International Journal of Pediatrics and Adolescent Medicine (2015) 2, 29—33

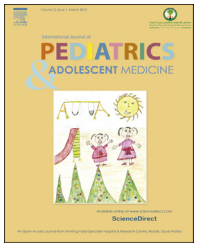
HOSTED BY

ELSEVIER

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.elsevier.com/locate/ijpam



INSTRUCTIVE CASE

Near missed reversible cardiomyopathy: The value of the electrocardiogram

Majid Al-Fayyadh*, Abdualah Alwadai, Abdulah Al Huzaimi, Zohair Al Halees

Heart Centre, King Faisal Specialist Hospital & Research Centre, Saudi Arabia

Received 6 January 2015; received in revised form 17 February 2015; accepted 25 February 2015
Available online 4 April 2015

KEYWORDS
Dilated cardiomyopathy;
Electrocardiogram;
Pediatrics;
ALCAPA;
Tachycardia

Abstract Dilated cardiomyopathy is a devastating disease affecting the myocardium that is characterized by cardiac chamber dilatation with contractile function impairment in the absence of structural heart disease. The majority of cases are idiopathic; these patients have a poor outcome. However, identifying a reversible etiology and instituting appropriate intervention could reverse the disease process and result in complete recovery. An electrocardiogram is a simple, non-invasive and quick test that should be performed in each patient presenting with dilated cardiomyopathy. Failure to perform this test or misinterpreting its result could result in a tragic misdiagnosis of idiopathic cardiomyopathy, depriving the patient of a potentially curative intervention. Here, we report two cases of dilated cardiomyopathy caused by clinical conditions with recognizable ECGs. In both cases, the diagnosis was missed initially, delaying corrective interventions. These cases draw attention to the importance of performing and correctly interpreting ECGs in patients with dilated cardiomyopathy.

1. Introduction

Dilated cardiomyopathy (DCM) refers to a large group of heterogeneous myocardial disorders that are characterized by ventricular dilation and depressed myocardial contractility in the absence of abnormal loading conditions, such as hypertension or valvular disease [1]. The Pediatric Cardiomyopathy Registry in North America reported the annual incidence of DCM to be 0.57 per 100,000 children per year [2]. Almost half of these patients die or undergo a heart transplant within 2 years of diagnosis [2,3]. In addition,

* Corresponding author. Heart Centre, MBC-16, King Faisal Specialist Hospital & Research Centre, Takhassusi Street, PO BOX 3354, Riyadh 11211, Saudi Arabia. Tel.: +966 11 4423921; fax: +966 11 4427791.
E-mail addresses: mfayyadh@kfshrc.edu.sa (M. Al-Fayyadh), awadai@kfshrc.edu.sa (A. Alwadai), aalhuzaimi@kfshrc.edu.sa (A. Al Huzaimi), alhalees@kfshrc.edu.sa (Z. Al Halees).
Peer review under responsibility of King Faisal Specialist Hospital & Research Centre (General Organization), Saudi Arabia.

http://dx.doi.org/10.1016/j.ijpam.2015.02.002

two-thirds of patients with DCM are thought to have DCM that is idiopathic in origin. Such a high frequency of idiopathic DCM may lead the practitioner to pay less attention during the search for an etiology of a newly diagnosed case, thereby committing the patient to a poor prognosis with high mortality.

However, finding and appropriately treating the primary cause of DCM may result in the total restoration of cardiac function and reversal of its sequela.

One of the routine and important tests in investigating DCM is an electrocardiogram (ECG). Unlike blood tests that produce quantitative results, the ECG result is based on physician interpretation that requires prior knowledge and experience. Failure to recognize the cardiac etiology of DCMs that manifest on an ECG can result in a delayed or missed diagnosis with potentially morbid or even fatal outcomes.

We report two patients who, on initial assessment, were misdiagnosed as idiopathic DCM or DCM following presumed myocarditis. Twelve months later, after careful interpretation of their ECGs, these patients were appropriately diagnosed and treated. Both underwent corrective intervention. One had complete recovery of cardiac function with reverse remodeling; the other patient is still undergoing follow-up treatment and is expected to improve.

2. Case 1

The first patient is a 2-year-old boy born at 36 weeks of gestation with a birth weight of 2.6 kg. As a one year old, he was admitted for two weeks with a history of tachypnea and poor weight gain. His weight at the time was 5.7 kg, and he had a height of 62 cm and a head circumference of 44 cm (all below the 5th percentile).

As part of the initial work-up, a chest X-ray and ECG were performed. The chest X-ray showed cardiomegaly, while the ECG showed sinus tachycardia, with a deep Q wave in leads I and aVL and left ventricular hypertrophy with a strain pattern in leads V5 and V6 (Fig. 1). These findings were not reported at the time.

The patient was referred for cardiac evaluation, and the diagnosis of idiopathic DCM was made after the echocardiogram showed a dilated left ventricle with an ejection fraction (EF) of 35% and mild mitral regurgitation. No comment was made about the origin of the left coronary artery.

The patient was started on anti-failure medication (furosemide, captopril and digoxin) and was discharged home. One year later, the patient was seen by a different cardiologist who spotted the ECG abnormalities and referred the patient to our center with a suspected anomalous left coronary artery from the pulmonary artery (ALCAPA). The clinical evaluation at our center was not different from the referring site; however, echocardiography showed the right coronary artery (RCA) to be dilated with multiple collaterals, while the left coronary artery (LCA) origin was not clearly observed. With color Doppler, the flow in the LCA was retrograde, which is highly suspicious for ALCAPA. To confirm the diagnosis, cardiac catheterization was performed. A selective angiogram showed a dilated RCA with multiple collaterals filling the LCA that originated from the pulmonary artery instead of the aorta (Fig. 2). The patient was taken for surgery, and the LCA was found to originate high on the MPA at the takeoff of the right pulmonary artery (Fig. 3). Such a high LCA takeoff is unusual for cases of ALCAPA, which explains the difficulty in visualizing its origin on echocardiography. The LCA was implanted to the aorta, and after a smooth post-operative course, the patient was discharged home on anti-failure medications four days after surgery. One month after surgery, he continues to be on medical therapy for his heart failure with the hope that cardiac function will improve with time.

3. Case 2

A 3-year-old boy presented for the first time to a local hospital with a one month history of shortness of breath, lethargy, and weight loss. These symptoms were preceded

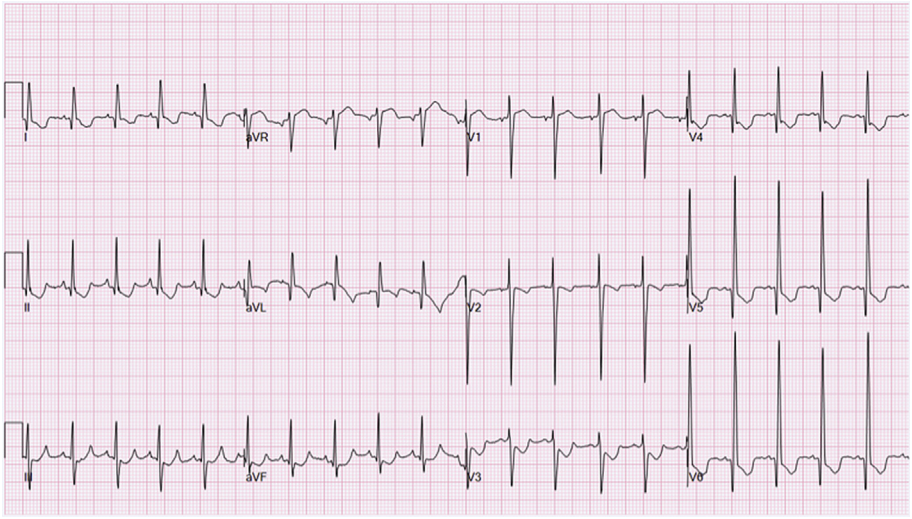


Figure 1 ECG of case 1. There is sinus tachycardia with deep Q wave in leads I and aVL strongly suggesting ALCAPA. There is also LVH with ST segment depression and T wave inversion most evident in V5 and V6.

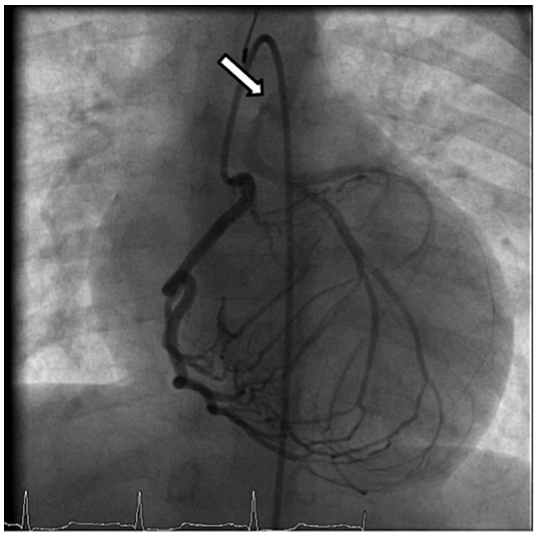


Figure 2 Selective right coronary artery angiogram showing dilated right coronary system with multiple collaterals to the left coronary artery which fills retrograde. There is spill of the contrast into the pulmonary artery (arrow) further confirming ALCAPA.

by an upper respiratory tract infection. His chest X-ray showed cardiomegaly, and echocardiography showed a dilated LV with depressed systolic function and an EF of 20%. The initial evaluation did not include an ECG.

He was treated as having viral myocarditis and was started on anti-failure medications (furosemide, captopril and digoxin). With no improvement in his condition, he was referred to our center for further management as a case of DCM. At our initial evaluation, his weight was 14.2 kg and his heart rate was regular at 160 beats per minute. The mother reported that the heart rate had been rapid for the last nine months.

The clinical examination was otherwise unremarkable apart from a pansystolic murmur at the apex.

A chest X-ray showed marked cardiomegaly, while the ECG showed narrow complex tachycardia at a rate of 160

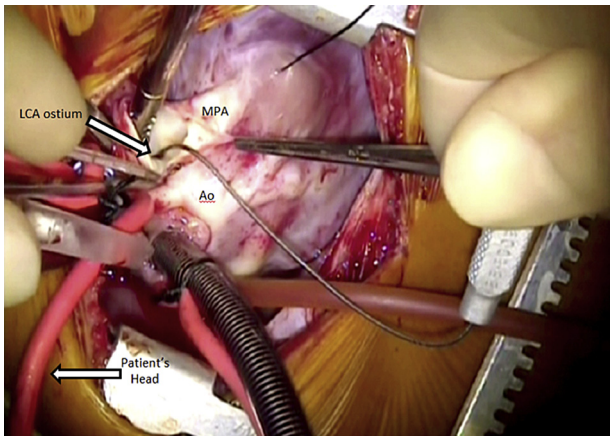


Figure 3 During surgery, the main pulmonary artery (MPA) is opened and a probe is placed in the LCA which arises close to the takeoff of the right pulmonary artery. The aorta (Ao) is posterior and to the right of the pulmonary artery.

beats per minute and an abnormal P-wave shape consistent with ectopic atrial tachycardia (EAT) or a permanent form of junctional reciprocating tachycardia (PJRT); both are a common cause of tachycardia-induced DCM (Fig. 4). The echocardiogram showed a severely dilated left atrium (SD + 8) and left ventricle (SD + 18) with severely depressed left ventricular systolic function (EF 10%). Based on the initial evaluation, a preliminary diagnosis of EAT was made and was confirmed by a 24-h ambulatory ECG recording that demonstrated the incessant nature of the rhythm. Amiodarone was introduced to the patient's medical regime to restore a sinus rhythm. Based on the behavior of the tachycardia and the good response to therapy, the tachycardia is likely to be EAT; however, PJRT remains a possibility that can only be excluded by an electrophysiology study. Nevertheless, the medical treatment and approach to radiofrequency ablation are the same in both situations. One month after the initiation of treatment with amiodarone, all symptoms disappeared and the EF increased to 30%. Within nine months, the cardiac function was normalized to an EF of 62% and the cardiomegaly was no longer evident on chest X-ray (Fig. 5).

4. Discussion

The outcome of symptomatic heart failure with DCM has not changed significantly over the last few decades [2—4]. Data from the pediatric DCM registry showed that about half of the children with DCM and symptomatic heart failure will die or require a cardiac transplant within two years of diagnosis. Controlling the symptoms of heart failure can be challenging in these patients; however, identifying the etiology causing the cardiomyopathy in the first place is the initial challenge.

Multiple disorders can affect the heart [5], ranging from primary genetic abnormalities involving the myocardium, infectious pathogens causing myocarditis, storage diseases and exposure to toxins. However, in most pediatric cardiomyopathy studies, an etiology is found in only about one third of the cases, while the remainders are labeled as idiopathic.

The search for a cardiomyopathy cause may be incomplete or misinterpreted if the clinician is inexperienced or careless. In one case, our patient had an ECG that was misinterpreted, while in the other case, an ECG was not performed at all. Both patients were assumed to have idiopathic DCM based on the initial incomplete evaluation. Although statistics indicate that the majority of DCM cases are idiopathic, it is unwise to assume this diagnosis unless a complete work-up is performed and other etiologies are ruled out.

In our first case, the diagnosis was evident on the initial ECG obtained at the time of presentation, and in both cases, the cause was treatable and the DCM was potentially reversible. Case 1 had an abnormal ECG that was suggestive of ischemia in addition to changes resulting from the disease process. The presence of Q waves in leads I and aVL should raise suspicion for ALCAPA in any child or infant presenting with cardiomyopathy [6]. Additionally, the ECG showed features of left ventricular strain with an ST-segment depression and T-wave inversion in the lateral

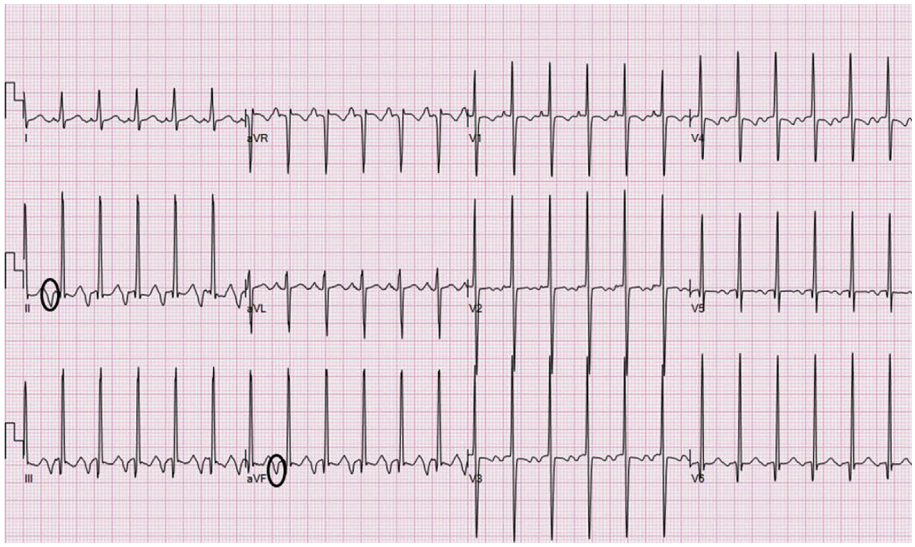


Figure 4 Case 2, ECG showing Narrow QRS long RP tachycardia at a rate of 160/minute with abnormal deep "P" wave in the inferior leads (circled) suggesting a non-sinus i.e. ectopic atrial tachycardia or permanent form of junctional reciprocating tachycardia.

leads. These findings should prompt the clinician to exclude coronary anomalies as a cause of the DCM. Systematic coronary artery evaluation by echocardiography using both 2D and color Doppler will identify the majority of these cases [7]. However, echocardiography can be misleading in some cases as a result of the "drop-out" at the coronary origin and the close proximity of the left coronary artery, which runs beside the aorta and redirects itself to the pulmonary artery [8]. In addition, color Doppler may not show the abnormal flow into the main pulmonary artery clearly if the pulmonary artery pressure is significantly elevated. In such cases, a more definitive study, such as a coronary angiogram or CT angiography, is indicated. In our patient, the echocardiogram was highly suggestive of ALCAPA, and to confirm the diagnosis, we elected to proceed with coronary angiography.

Typically, patients with ALCAPA present with heart failure in early infancy once the pulmonary pressure and resistance drops [6]. This results in the under-perfusion of the left ventricular myocardium, and eventually, the flow is reversed, flowing from the left coronary artery into the pulmonary artery. This in turn leads to myocardial ischemia, then infarction, dilated cardiomyopathy and eventually congestive heart failure. Initially, myocardial ischemia is transient, and if caught early enough, the damage is reversible with surgical correction. This outlines the importance of early recognition of ALCAPA as a cause of DCM in children.

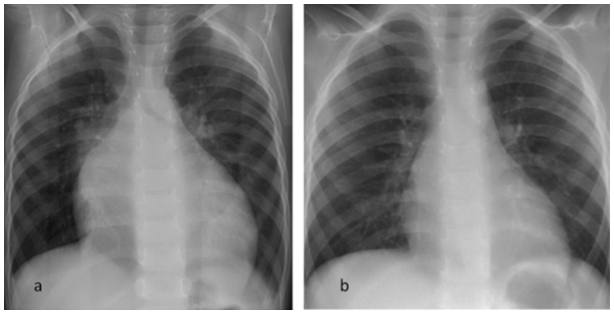


Figure 5 Case 2. Chest X-ray at presentation (a) showing marked cardiomegaly and (b) nice months of restoration of sinus rhythm showing reverse cardiac remodeling and normalization of cardiac size.

Case 2 presented with an ECG, also indicating an etiology for DCM. The persistent narrow QRS tachycardia with long R—P interval and abnormal P-wave axis raised suspicion of tachycardia-induced cardiomyopathy. After successful treatment of the tachycardia, most patients enjoy a favorable outcome with the recovery of ventricular function and reverse ventricular remodeling [9]. However, failure to recognize the tachycardia for an extended period of time could result in permanent myocardial damage or even sudden death. In a multicenter study that included eighty-one patients with tachycardia-induced DCM from 17 centers, Moore et al reported LV function recovery in 92% of the patients following control of the arrhythmia [9]. In that cohort, there was one mortality and two patients underwent heart transplant after controlling the arrhythmia. In a multivariate analysis, younger age and a faster tachycardia rate were associated with a higher chance for function recovery.

Our patient had tachycardia-induced DCM. With the restoration of a sinus rhythm after initiating amiodarone, the DCM resolved and reverse ventricular modeling led to normalized values. The plan is to consider ablation of the tachycardia mechanism once the patient is in an appropriate size to perform this procedure safely.

In both cases, the computer interpretations of the ECGs were not accurate in identifying the abnormalities. Despite the improvement in algorithms and programs that read ECGs, computer interpretation demonstrates frequent errors in the interpretation of rhythm disturbances [10,11]. Pediatricians' interpretation of ECGs also has a high rate of inaccuracy. A wide United Kingdom online study that

included 764 pediatricians demonstrated an accuracy rate of 64% only when interpreting 10 common clinical conditions with recognizable ECGs [12]. This alarming rate underscores the importance of teaching the skill of ECG interpretation in pediatric training programs and continuous medical education. Our cases suggest that having these skills will enable the ability to save lives and/or prevent morbidities.

5. Conclusion

It cannot be overstated that early recognition of a primary treatable cause of DCM along with relatively simple interventions could save patients' lives. Practicing pediatricians, as well as cardiologists, must be thorough and diligent in searching for a primary cause of DCM. The findings on an ECG, in particular, can be missed because they are dependent on the clinician's own expertise in interpreting the recording. In both cases that are reported here, the automated computer reading did not identify the diagnosis nor suggest the etiology, which emphasizes the importance of interpreting the ECG correctly. A second opinion is advised if the ECG findings are not clear.

Conflict of Interest

The authors declare that they have no financial or other relationship that constitute a conflict of interest.

References

[1] Yancy C, Jessup M, Bozkurt B, Wilkoff B. A report of the American College of Cardiology Foundation/American heart association task force on practice guidelines. Circulation 2013;128:e327.

[2] Wilkinson JD, Landy DC, Colan SD, Lipshultz SE. The pediatric cardiomyopathy registry and heart failure: key results from the first 15 years. Heart Fail Clin 2010 Oct;6(4):401—13.

[3] Jefferies J, Towbin J. Dilated cardiomyopathy. Lancet 2010 Feb;375(9716):752—62.

[4] Everitt M, Lu Minmin, Lipshultz S. Recovery of echocardiographic function in children with idiopathic dilated cardiomyopathy. J Am Coll Cardiol 2014;63(14):1405—13.

[5] Kantor P, Mertens L. Heart failure in children. Part I: clinical evaluation, diagnostic testing, and initial medical management. Eur J Pediatr 2010;169:269—79.

[6] Aladulgader A, Bulbul Z. Anomalous left coronary artery arising from the pulmonary artery in an adolescent: normalization of LV dimension and function following repair. Ann Saudi Med 2001;21:3—4.

[7] Houston A, Pollock J, Doig W, Murtagh E. Anomalous original of the left coronary artery from the pulmonary trunk: elucidation with colour doppler flow mapping. Br Heart J 1990 Jan; 63(1):50—4.

[8] Yarrabolu T, Ozcelik N, Quinones J, Balaguru D. Anomalous origin of left coronary artery from pulmonary artery — duped by 2D; saved by color doppler: echocardiographic lesson from two cases. Ann Pediatr Cardiol 2014 Sep;7(3):230—2.

[9] Moore J, Patel P, Shannon K, Balaji S. Pediatric of myocardial recovery in pediatric tachycardia-induced cardiomyopathy 2014 July;11(7):1163—9. http://dx.doi.org/10.1016/j.hrthm.2014.04.023.

[10] Chiu C, Hamilton R, Gow R, McCrindle B. Evaluation of computerized interpretation of the pediatric electrocardiogram. J Electrocardiol 2007 Apr;40(2):139—43.

[11] Shah A, Rubin S. Errors in the computerized electrocardiogram interpretation of cardiac rhythm. J Electrocardiol 2007 Sep-Oct;40(5):385—90.

[12] Jheeta J, Narayan O, Krasemann T. Accuracy in interpreting the paediatric ECG: a UK-wide study and the need for improvement. Arch Dis Child 2014 Jul;99(7):646—8.