prognosis It was also noted that the patient still had frequent hospitalizations with CHF exacerbations complicated with AKI despite maximum medical therapy. With Palliative care, pediatric cardiologist and Nephrologist all involved, the ultimate decision made to discharge home under strict hospice care plan which will be taken care by a home visit nurse provided thought hospice care Agency.

Conclusion: The report shed lights on shone syndrome related CHF, the course of the disease, treatment options and ultimate goals of care. In patients who continues to be symptomatic despite maximum medical and surgical therapy, the decision of implementing more invasive treatment options will greatly depend on goals of care and overall weight of risk vs. benefits.

Introduction

Born and raised in UK, Dr. John Desmond Shone initially served as a soldier during world war before he joined school of medicine and graduated from university of London in 1953 and immigrated to North America in 1957 to practice as a general pediatrician. Under supervision of Dr Jesse Edward, he noticed series of congenital heart defects including "parachute mitral valve", supra valvular ring of the left atrium, subaortic stenosis, and coarctation of the aorta, he then described "Shone" syndrome with help of Dr Edward [1]. Paucity of literature description of the condition contributes to the deficiency of overall management recommendations of the condition [2]. Despite being congenital anomaly with relatively short life expectancy, very rare cases are reported in adults, the older I found is a case of Incomplete Shon's complex in a 55-year-old woman [3]. Common manifestations are Variants of Heart failure syndromes, arrhythmias, and resistant hypertension.

Case Report

Past Medical History

Immediate post-delivery assessment showed evidence of a group of congenital cardiac defects, including coarctation of the aorta, bicuspid aortic valve, a large mid muscular ventricular septal defect, and large secundum atrial septal defect. His ventricular septum was posteriorly deviated. Three days after delivery, his coarctation of the aorta was repaired with a subclavian flap, and in a month, he underwent surgical repair of a muscular ventricular septal defect and a secundum atrial septal defect with no evidence of residual defects.

At 16 months, an aortic valvotomy and myectomy was done, and at 6-1/2 years, balloon dilation of the aortic valve was done. The gradient was reduced from 60 down to 20 mmHg. With another Balloon dilatation done at 13 years old, he then developed complete heart block with tem-

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porary pacing lead which resolved after approximately six days. In 2014, he was admitted with the first ever exacerbation of congestive heart failure treated with Lasix. His Echo showed systolic dysfunction LVEF=31.4%, CHF improved on milrinone, lisinopril, digoxin, and diuretics to LVEF=45%. At the age of 16 years, he underwent aortic valve replacement with a 25 mm St Jude valve and complete heart block requiring placement of a tri-chamber biventricular pacemaker. Further genetic investigation was discussed as there may be a genetic component of his cardiomyopathy, whole genome microarray was normal, whole exon sequencing recommended however additional studies have not been pursued.

At 18 years, admitted for congestive heart failure, and at 21 years admitted again with symptoms of congestive heart failure with shortness of breath and grunting. LVEF was 16.6%. Lisinopril, diuretics, and metoprolol have been titrated up. At 21 years, 8/20/2020, seen at UW health heart failure clinic, Mom's question was regarding heart transplant and process that he will go through if he requires that. Stress test was recommended as well as increasing his metoprolol to 50 mg daily because his blood pressure was 124/60 the day he was seen. This suggests that he may tolerate a higher dose of metoprolol. LVEF was slightly improved to 28.4% and stress test, He tolerated around 6 minutes, getting up to a brisk walk. No evidence of ischemia, arrhythmia, or chest pain. He stopped early because he was tired.

Clinical Presentation

23-year-old male with a complicated cardiac history including congenital heart disease, Shone's syndrome with multiple surgeries in the past, CHF with reduced ejection fraction (20-25%), diabetes mellitus type 2, sleep apnea on CPAP, factor V Leyden deficiency on Coumadin, cognitive impairment, chronic hypoxemic respiratory failure on 4 L/min of oxygen at rest and 6 L with activity, as well as other comorbidities who was just recently admitted from 10/10-6 10/14, and again October 23 through November 3 for acute on chronic heart failure, acute on chronic respiratory failure, acute kidney injury, presented to the ED with complaints of weight gain and shortness of breath. In the ED his labs notable for creatinine 1.8, BUN 107, alkaline phosphatase 135, AST 81, bilirubin 2.2, BNP 790. Chest X-ray showed pulmonary congestion. Troponin was elevated at 400s. EKG showed Ventricular paced, rate 103, PR 192, QRS 176, QTc 555, like prior. CXR done with an impression of Suspect CHF exacerbation. Cardiology was consulted and that he was not a transplant candidate, with symptom management being recommended, and was also seen by nephrology for discussion of potentially starting dialysis for his compromised renal function, diuretics therapy initiated.

He was started furosemide 120 mg IV, his ABGS show metabolic alkalosis therefore 250mg of acetazolamide was given. A strict monitoring of his input output and weight. He was fluid restricted, and - Due to AKI we held Entresto. His acute-on-chronic respiratory failure with Hypoxia was thought due to pulmonary edema secondary to heart failure decompensation, CKD along with obstructive sleep apnea on top of obesity hypo ventilatory syndrome and underlying congenital heart disease. He was placed on currently of 5-6L oxygen via nasal cannula to keep saturations above 92%. His Acute kidney injury was attributed to cardiorenal syndrome, the plan was to continue him on diuretics with temporal improvement recorded. A discussion about the potential need of Dialysis was going on with the nephrologist but the ultimate decision was no dialysis recommendations as AKI improved as well as poor prognosis of his underlying multiple comorbidities.

He overall did well from a diuretic standpoint and renal function continues to improve. After that his oxygen needs and Lab parameters remained static, with his shortness slowly improved. He had been having constipation for which he was prescribed bisacodyl, MiraLAX and senna with good subsequent bowel movements. His Potassium level was found to be below level likely due to excessive diuretics use, this was corrected using KCL. He also had a SIR'S alert with Tachycardia, Tachypnea and hypothermia, Likely misinterpretation of his Hemodynamics response to the CHF state, managed conservatively.

Towards the end of his hospital course plans regarding disposition were discussed. As per his Pediatric cardiologist he is an advanced case of CHF with too frequent exacerbations and hospital admissions, and not a candidate of Heart transplant because of multiple comorbidities, poor expected out come and the underlying genetic disease with overall reduced life expectancy. It was also noted that the patient still had frequent hospitalizations with CHF exacerbations complicated with AKI despite maximum medical therapy. As per cardiology consult, this patient is high risk of having ventricular arrhythmias, he was offered upgrading of his CRT-P to CRT-D but the mother opted to decline. With Palliative care, pediatric cardiologist and Nephrologist all involved, the ultimate decision made to discharge home under strict hospice care plan which will be taken care by a home visit nurse provided thought hospice care Agency. Hospice care drugs recommended by palliative care team added to his medication's reconciliations. Throughout hospitalization, his chronic diseases managements were all addressed appropriately, his blood glucose was little bit difficult to be controlled with long-acting insulin, pre meal act rapid insulin and as needed sliding scale, his warfarin dosages were appropriately managed according to INR. With discharge, we held it as the INR was found to be 4.7, the decision of resuming it would be deferred to hospice care. We also stopped Entresto, Metoprolol and metolazone because of hemodynamics and philosophy of hospice. Spironolactone was added. Other home medications remained unchanged. Significant time spent with the patient and his family trying to explain the elements of hospice care, mother appeared deeply sad. Efforts made to support her with counselling, sympathy and providing answers for her questions. 2 weeks later, he passed away peacefully at home.

Discussion

Although theoretically this patient might be a good candidate of heart transplant having persistent New York Heart Association (NYHA) functional class IV Heart Failure (HF) symptoms refractory to optimal medical and surgical therapy (including use of CRT), it wasn't recommended for this patient. The decision of whether a patient with advanced HFrEF a candidate for invasive treatment modalities should take into consideration the overall health status, cardiac history, concurrent comorbidities, Life expectancy and quality. This patient was clearly of poor overall health status with multiple complex cardiac history, despite having many corrective cardiac surgeries and an implanted CRT-P device with maximized dose of diuretics up to the level of getting AKI and contraction alkalosis, he continued to be a regular guest to emergency departments and hospitals medical wards. In the last year of his Life only, he was admitted to hospital on five different occasions with acute exacerbation of CHF. Furthermore, this patient had a baseline cognitive impairment,

it wouldn't be surprising if he continued to be dependent on others care even if his cardiac condition managed to be solved. Overall risk Vs benefit was in favor of comfort-based care rather than invasive treatment modalities. This decision was made with contributions of adult cardiologist, pediatric cardiologist, nephrologist, and primary hospital team.

Conclusion

Shone syndrome is a rare form of congenital heart diseases in which early surgical intervention might be of survival benefit. In patients who continues to be symptomatic despite maximum medical and surgical therapy, the decision of implementing more invasive treatment options will greatly depend on goals of care and overall weight of risk vs. benefits. If expectations are not great enough, invasive treatment options would not be recommended over comfort-based care.

Author Statements

Acknowledgement

I wanted to take a moment to acknowledge and express my sincere appreciation for patient's mother for her cooperative attitude towards the team. Her deep understating of the importance of reporting this rare condition for scientific and educational purposes was crucial for us to continue working on the report. I would like also to express my appreciation for my team worker, my supervisor and Marshfield clinic department of internal medicine and the residency program. I would also express my tributes to my supporting family and who ever put and effort helping or motivating.

Financial Support

Here to declare that our Manuscript didn't cost us a lot so far. But in case something comes up in the future, we will use our annual educational allowance granted by Marshfield clinic. With this financial support, we will be able to cover the costs associated with publications, presentations, and travel expenses for conferences where we can disseminate our findings. We will be truly grateful for this support, as it will allow us to delve deeper into our topic objectives and ultimately advance the understanding of treatment of Shone's syndrome. We look forward to sharing our outcomes and making a meaningful impact in the scientific community and beyond.

Conflict of Interest Statement

I (Ali Elkhedr,) hereby declare that I have no financial or personal conflicts of interest that could potentially bias the outcomes or findings of the case report (Management options of Terminal stage CHF due to Shones syndrome in a 23 Y old patient) . I affirm that I have no financial relationships with any organizations or individuals that could be perceived as having a direct interest in the Manuscript, such as pharmaceutical companies, medical device manufacturers, or any other entities that may benefit financially from the research results. Furthermore, I confirm that I have no personal relationships, such as familial, romantic, or close friendships, that could potentially influence my objectivity or create a conflict of interest. I understand the importance of maintaining integrity, transparency, and impartiality in the Manuscript writing, and I pledge to disclose any potential conflicts of interest that may arise during this process. Should any conflicts of interest arise in the future, I will promptly disclose them to all relevant parties, including my team, funding agencies, and any publications or presentations related to the manuscript. By signing this statement, I affirm my commitment to upholding the highest standards of ethical conduct and ensuring the integrity and credibility of the research project."

Patient Permission/Consent Declarations

I participated in management of this patient last year and I took verbal permission form his mother that we may scientifically write/report it for educational purposes. She is also ok with giving us written signed consent to publish/present it for scientific and educational purposes.

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