Childhood dilated cardiomyopathy (DCM) is characterized by a dilated left ventricle and systolic dysfunction, and in some patients also by right ventricle failure. It is a serious myocardial disease, usually idiopathic, but its infectious, metabolic and genetic aetiologies are increasingly revealed. DCM frequently results in heart failure and sometimes death. It is the most common form of cardiomyopathy and the most common reason for heart transplantation in children.

The original article by Inas Adullsatar Saad in the September 2007 issue of the Libyan Journal of Medicine presents a study of natural history and predictors of prognosis in Arab children with idiopathic DCM [1]. Poor clinical outcome was found to be related to older age and echocardiographic findings of increased end-diastolic volume z-score at presentation. On the other hand, a higher z-score value for interventricular septum and left ventricular posterior wall dimension in diastole was related to a favourable outcome. At short to medium follow-up (up to 4 years), 50% of the patients improved and another 25% had a stationary course. Most of the patients who deteriorated died as heart transplantation was not an option.

Because many children with DCM die early, emphasis has recently been placed on using echocardiographic and electrocardiographic measures to identify those who have the best chance of survival by early heart transplantation [2,3,17]. However, we should be aware of the technical difficulties of echocardiography in children with DCM and the large overlap of individual echocardiographic measures among the outcome groups. We still lack a more comprehensive, predictive risk algorithm to stratify children with DCM for death, transplant or recovery [3,17].

Improving causal diagnosis by more intense evaluation

At least one third of children with DCM have a known cause at the time of diagnosis, the rest are classified as idiopathic [4]. Increasing the causal diagnostic rate is important to consider a cause-specific treatment, to predict prognosis, and to estimate the recurrence risk when the cause is genetic. After initial viral and metabolic testing, early endomyocardial biopsy and molecular viral testing might improve the diagnostic evaluation, at least in older children [4]. With the extended use of early heart biopsy, a potential viral contribution and increasing diagnosis of lymphocytic myocarditis has been reported [4,5]. Viral testing, including polymerase chain reaction (PCR), has predominantly shown positive findings of coxsackie and adenovirus [4,5]. In viral myocarditis, the effects of the virus on the cellular immune system, and the activation of viral proteases that cleave membrane-stabilizing proteins such as dystrophin, seem to be an important mechanism in enteroviral myocarditis and leading to later DCM. [4,6,7]. These findings raise the possibility of future specific treatment by vaccination, administration of cytokine and protease inhibitors or, in general terms, immunosuppressive or immunomodulating therapy [4,6]. Immunoglobulin, sometimes combined with corticosteroids, has been used as an immunomodulating therapy in acute fulminant myocarditis. This treatment is based mainly on case reports of favourable outcome [8].

An increasing number of metabolic and genetic causes of DCM are being revealed, especially in young patients. Thus, appropriate metabolic (urine and blood) laboratory testing, a thorough family history, and echocardiography of first-degree relatives should be considered [4] Molecular genetic studies, now increasingly available for clinical use, have shown that DCM is caused by defects in genes encoding cytoskeletal proteins eg defects in dystrophin and tafazzin. [4]. A recent Australian study showed that about 10% of paediatric DCM patients had parental consanguinity [5]. This finding may contribute to familial DCM and was probably due to frequent intra-family marriages in the original Australian population. In familial DCM with unsatisfactory prognosis, cardiac autoantibodies were predictive of disease development even in asymptomatic relatives [9].

Management of heart failure- clinical practice and new treatment strategies.

Conventional management to stabilise congestive heart failure in childhood DCM consists of digoxin, diuretics and inhibition of angiotensin converting enzyme (ACE). Aldosterone inhibitor might be added to spare electrolytes, and their long-term use is intended to decrease myocardial fibrosis [10]. In children with low ejection fraction on echocardiography, anticoagulation therapy based on clinical experience seems to be important for preventing mural thrombi. This group of severely ill young patients might also be managed by intravenous positive inotropic agents (beta-adrenergic agonists and phosphodiesterase
inhibitors) and the new calcium-sensitising agent, levosimendan [11]. Most recommendations for managing heart failure are based on trials on adults. However aetiologies and pathophysiology in children with heart failure might differ from those in adults, and therefore these young patients with heart failure and DCM might need different treatment strategies.

Retrospective studies in DCM children indicate that prolonged use of beta-blockers (carvedilol and metoprolol) seem to improve ventricular function [12,13]. The results of a prospective multi-centre trial of carvedilol in pediatric DCM recently published, do not show improvement in heart failure, however there might be a differential effect of carvedilol based on ventricular morphology. Furthermore the trial was underpowered [14,15].

Mechanical circulatory support, such as left ventricular assist devices, even in young severely ill patients with DCM, is now available for short-term cardiac support, or more prolonged use as a bridge to heart transplantation [16]. There have also been successful trials with devices in small groups of grown-up children with heart failure using implantable cardioverter defibrillators and both acute and long-term resynchronisation therapy in children with ventricular dyssynchrony [16].

These new therapies for severely ill paediatric patients with DCM might improve the final outcome considering the limitations in use of paediatric heart transplantation.

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