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Original Research Article

The Outcome of Dilated Cardiomyopathy in Pediatric Age Group After One Year Treatment for Acute and Chronic LV Dysfunction

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Abstract

Dilated cardiomyopathy (DCM) means congestive heart failure as a result of dilation and systolic dysfunction of cardiac ventricles (mainly the left ventricle). It is a common type of cardiac muscle disease in pediatric age group. Many children with DCM are without symptoms, but known patients with DCM may present with acute heart failure, others may be developed ventricular abnormal rhythm and atrioventricular block, syncopal attack and sudden death . Dilated cardiomyopathy (DCM) in pediatric age group has a worse prognosis, the survival rate was 41% at 1 year and 20% at 3 years.

The participants belongs to both 48 patients(15 patients with acute DCM , mean age (10.24±11.38 months; 8 females and 7 males) and 33 patients with chronic DCM (mean age (10.46±10.72 months;18females and 15 males) were selected and examined in the cardiology clinic of Karbala pediatric teaching hospital between April 2013 and October 2014. 2- D echocardiography, M- mode for assessment of cardiac function (Ejection fraction (EF).

At the time of attendance , all the patients with acute DCM (NO.=15) had low EF, with treatment , there was improvement in the EF during the period of treatment and follow up, after 1 year 66.7% of patients had normal EF.

Also, all patients with chronic DCM (NO.=33) had low EF, with treatment , there was improvement in EF, after 1 year 60.6% of patients had normal EF. At the time of attendance , all the patients with acute and chronic DCM had low EF, with treatment, there was improvement in the EF during the period of treatment and follow up, after 1 year of treatment and follow up.

الخلاصة

اعتلال العضلة القلبية الموسع أو الممدد يعني قصور القلب ألاحتقاني نتيجة لتمدد وتوسع البطينات القلبية(بصورة رئيسية البطين الأيسر) مع قصور في الوظائف الانقباضية وهو نوع شائع من الأمراض العضلية القلبية عند الأطفال.كثير من الأطفال المصابين باعتلال العضلة القلبية الموسع يظهرون احتقان قلبي حاد لا معوض.عدد من الأخطار التي تهدد حياة المصابين يعود إلى تواتر بطيني غير طبيعي وأعاقه أذينية بطينية مع نوبات إغماء وحتى موت مفاجئ ان اعتلال العضلة القلبية الموسع لدى الأطفال له تكهنات سيئة وحيث تكون نسبة العيش لسنه واحدة ٤١% مقابل ٢٠%لثلاث سنوات لذا فان العلاج المكثف والقريب لهذه الحالات يجب ان يقر في المراحل الابتدائية المبكرة للمرض في المجاميع ذات الخطورة العالية.ان هدف هذه الدراسة هو بيان نتيجة متابعة الأطفال المصابين باعتلال العضلة القلبية الموسع لسنة واحدة من العلاج والمتابعة. شملت هذه الدراسة ٤٨ طفلا مصابا باعتلال عضلة القلب الموسع والذين يعانون من قصور وظائف البطين الأيسر ثم تشخيصهم سريريا بواسطة اخذ تاريخ المرض والفحص السريري وتخطيط القلب الكهربائي وفحص القلب بالا يكو أحادي وثنائي الأبعاد والدوبلر .ويعتبر الايكو حجر الزاوية في تشخيص حالات اعتلال العضلة القلبية الموسع.وقد تم تقسيم هؤلاء المرضى إلى ١٥ طفلا من الذين اظهروا قصورا قلبيا حادا عند حضورهم للمستشفى وكانوا٨ إناث و٧ ذكور بالإضافة إلى ٣٣ طفل هم معروفين ومسجلين لدى سل الاستشارية القلبية ومعروفين بقصور وظيفي مزمن للبطين الأيسر وهم ١٨ إناث مقابل ١٥ ذكور. كلا المجموعتين تمت متابعتهم بعد بدء العلاج ولسنة كاملة بصورة منتظمة بالفحص السريري والمختبري وفحص الايكو خلال فترة العلاج لهذا القصور القلبي الوظيفي. في كثير من المرضى المصابين لوحظ زيادة في وظيفة الضخ للجانب الأيسر للقلب خلال عام كامل من العلاج المصاحب لعجز القلب بشكل ملحوظ

متمثل بتحسن الكسر الجزيئي النسبي (EF), والقصر الكسري (FS)وكلاهما لوحظوا بإظهار الزيادة مع التحسن الوظيفي. وقد لوحظ بان عدد من المرضى يظهرون تلكؤ بالوظيفة القلبية عند إيقاف العلاج لديهم لذا لوحظ ضرورة إبقائهم على العلاج لفترة أطول.

Introduction

Dilated cardiomyopathy (DCM) means congestive heart failure as a result of dilation and systolic dysfunction of cardiac ventricles (mainly the left ventricle).It is a common type of cardiac muscle disease in pediatric age group. Many children with DCM are without symptoms , but known patients with DCM may present with acute heart failure,others may be developed ventricular abnormal rhythm and atrioventricular block, syncopal attack and sudden death.Dilated cardiomyopathy (DCM) in pediatric age group has a worse prognosis, the survival rate was 41% at 1 year and 20% at 3 years. Close intensive therapy should be considered in the early stages of (DCM) in this high-risk group.In children the incidence is 0.57 cases per 100000 per year overall, but is higher in male children than in female (0.66 in male children and 0.47 patient per 100000and is more in babies younger than 1 year than in children (4.40 case in <1 year age while0.34 cases in older children per 100000. Two third of the affected children,with DCM might have idiopathic disease. In adult, the prevalence of DCM is one case in 2500 individual, and the incidence of DCM is (7) cases per 100000 per year (but many patients may pass undiagnosed) (Taylor MR et al, 2006).The disease may be familial in 20-48% of cases. A good understanding of the causes of the disease is important in children and adult.

Materials and Methods

The participants belongs to both48 patients(15 patients with acute DCM, mean age (10.24±11.38months; 8 females and 7 males) and 33 patientswith chronic DCM(mean age (10.46±10.72months;18females and 15 males) were selected and examined in the cardiology clinic of Karbala pediatric teaching hospital between April 2013 and October 2014, the type of test was discussed for the parents , starting with history taking especially family history, and presence of familial cardiac disease, the age and degree of relatives of both parents, past medical history and antenatal history followed by physical examination. Electro-cardiography (ECG), were done for all patients, then ultrasonic examination of the heart by using, 2- D echocardio-graphy, M-mode for assessment of cardiac function (EF, FS), chamber size, cardiac wall thickness in addition to Doppler study of cardiac valves for detection of valvular complications and presence of regurgitation, and to exclude other cardiac wall defects or congenital anomaly.Treatment of patient diagnosed with DCM involve mainly Digoxin,Furosamide and Enalopril.

This regime of examination was repeated for all patients during the period of the study with regular spacing and periods after starting medical treatment for the first group (those with acute presentation), after 3 months,6 months and 1year, and for the second group (those who are known cases of dilated cardiomyopathy), after 6 month and 1 year.

Results

Outcome ofDilated Cardio-myopathy with acute LV dysfunction

At the time of attendance, all the patients (NO.=15) had low EF, with treatment, there was improvement in the EF during the period of treatment and follow up, after 1 year 66.7% of patients had normal EF as shown in figure (1).

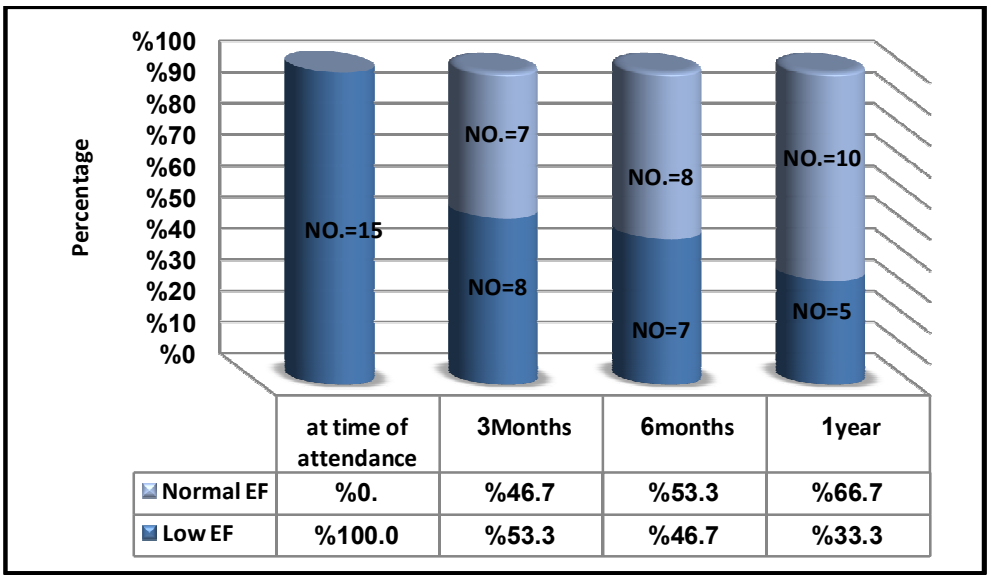

Figure 1: Outcome of DCM with acute LV dysfunction after 3, 6 and 12 month of treatment

The Outcome ofDCM with chronic LV dysfunction

At time of attendance , all patients (NO.=33) had low EF, with treatment, there was improvement in EF, after 1 year 60.6% of patients had normal EF as shown in figure (2).

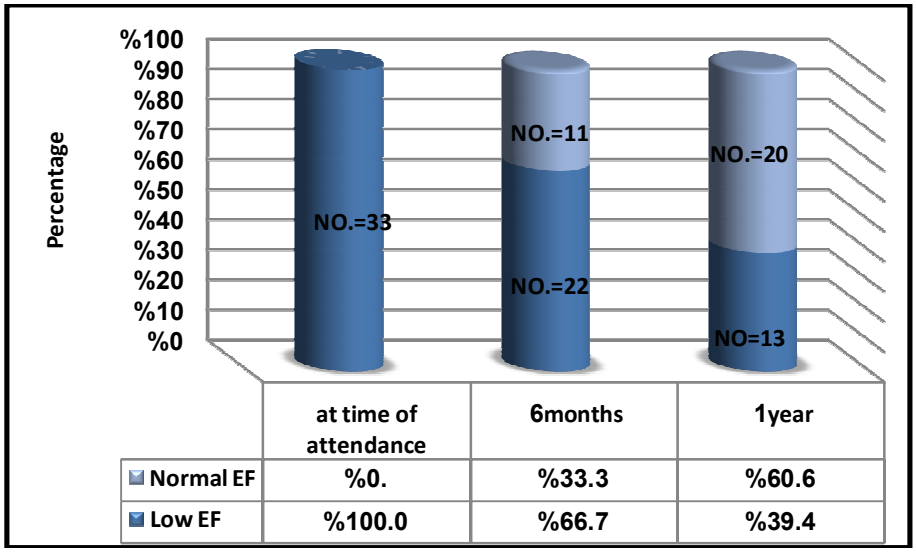

Figure 2: Outcome of DCM with chronic LV dysfunction after 6 and 12 month of treatment

Outcome of Acute and Chronic DCM

At the time of attendance, all patient with acute and chronic DCM had low EF (100%), with treatment, there was improvement in EF in both group, after 6 month and 1 year later , there was insignificant differencebetween acute and chronic DCM outcome (P>0.05) as shown in figure (3).

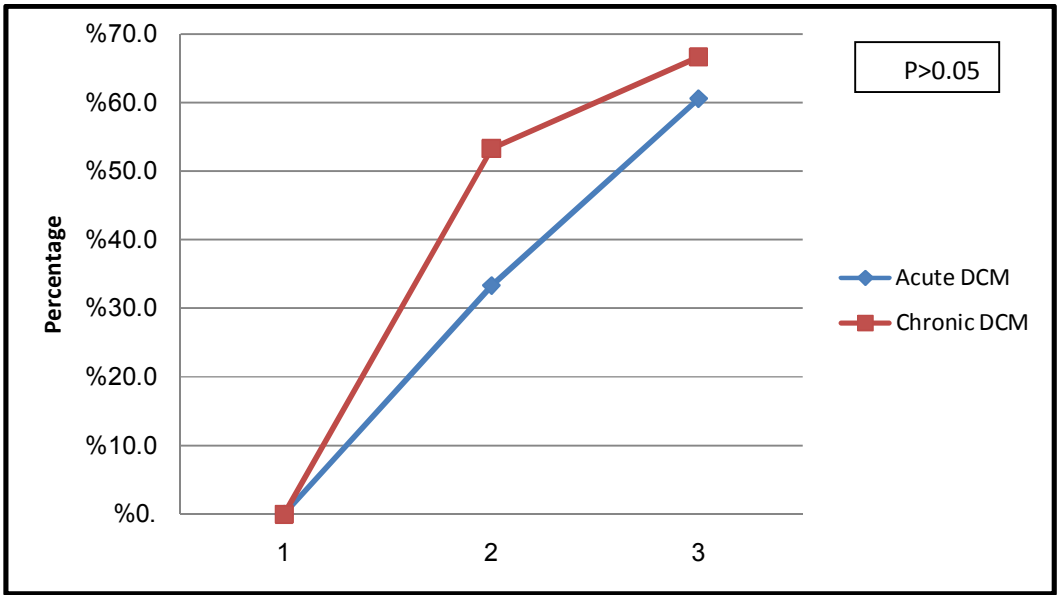

Figure 3: Outcome of acute and chronic DCM

Discussion

In this study we found that ,those patients that have dilated cardiomyopathy with acute left ventricular dysfunction who had higher level of troponin at the time of attendance shown to develop higher rate of improvement of the ejection fraction (EF) after one year than more other patient in the same group .At the time of attendance, all the patients had low ejection fraction (100) %, with treatment, there was improvement in the ejection fraction during the period of treatment and follow up, after 1 year of treatment, 66.7% of patients with acute cardiac failure had normal ejection fraction as shown in figure (1), probably may be due to early diagnosis and treatment and 66.6 of those who known as a chronic patient with chronic cardiac failure had improving their cardiac function after more than one year of treatment. In this study we found that,those patients with acute dilated cardiomyopathy that had higher level of troponin at the time of attendance shown to develop higher rate of improvement of the ejection fraction after one year than other patient in the same group and those children with acute presentation who are under the age of 2 years show a higher rate of improvement in their cardiac function, and this may be partly due to early diagnosis because of early acute presentation, and as it mention in many previous studies that regard the age under 2 years as a prognostic factor. While in those children with chronic left ventricular dysfunction, the rate of improvement in their cardiac function shown to be slower than those with acute presentation and this is probably because of more damage to myocardium that occur in such prolong period of the disease, and this is shown in many researches and studies that involve children with congenital heart disease with cardiac failure.

Conclusion

At the time of attendance, all the patients with acute and chronic DCM had low EF, with treatment, there was improvement in the EF during the period of treatment and follow up, after 1 year of treatment and follow up.It is important to follow patient with dilated cardiomyopathy for longer period to detect the five year survival rate and follow the improvement and deterioration among those children with DCM after cessation of treatment for long time.

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