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Dilated cardiomyopathy in childhood

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Abstract

Dilated Cardiomyopathy is an uncommon disease in children but morbidity and mortality in affected patients are high. This review discusses clinical presentation, diagnosis, medical management and prognosis of the condition, with an emphasis on recent advances that have influenced management of these children.

MeSH: Cardiomyopathy, congestive, Myocarditis/diagnosis, Heart transplantation, Prognosis

Presentation and investigation of dilated cardiomyopathy

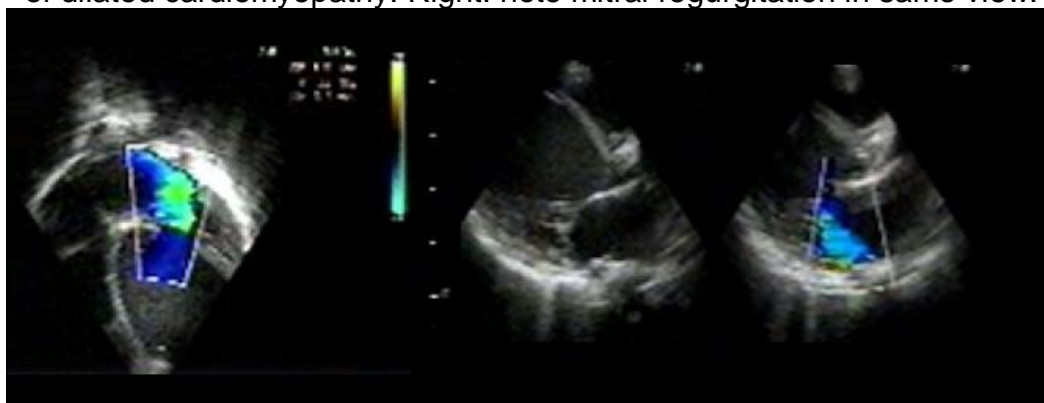
At the time of presentation the child with dilated cardiomyopathy (DCM) is usually in symptomatic cardiac failure, and at initial assessment, it is important to differentiate this condition from bronchiolitis. The chest x-ray features of increased cardiothoracic ratio, with evidence of lateral bronchial displacement due to left atrial enlargement and pulmonary plethora in association with hepatomegaly, raises clinical suspicion of the diagnosis (figure 1).

Figure 1 Chest x-ray in a child with dilated cardiomyopathy



Echocardiography is confirmatory showing dilatation of cardiac chambers, with or without mitral regurgitation (figure 2), and reduced ventricular function on M Mode analysis. It is very important to exclude mural thrombus on echocardiogram as its presence requires urgent treatment.

Figure 2 On left: echocardiographic apical four chamber view of dilated cardiomyopathy - note mitral regurgitation. Middle: parasternal long axis view of dilated cardiomyopathy. Right: note mitral regurgitation in same view.



Both coronary arteries should be identified in an attempt to exclude anomalous origin of the left coronary artery from the pulmonary artery although this can be difficult¹ and angiography may be required (figure 3).

The electrocardiogram may be more useful in this respect showing deep Q waves in lead I and wide Q waves in lead AVL, where the origin of the left coronary artery is anomalous.²

More commonly in idiopathic DCM sinus tachycardia, increased left ventricular voltages and ischaemic changes are seen on ECG at presentation. The QRS complex may be broad due to conduction disturbance and evidence of right atrial hypertrophy and left atrial hypertrophy is sometimes evident. 24

hour ECG monitoring will exclude chronic tachycardia with secondary ventricular dilatation e.g. permanent junctional reciprocal tachycardia, which is associated with a moderate increased heart rate and an abnormal P axis and will also identify children with secondary arrhythmias.

Figure 3 Aortogram in anomalous origin of left coronary artery



Differential diagnosis

There are a number of alternate diagnoses that need to be excluded at the time of presentation, as management and therapeutic strategies may need to be altered accordingly. Anomalous origin of the left coronary artery, the ECG features of which are described above, usually presents at 2-3 months and can be confirmed at cardiac catheter. This condition is surgically correctable. Myocarditis is more difficult to diagnose with accuracy but an attempt should be made as spontaneous resolution is likely and short-term mechanical support may be more appropriate than referral for transplantation. Positive viral cultures or increased antibody titres on paired serum samples may help. Common causes include viral infection with coxsackie, echo, HIV, measles, mumps and rubella but all families of micro-organisms have been implicated. Myocardial biopsy is rarely indicated, because of the risk.

In most children dilated cardiomyopathy is a sporadic condition of unknown cause. However familial cases have been reported with autosomal dominant with incomplete penetrance, recessive and x-linked inheritance patterns described. Michels et al³ demonstrated a prevalence of familial disease in 20% of index cases in a prospective study where asymptomatic first-degree relatives were screened. No features specific to familial disease have been identified. It is our policy to offer such screening, and this needs to be handled sensitively.

DCM as a secondary disease

DCM is rarely due to systemic disease however as myocardial damage may be reversible with treatment of the underlying pathology it is essential to attempt to rule out metabolic, endocrine, storage, mitochondrial and connective tissue disorders at first presentation. Blood may be taken for lactate, glucose, amino acids, carnitine and acylcarnitine, cholesterol and triglycerides, thyroid function, creatinine kinase, iron and iron binding capacity and uric acid. A full blood count to assess absolute neutrophil count and vacuolated lymphocytes may also be helpful. Early morning urine analysis for amino-acids, organic acids and glycosamine glycans may further exclude metabolic disease.

Medical management

The objective of drug therapy in DCM is to give supportive relief and maximize cardiac function. There is not as yet a treatment that offers a cure.

Diuretic have an established role particularly in their ability to produce rapid symptom relief, but earlier use of ACE inhibitors has probably resulted in lower doses being used in recent years. ACE inhibitors have been consistently shown to reduce morbidity and mortality in adult studies,^{4,5} Enalapril maleate was used in these series, there is limited information available on its use in children where captopril is most commonly prescribed. ACE inhibitors are generally well tolerated, their principle side effects include first dose hypotension, non-productive cough, and a risk of hyperkalaemia, especially in patients already on potassium supplements or potassium sparing diuretics.

Treatment with B-blockers needs also to be considered. The increased sympathetic drive that occurs as a compensatory mechanism in chronic heart failure appears to have an inverse relationship with prognosis^{6,7}. B-blockers down regulate this sympathetic overdrive⁸ and the evidence that they improve outcome in adult patients with chronic heart failure is increasing^{9,10,11}. The a-adrenergic effects of the third generation B-blockers e.g. Carvedilol and Timolol also cause vasodilatation and this may be helpful.

Digoxin has a place as an orally active inotrope, but its failure to actively reduce mortality in adults¹² means it is increasingly relegated to second line therapy. Intravenous inotropes may occasionally be necessary to support the child in end stage cardiac failure. Their use is an indication for transplant assessment in some centers.¹³

Arrhythmias may also require treatment but this needs to be carefully evaluated, as many anti-arrhythmic drugs are negatively inotropic. All children with poor LV function are at risk of mural thrombus and should be treated prophylactically with aspirin, the detection of clot requires urgent anticoagulation (fig. 4).

Growth hormone may be effective as add on therapy in the child who is failing to improve on conventional medical management. Early studies in adults¹⁴ have shown improvement in LV function and exercise tolerance and reduced myocardial oxygen consumption with GH therapy that deteriorated when therapy was discontinued. In the future subcutaneous GH could have a role as a medical 'bridge to transplantation' but further work need to be done.

Figure 4 Mural thrombus (MT) in a child with dilated cardiomyopathy



Prognosis

Mortality for DCM is highest in the first year after diagnosis with a reported survival at 1 and 5 years after first presentation of 79% and 61% respectively.¹⁵ Early deaths are principally caused by severe heart failure. Some late deaths are sudden, presumably due to arrhythmia, in children who fail to recover to normal ventricular function. While it is accepted that the risk of mortality is high there is less agreement as to predictors of poor outcome. Failure of improvement or deterioration in shortening fraction, ventricular arrhythmias, detection of mural thrombus, presentation at age >2years, endocardial fibroelastosis and left ventricular end diastolic pressure > 20mmHg have all been put forward as adverse prognostic factors.^{15,16,17,18}

Mechanical and surgical support

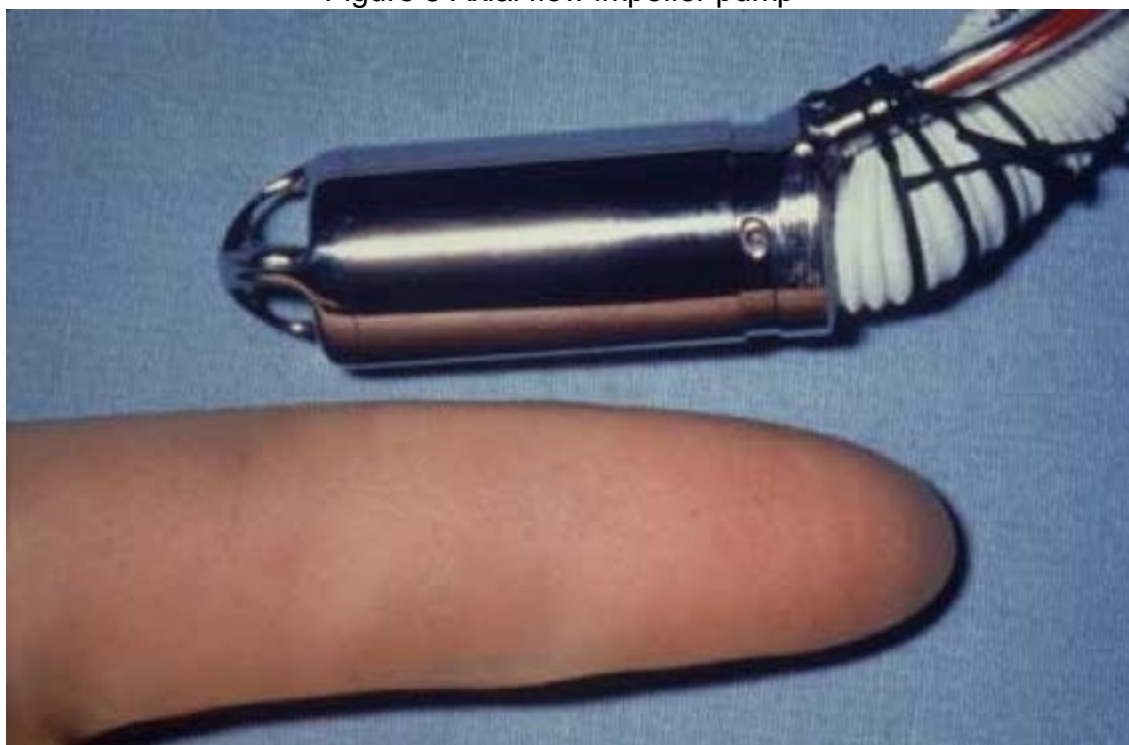
Left ventricular assist devices and biventricular assist devices, although not widely available yet in the UK will have a role in the short term, providing a bridge to transplant for the child with intractable cardiac failure. In the future indwelling axial flow impeller pumps such as the JARVIK2000 Heart may have a place in the long term mechanical support of this difficult group of patients (fig. 5).¹⁹

End stage cardiac failure secondary to DCM has been the most common indication for heart transplantation in children and adolescents.²⁰ Survival statistics post transplantation are improving. In a series reported from Great

Ormond Street, Adwani reported survival for 95% at 1 year and 87% at 3 years, in patients transplanted for dilated cardiomyopathy.¹³ Currently the principle limiting factor in paediatric cardiac transplantation is a shortage of donor organs hence the importance of developing a mechanical support system suitable for use in the long term.

A second surgical option is the Batista operation where a partial left ventriculectomy is combined with mitral valve repair to restore left ventricular dimensions to normal thus improving pump function. Paediatric experience of this operation is limited but a 55% 2 year survival was reported in adult patients in the US, with most survivors showing symptomatic improvement.²¹

Figure 5 Axial flow impeller pump



Conclusion

Management of children with dilated cardiomyopathy remains difficult but recent advances including early introduction of ACE inhibitors and B-blockers may improve what is currently a bleak outlook. In the future implantable left ventricular assist devices may provide interim mechanical support, but referral for transplantation remains the cornerstone of treatment.

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