CASE REPORT

Clinical improvement and no further need of transplant after closure of PDA with transcatheter approach in an end-stage heart failure patient with hypertrabeculation

Trabekülasyon artışının eşlik ettiği son dönem kalp yetersizliği olan olguda PDA'nın transkateter kapatılması sonrası görülen klinik düzelme ve kalp nakli ihtiyacının ortadan kalkması

Serhat Koca, M.D., Feyza Ayşenur Paç, M.D., Ajda Mutlu Mıhçıoğlu, M.D.,[#] Vedat Kavurt, M.D.,* Denizhan Bağrul, M.D.

Department of Pediatric Cardiology, Yüksek İhtisas Training and Research Hospital, Ankara, Turkey
[#]Department of Pediatric Cardiology, Sadi Konuk Training and Research Hospital, İstanbul, Turkey
*Department of Pediatric Cardiology, Diyarbakır Children Hospital, Diyarbakır, Turkey

Summary– Although heart failure is managed medically most of the time, heart transplantation is still last resort for selected end-stage heart failure patients with noncompaction cardiomyopathy. Presently described for the first time is case of pediatric patient with noncompaction cardiomyopathy who was initially referred to our hospital for heart transplant but underwent PDA repair and improved clinically without need for heart transplant.

Özet– Nonkompaksiyon kardiyomiyopatiye kalp yetersizliği sıklıkla eşlik eder. Bu hastalıkta görülen kalp yetersizliği tedavisinde, dilate kardiyomiyopatiye benzer şekilde tıbbi tedavi ya da tıbbi tedaviye dirençli olgularda kalp nakli uygulanabilir. Bu yazıda, kliniğimize kalp nakli planıyla yönlendirilen son dönem kalp yetersizliği olan nonkompaksiyon kardiyomiyopatili bir pediyatrik olguda, PDA'nın kapatılması ile düzelme sağlandı ve kalp nakiline ihtiyaç kalmadı.

Abbreviations:

PDA	*Patent ductus arteriosus*
VSD	*Ventricular septal defect*

Noncompaction cardiomyopathy, defined 80 years ago for the first time, is a type of cardiomyopathy that results from interruption during development of ventricular myocardium that takes place at 5 to 8 weeks of intrauterine life.[1] This cardiomyopathy comprises 9.5% of all childhood cardiomyopathies. Patients are diagnosed with echocardiography, noncompact to compact ratio of >1.4 in children and >2 in adults, trabeculae present in lateral/apical/inferior sites of left ventricle, and blood flow between intertrabecular recesses.[2,3] Primary goal in treatment is control of developing complications. Thromboembolic events, ventricular arrhythmias, and heart failure are commonly seen in this clinical entity. Heart failure can be treated with medical treatments as in dilated cardiomyopathy, and heart transplant can be performed in cases of resistance to medical treatment.[4]

Failure of ductus arteriosus, a blood vessel connecting the pulmonary artery to the proximal descending aorta, to close after birth is called patent ductus arteriosus (PDA). Due to left-to-right shunt formed by PDA, preload increases and dilatation in left heart chambers is seen. PDA should be closed when flow of shunt is excessive.

Heart failure becomes more evident in presence of lesion with left-to-right shunt and noncompaction. In the literature there are documented cases of ventricular septal defect (VSD) or PDA together with noncompaction for which correction of lesions with left-to-right shunts was preferred treatment.[5–8]

Received: June 18, 2016 *Accepted:* August 15, 2016

Correspondence: Dr. Serhat Koca. Türkiye Yüksek İhtisas Eğitim ve Araştırma Hastanesi, Pediatrik Kardiyoloji Kliniği, Kızılay Sokak, No: 4, Sıhhiye, Ankara, Turkey.
Tel: +90 312 - 306 11 36 e-mail: drserhatkoca@gmail.com

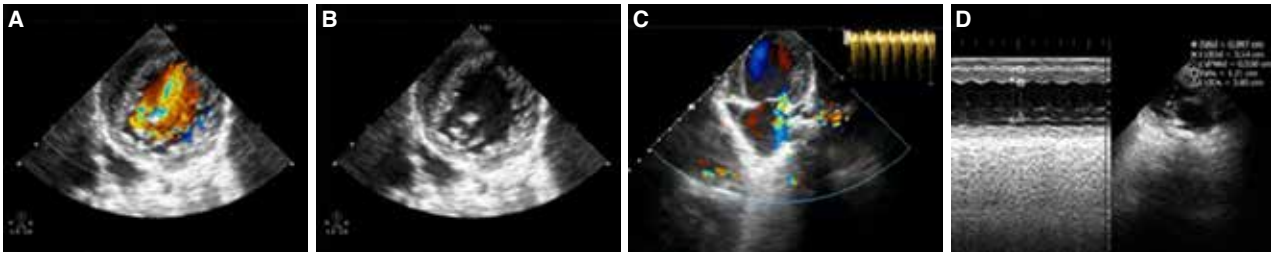

Figure 1. Echocardiographic findings of the patient at time of presentation: **(A)** prominent noncompaction, **(B)** enlarged left heart chambers, **(C)** mitral insufficiency, and **(D)** decreased left ventricular function.

Presently described is case of noncompaction cardiomyopathy leading to end-stage heart failure in which heart transplant was avoided due to clinical improvement with repair of PDA.

CASE REPORT

A 9-month-old child with heart failure due to noncompaction cardiomyopathy was referred to our hospital for heart transplant as last resort. Echocardiography revealed prominent noncompaction at apical and lateral sites of left ventricle (noncompact/compact: 0.95/0.48: 1.98), decreased left ventricular function (ejection fraction: 25%; fractional shortening: 12%), mild mitral insufficiency due to annular dilatation, PDA, and enlargement of the left ventricle (Figure 1).

Due to presence of heart failure and large PDA, PDA was closed with transcatheter approach under general anesthesia (Figure 2). After procedure, patient was transferred to the intensive care unit, extubated following day, and closely monitored for 5 days, during which patient showed clinical improvement (Figure 3). Patient was then monitored in ward and eventually discharged home with medical therapy. At 2-month follow-up, cardiac function showed tremendous improvement (ejection fraction: 62%; fractional shortening: 30%). Patient continued to gain weight

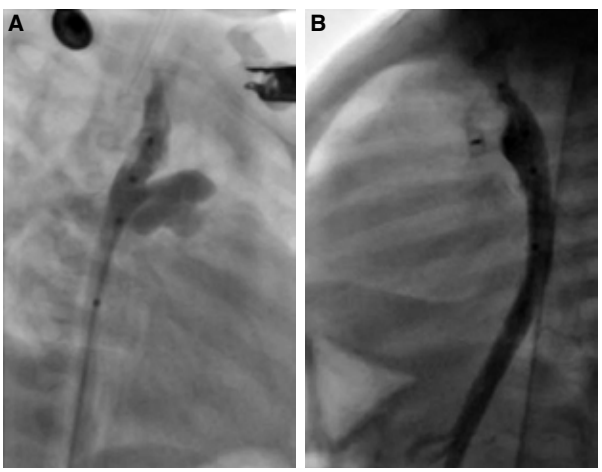

Figure 2. Closure of patent ductus arteriosus (PDA) with transcatheter approach under general anesthesia. **(A)** Right front oblique position, PDA is visible; **(B)** lateral position, occlusion of PDA with device.

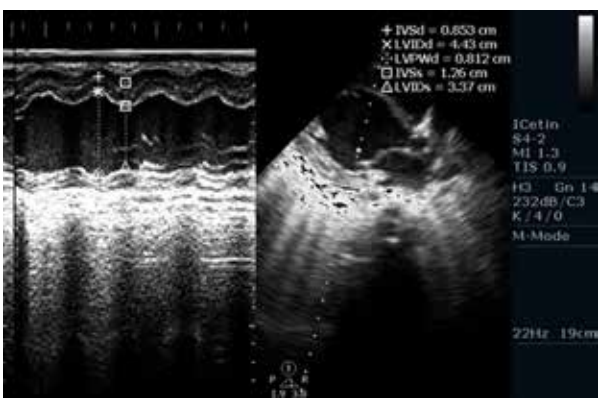

Figure 3. Improvement in cardiac contractility was seen after procedure.

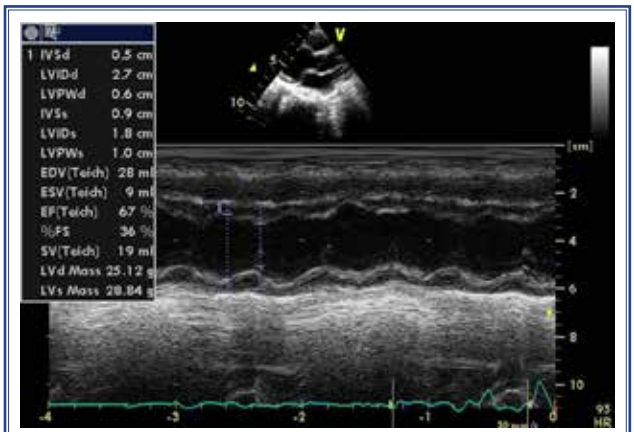

Figure 4. Echocardiographic findings show improvement in cardiac function.

normally and was followed-up without any clinical difficulties or complaints (Figure 4).

DISCUSSION

In treatment of heart failure, repairing underlying pathology, decreasing preload and afterload, and increasing contractility of heart are goals. In advanced cases of heart failure with primary cause, heart transplant is the last choice. Noncompaction cardiomyopathy is one cause of heart failure arising from impaired contractility of heart. PDA is another cause of heart failure, in which anatomical defect causes increasing preload as result of left-to-right shunt. In treatment of left-to-right shunt case, decreasing preload either medically or surgically is primary aim.

There are reports in the literature of heart failure cases due to noncompaction cardiomyopathy accompanied by left-to-right shunt (VSD, PDA), as well as other cardiac pathologies, such as aortic insufficiency and aortoventricular tunnel. In these cases, repairing defect resulted in improvement of cardiac function.[5–6] In some asymptomatic cases, or cases with minor symptoms in which PDA and noncompaction cardiomyopathy present together, although the patients did not have clinical heart failure, patients were treated with repair of PDA due to enlargement of left heart chambers seen on echocardiography.[7,8] In our case, advanced heart failure resulted from left-to-right shunt and noncompaction cardiomyopathy. Repair of shunt resolved patient's heart failure. To our knowledge, a case with noncompaction cardiomyopathy leading to heart failure treated by repairing isolated PDA has not yet been reported. Although the patient was followed-up asymptomatically, it is still possible that development of primary myocardial insufficiency or other complications due to noncompaction (thromboemboli, ventricular arrhythmia) may occur.

Conflict-of-interest issues regarding the authorship or article: None declared.

REFERENCES

1. Sedmera D, Pexieder T, Vuillemin M, Thompson RP, Anderson RH. Developmental patterning of the myocardium. Anat Rec 2000;258:319–37.
2. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation 1990;82:507–13.
3. Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart 2001;86:666–71.
4. Almeida AG, Pinto FJ. Non-compaction cardiomyopathy. Heart 2013;99:1535–42.
5. Myers PO, Tissot C, Cikirikcioglu M, Kalangos A. Complex aortic coarctation, regurgitant bicuspid aortic valve with VSD and ventricular non-compaction: a challenging combination. Thorac Cardiovasc Surg 2011;59:313–6.
6. Malakan Rad E, Zeinaloo AA. Postoperative normalization of left ventricular noncompaction and new echocardiographic signs in aorta to left ventricular tunnel. Echocardiography 2013;30:102–7.
7. Calzolari M, Gaddi O, Ilari B, Iotti R, Tortorella G, Muià N, Guiducci U. Association of non-compacted myocardium and patent ductus arteriosus in a young asymptomatic male patient: findings in screening for contact sport eligibility. [Article in Italian] Ital Heart J Suppl 2002;3:659–64.
8. Yang HJ, Song BG, Ma BO, Jeon GJ, Kim TH, Lee JU, et al. A rare combination of left ventricular noncompaction, patent ductus arteriosus, and persistent left superior vena cava demonstrated by multidetector computed tomography and echocardiography. Heart Lung 2012;41:35–8.

Keywords: Heart transplantation; noncompaction cardiomyopathy; patent ductus arteriosus.

Anahtar sözcükler: Kalp nakli; nonkompaksiyon kardiyomiyopati; patent duktus arteriyozus.