Case Report

Idiopathic dilated cardiomyopathy as cor bovinum in infancy: "A foggy road in winter"

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ABSTRACT

Dilated cardiomyopathy, when diagnosed in infancy, poses an array of difficulties from reaching an etiological diagnosis to prognosticating the long-term outcome. Here, we report a case of idiopathic dilated cardiomyopathy in a 6-month-old child who responded well to beta-blocker (Carvedilol) in optimum dosage and revealed favorable cardiac remodeling over 6 months with substantial improvement in ejection fraction (EF) (EF of 22–44%) with significant amelioration of child's symptoms. Our case has a unique message that while treating idiopathic dilated cardiomyopathy (DCM) in infancy, optimized use of the beta-blockers is most often the only way to clear the foggy road of idiopathic DCM and obtain a favorable outcome.

Key words: Ejection fraction, Idiopathic dilated cardiomyopathy, Infancy

To ascertain the etiology of dilated cardiomyopathy (DCM) in infancy is always a puzzle to solve. After ruling out a history of viral myocarditis, echocardiographic exclusion of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), and ruling out a reversible cause of cardiomyopathy in the form of hypothyroidism and selenium deficiency, a battery of investigations emerges in solving the puzzle. Although genetic study sometimes points at a definite diagnosis, it does little help so far as the prognosis is concerned. Most often physicians stick to the age-old favorite "Idiopathic dilated cardiomyopathy." Although diuretics, 0 angiotensin-converting enzyme (ACE) inhibitors, and aldosterone antagonists constitute the major armamentarium, beta-blocker (Carvedilol) stands as the cornerstone in those cases [1]. Beta-blockers provide rest to the ailing heart, bring out a favorable ventricular remodeling, prevent sudden cardiac death in children by stabilizing the membrane and thereby preventing the occurrence of ventricular fibrillation and immediate cardiac arrest [2]. Optimizing Carvedilol in the dose of 0.05 mg/kg/dose–0.4 mg/kg/dose twice daily achieves favorable ventricular remodeling and brings out a better outcome [3]. Multivitamin and levocarnitine supplementation although prescribed by a few physicians lacks definite literature data in achieving a better outcome.

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CASE PRESENTATION

An 8-months-old male child with a body weight of 6 kg presented to the cardiology outpatient department with the chief complaint of poor feeding, forehead sweating, and failure to thrive. He was born out of non-consanguineous marriage and his perinatal history was uneventful. The mother noticed difficulty in feeding 3 months back and the baby was able to satisfy his hunger after one hour of sucking with the suck-rest-suck cycle. Chest X-ray revealed the presence of a big "Wall to Wall heart" suggestive of Cor Bovinum (Fig. 1). Electrocardiogram (ECG) revealed the presence of high voltage in precordial leads and poor R wave progression from V_1 to V_4 (Fig. 2). The child was not anemic, euthyroid and all the serum chemistries were within the normal limit including blood sugar and serum calcium. Echocardiography revealed a significantly enlarged globular left ventricle (LV) (Fig. 3) with severe left ventricular systolic dysfunction (ejection fraction or EF 22%). There was no mitral regurgitation (MR) (Fig. 3), pulmonary arterial hypertension (PAH), pericardial effusion, and left ventricular apical thrombus. There was D shaped right ventricle (Fig. 3) due to bulging of interventricular septum toward right ventricle. The bulging was because of the high left ventricular end-diastolic volume. Echocardiography also ruled out the presence of anomalous origin of the coronary artery from pulmonary artery as they were having a normal origin. There was no papillary muscle calcification, dilatation of right coronary

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artery, and no reverse flow from coronary to pulmonary artery in parasternal short-axis view. The child was on enalapril, diuretic, levocarnitine, multivitamin, and calcium supplementation prescribed from the local physician for the past 3 months. Previous chest X-ray and echocardiography done by the local physician revealed the same as huge cardiomegaly (cor Bovinum). There was a partial clinical response with treatment from the local physician. During the presentation, he had a blood pressure of 90/56 mm Hg in the right arm with a heart rate of 120 beats/min. Betablocker carvedilol was started in the dose of 0.2 mg/kg twice daily and the hemodynamic response was assessed after 15 days. The beta-blocker therapy was well tolerated by the patient. The dose of Carvedilol was escalated to 0.4 mg/kg twice daily and the child was advised for review in every 3 months. Aldosterone antagonist spironolactone (Dose: 6.25 mg twice daily) was also added which is well proved to reduce mortality in heart failure by lowering the angiotensin toxicity on myofibers.

The child's milestones were normal as per the age at the time of the index visit. On follow-up at 6 months, baby's LV had almost regressed to near normal size (Fig. 4) with good improvement in EF (44%). During follow-up his weight was normal (12 kg). The child had no feeding difficulty. The optimum dose of beta-blocker provided a favorable outcome in idiopathic DCM during infancy.

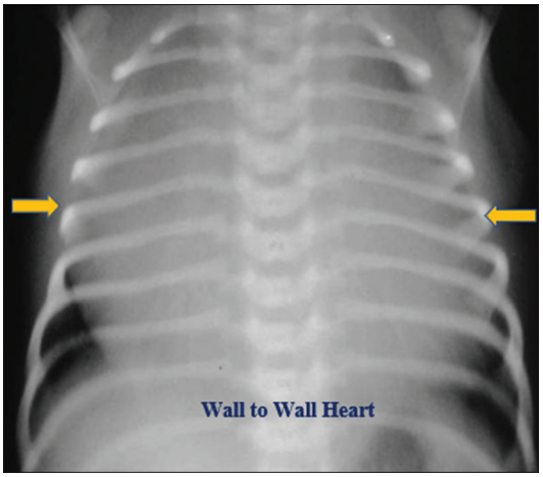

Figure 1: Cor bovinum of infancy

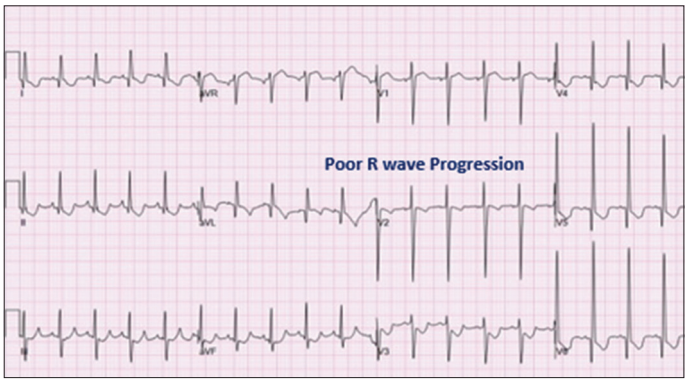

Figure 2: Electrocardiogram showing poor R wave progression and left ventricular hypertrophy

DISCUSSION

DCM in infancy poses a multitude of problems from an etiology to prognosis. Most often after excluding a history of viral myocarditis suggested by the triad of fever, exanthematous rash, and acute onset shortness of breath, physicians predict a reversible cause like hypothyroidism and selenium deficiency. ALCAPA needs specific exclusion and this entity has few characteristic features. These include the presence of Q waves with T wave inversion in leads I and a VL suggestive of lateral wall ischemia as revealed from ECG [4]. ALCAPA reveals papillary muscle scarring and calcification and provides the echocardiographer an indirect clue. Echocardiography reveals huge dilatation of the right coronary artery, left coronary sinus being empty, and low gain color doppler reveals a reversal of blood flow from left coronary to pulmonary artery [5]. Pompe disease, although a rare entity reveals echoluscence of the interventricular septum due to glycogen deposition. In the current case, the baby was diagnosed with a huge cardiomegaly with a "Wall- to- wall" heart, a feature

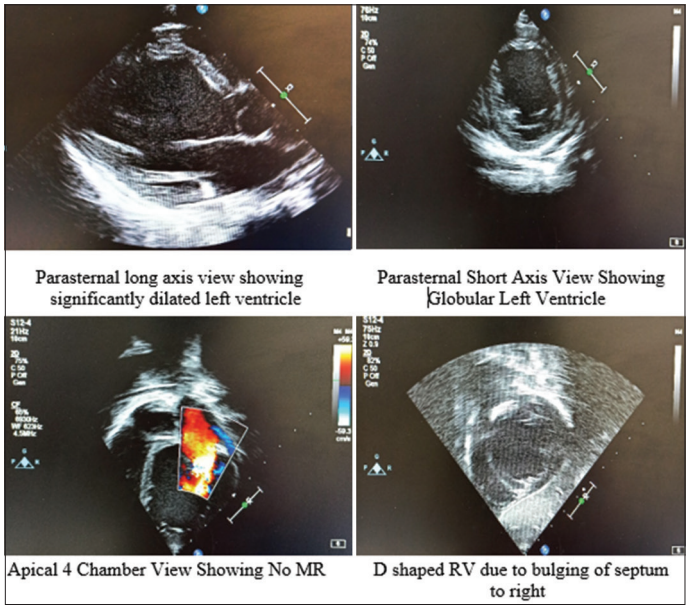

Figure 3: Echocardiography revealing dilated cardiomyopathy with a globular left ventricle, with no mitral regurgitation and D shaped right ventricle

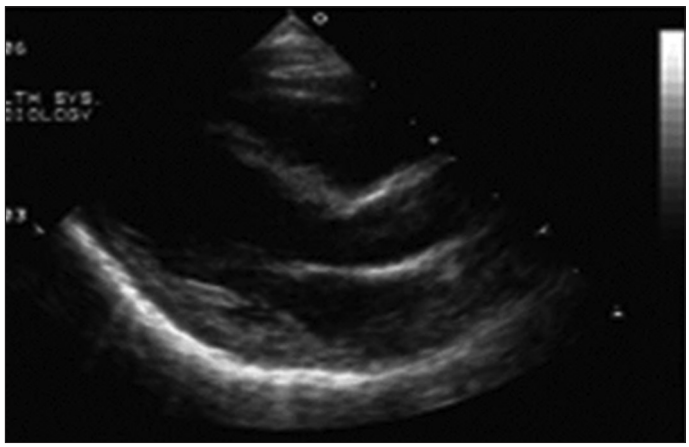

Figure 4: Near normal left ventricle in follow up after 6 months

of Ebstein's anomaly. However, this feature is also found in the hugely enlarged heart or Cor Bovinum of DCM. Cor Bovinum in old literary terms referred to the presence of huge cardiomegaly in the syphilitic heart [6] with severe aortic regurgitation with a tamboor quality early diastolic rumble. The presence of this type of huge cardiomegaly always needs to rule out the presence of pericardial effusion. Echocardiography in such huge dilatation of heart needs exclusion of MR secondary to annular dilatation and also needs exclusion of presence mural thrombus. Tachycardiomyopathy is also of special mention in infancy, especially paroxysmal junctional reciprocating tachycardia (PJRT) [7]. PJRT is an incessant narrow QRS tachycardia with negative P wave in II, III, aVF and V_5, V_6 through the right posteroseptal pathway. PJRT has a unique feature in ECG that it is the only reentrant arrhythmia that is initiated by sinus beat whereas all other reentrant arrhythmias are initiated by the ectopic beat. Myocarditis in infancy is caused by Coxsackie, Echo, HIV, Measles, Mumps, and Rubella [8]. The increased antibody titer in paired serum is diagnostic [8]. But most often inflammatory cardiomyopathy induced by viral myocarditis presents late after an insult; that's why it is commonly described as "virus hits and run" for which, obtaining a raised antibody titer is often quite difficult. Endomyocardial biopsy in children carries a high risk of inciting arrhythmias and chamber perforation.

DCM can be familial too. Autosomal dominant inheritance with incomplete penetration, recessive, and X-linked inheritance have been described. It is prudent to search for DCM in parents and other siblings as a familial incidence of DCM in up to 20% of cases. DCM in early infancy can be due to metabolic, endocrine, storage, mitochondrial and connective tissue disorders. Anemia can also cause dilatation of the heart which is reversible and serum iron and iron-binding capacity should be sent routinely in each patient. Blood should be sent for lactate, glucose, amino acid, carnitine, acylcarnitine, cholesterol, and triglycerides. A complete blood count should be sent for an absolute neutrophil count and vacuolated lymphocytes. Early morning urine analysis for amino acid, organic acid, and glycosaminoglycans rule out metabolic disease [8]. All those aforesaid parameters were normal in the child we studied.

Most often diagnosis of idiopathic DCM is reached after a battery of investigations. The objective of drug therapy in DCM is to provide symptomatic relief and maximize cardiac function. ACE inhibitors such as enalapril maleate decrease morbidity and mortality, favors a positive ventricular remodeling, decreases the severity of MR, decreases the angiotensin toxicity on myofibers, and improve the EF. ACE inhibitors are generally well tolerated by children and they may result in first-dose hypotension, cough, and hyperkalemia in children on potassium-sparing diuretics. In-hospital patients are usually monitored for hypotension induced by ACE inhibitors, the lowest dose is started and it is gradually escalated. Beta-blocker decreases the heart rate, gives rest to the failing heart, decreases the myocardial oxygen demand, decreases the sympathetic drive which is compensatory in heart failure, improves the coronary perfusion by prolonging the diastole, prevents the occurrence of arrhythmias by its membrane-stabilizing property and improves the outcome in chronic heart failure. Carvedilol is promising in this aspect with an additional role of having the potency to provide vasorelaxation which increases the forward flow. Digoxin is used as a second-line agent because it does not decrease mortality although it decreases the frequency of heart failure hospitalization. Digoxin is out of use in current days as it is emetogenic, proarrhythmic, even the presence of a single premature ventricular complex in ECG is a contraindication for its use. Growth hormone therapy may be used as an add-on during the bridge to transplantation. Survival rate in patients with DCM at 1–5 years are 79–61%, respectively. The optimal period to intervene in a DCM of infancy is within the 1st year after diagnosis. Early deaths in pediatric DCM is due to heart failure and late deaths are due to arrhythmias. Adverse prognostic factors in DCM include age at initial presentation >2 years, non-improvement or deterioration of left ventricular systolic dysfunction, presence of moderate to severe MR, development of severe PAH and right ventricular systolic dysfunction, presence of left ventricular thrombus, development of ventricular arrhythmias, presence of diffuse endocardial fibroelastosis and invasive LVEDP >20 mmHg [9]. Surgical options in DCM include Batista operation where partial left ventriculotomy is combined with mitral valve repair to restore left ventricular systolic function but till now there is no experience in children. High ionotropic support with a left ventricular assist device (Impeller) is the only option for the bridge to transplantation in children. End-stage refractory heart failure is managed with transplantation although this option is less promising to be exercised in India. Optimizing the ACE inhibitor and betablocker are the only options to be exercised to achieve a favorable outcome. Carvedilol can be started in the dose of 0.05 mg/kg twice daily in children with a low range of blood pressure and if it is well-tolerated the dose is increased to 0.4 mg/kg twice daily. We put the patient on the optimum dose of betablocker for 6 months which was not initially advised by the primary care physician and resulted in dramatical increase of the left ventricular EF from 23% to 44% over a period of 6 months. If we can achieve the maximum dose of enalapril (0.5 mg/kg/day), carvedilol (0.4 mg/kg/day), and spironolactone (3 mg/kg/day), we can provide the best treatment to the child. The take-home message learnt from the current case is that adequately optimized doses of betablockers can save children in idiopathic DCM.

CONCLUSION

DCM is a rare disease in children but morbidity and mortality in affected patients are high. Here we discussed the clinical presentation, diagnosis, medical management, and prognosis of the condition, with an emphasis on recent advances that have influenced the management of these children. Beta-blockers are the cornerstone in the management of idiopathic DCM of infancy. Optimum dosage of beta-blockers improves positive ventricular remodeling and can also normalize the ventricular function as exemplified in our case. Hence, we conclude that, although idiopathic DCM of infancy is a foggy road of winter

from diagnosis to management, optimum dosage of carvedilol may help in reaching the destination.

REFERENCES

1. Garcia VV, Calle DG, Garay CC, Monsalvo MG, Rodriguez JB, Sousa GB, *et al*. Use of betablocker in dilated cardiomyopathy and relationship with cardiovascular events. Eur Heart J 2020;41:2100.
2. de Araújo Rocha Reis Filho JR, Cardoso JN, dos Reis Cardoso CM, Pereira-Barretto AC. Reverse cardiac remodelling: A marker of better prognosis in heart failure. Arq Bras Cardiol 2015;104:502-6.
3. Askari H, Semizel E, Bostan OM, Cil E. Carvedilol therapy in pediatric patients with dilated cardiomyopathy. Turk J Pediatr 2000;51:22-7.
4. Hoffman JE. Electrocardiogram of anomalous left coronary artery from pulmonary artery in infants. Pediatr Cardiol 2013;34:489-91.
5. Yuan XC, Hu J, Zeng Z, Zhou AY, Chen L. Echocardiographic diagnosis of anomalous origin of left coronary artery from the pulmonary artery. Medicine 2019;98:e18046.
6. Madke BS, Agrawal NB, Vaideeswar P, Pradhan M, Rojekar AV, Khopkar US. Luetic aortopathy: Revisted. Indian J Sex Transm Dis AIDS 2010;31:118-21.
7. Rodriguez EV, Pina HR, Bouthillier AP, Mejia LM, Arteaga SJ, Sanchez AM, *et al*. Tachycardia induced cardiomyopathy in children and adolescents. Bol Med Hosp Infant Mex 2013;70:231-43.
8. Dancea AB. Myocarditis in infants and children: A review for the paediatrician. Paediatr Child Health 2001;6:543-5.
9. Alvarez JA, Wilkinson JD, Lipshultz SE. Outcome predictors for pediatric dilated cardiomyopathy: A systematic review. Prog Pediatr Cardiol 2007;23:25-32.

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