# Isolated Right Ventricular Noncompaction in an Adult Woman with Severe Pulmonary Hypertension Following at the Intensive Care Unit

**Mustafa Yıldız[1], Yasemin Özşahin[2], Hülya Yılmaz Ak[2], Doğaç Öksen[1]**

[1] Department of Cardiology, Istanbul University Cardiology Institute, Istanbul, Turkey
[2] Department of Anesthesiology and Reanimation, Istanbul University Cardiology Institute, Istanbul, Turkey

## ABSTRACT

Pulmonary hypertension is defined by a mean pulmonary artery pressure ≥ 25 mmHg at rest, measured during right heart catheterization. Ventricular noncompaction is a genetic cardiomyopathy which mostly effects left ventricle. It is related with deterioration of myocardial embryogenesis and commonly together with other cardiac diseases. Isolated ventricular non-compaction is characterized by modified morphology of myocardial wall, increased trabeculation in ventricular cavity and deep intertrabecular recesses. A 43-year-old woman presented exercise induced dyspnea and atypical chest pain. She has not any medical history prior. On admission, her 12 lead ECG showed complete righ bundle branch block, her blood pressure was 120/80 and pulse rate 80 per minutes. Transthoracic 2D echocardiogram and magnetic resonance imaging showed dilated and hypertrophied right ventricle with non-compaction of the right ventricular apex. The systolic pulmonary arterial pressure was 80 mmHg on the Doppler echocardiography. The coronary angiography revealed normal coronary arteries. The catheterization was showed pulmonary hypertension, right ventricle non-compaction and negative pulmonary vasoreactivity testing. Ventricular noncompaction, especially right ventricular noncompaction, complicated by severe pulmonary hypertension is exceptional. Only a few isolated right ventricular noncompaction has been reported but inclusion of pulmonary hypertesion cases are rare subsets. Diagnosis of pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic ventricular dysfunction secondary to right ventricular noncompaction. Widespread usage of cardiac magnetic resonance imaging, may enhance visual quality and evaluation of ventricular morphology, probably this will provide prevalance increment and clinical outcome improvements. Early diagnosis would bring better results.

**Key Words:** Pulmonary hypertension; ventricular noncompaction.

## Yoğun Bakım Ünitesinde Takip Edilen Şiddetli Pulmoner Hipertansiyonu Olan Kadın Hastada İzole Sağ Ventrikül Nonkompaksiyonu

## ÖZ

Pulmoner hipertansiyon, sağ kalp kateterizasyonu sırasında ölçülen, istirahatte ortalama pulmoner arter basıncı ≥ 25 mmHg olarak tanımlanır. Ventriküler nonkompaksiyon, çoğunlukla sol ventrikülü etkileyen genetik bir kardiyomiyopatidir. Miyokardiyal embriyogenezin bozulmasıyla ilişkilidir. İzole ventriküler nonkompaksiyon, miyokart duvarının morfolojisinin değişmesi, ventriküler kavitede trabekülasyon artması ve intertrabeküler girintilerin derinleşmesiyle karakterizedir. Tanı için iki boyutlu ekokardiyografi, kardiyak manyetik rezonans görüntüleme ve sağ ventrikülografi kullanılabilir. Kırk üç yaşında kadın hasta egzersizle ilişkili nefes darlığı ve atipik göğüs ağrısı şikayetiyle başvurmuştur. Bilinen kronik hastalığı olmayan hastanın başvuru sırasındaki EKG'sinde tam sağ dal bloğu görülmüş, kan basıncı 120/80 ve kalp atım hızı 80/dk bulunmuştur. Transtorasik 2D ekokardiyogram ve manyetik rezonans görüntülemede sağ ventrikül hipertrofik ve sağ ventrikül apeksinde nonkompaksiyon izlenmiştir. Doppler ekokardiyografide sistolik pulmoner arter basıncı 80 mmHg'dır. Bağ dokusu hastalıkları, kronik trombotik/embolik hastalık, anormal kardiyak ve pulmoner şantlar ve akciğer hastalığı gibi pulmoner hipertansiyonun ikincil nedenleri dışlanmıştır. Sol ve sağ kalp kateterizasyonu, koroner anjiyografi ve pulmoner vazoreaktivite testi yapılmıştır. Koroner anjiyografide koroner arterler normal bulunmuştur. Kateterizasyonda pulmoner hipertansiyon, sağ ventrikül nonkompaksiyon izlenmiş ve pulmoner vazoreaktivite testi negatif tespit edilmiştir. Şiddetli pulmoner hipertansiyon ile birlikte komplike sağ ventriküler nonkompaksiyon nadir görülen bir tablodur. Literatürde az sayıda izole sağ ventrikül nonkompaksiyonu bildirilmiştir. Nadir olması nedeniyle, tanısı son derece zordur. Pulmoner hipertansiyon tanısı, sağ ventrikül nonkompaksiyonuna sekonder sistolik ve diyastolik ventrikül disfonksiyonunun neden olduğu pulmoner venöz basınçların artmasıyla konulabilir. Altta yatan diğer mekanizma embolik olaylarla ilişkili olabilir. Kardiyak manyetik rezonans görüntülemenin yaygın kullanımı, ventriküler morfolojinin değerlendirilmesini kolaylaştırır. Erken teşhis klinik seyrin daha iyi olmasını sağlar.

**Anahtar Kelimeler:** Pulmoner hipertansiyon; ventriküler nonkompaksiyon.

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**Correspondence**

**Mustafa Yıldız**

**E-mail:** mustafayilldiz@yahoo.com
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## INTRODUCTION

Pulmonary hypertension is defined by a mean pulmonary artery pressure ≥ 25 mmHg at rest, measured during right heart catheterization. Connective tissue diseases, chronic thrombotic/embolic disease, cardiac and pulmonary shunts and lung disease are associated with pulmonary hypertension. Ventricular noncompaction is a genetic cardiomyopathy which mostly effects left ventricle, rarely involves right ventricle. It is related with deterioration of myocardial embryogenesis and commonly together with other cardiac diseases[1].

Isolated ventricular non-compaction is characterized by modified morphology of myocardial wall, increased trabeculation in ventricular cavity and deep intertrabecular recesses. Two-dimensional echocardiography, cardiac magnetic resonance imaging and right ventriculography can be used for diagnosis. The major clinical manifestations are heart failure, arrhythmias, sudden death and embolic events. In this case, we present right ventricular noncompaction complicated by severe pulmonary hypertension.

## CASE REPORT

A 43-year-old woman presented exercise induced dyspnea and atypical chest pain. She has not any medical history prior. There was not any family history of cardiomyopathy or cardiac death. On admission, her 12 lead ECG showed complete righ bundle branch block, her blood pressure was 120/80 and pulse rate 80 per minutes. Transthoracic 2D echocardiogram and magnetic resonance imaging showed dilated and hypertrophied right ventricle with non-compaction (ventricular trabeculations with deep intertrabecular recesses are seen; and color Doppler imaging demonstrates blood flow through these deep recesses in continuity with the ventricular cavity) of the right ventricular apex (Figure 1A, B). The systolic pulmonary arterial pressure was 80 mmHg on the Doppler echocardiography. Secondary causes for pulmonary hypertension like connective tissue diseases, chronic thrombotic/embolic disease, anomalous cardiac and pulmonary shunts and lung disease were excluded. She was received left and right heart catheterization, coronary angiogram and pulmonary vasoreactivity testing with inhalation of iloprost ($PGI_2$). The coronary angiography revealed normal coronary arteries. The catheterization was showed pulmonary hypertension, right ventricle non-compaction and negative pulmonary vasoreactivity testing (Figure 1C). Endothelin receptor antagonist (Bosentan) was prescribed for pulmonary artery hypertension.

## DISCUSSION

Ventricular noncompaction, especially right ventricular noncompaction, complicated by severe pulmonary hypertension is exceptional. Only a few isolated right ventricular noncompaction has been reported but inclusion of pulmonary hypertesion cases are rare subsets[2]. Due to rarity, diagnosis remains extremely difficult.

Mosty congenital heart diseases such as ventricular septal defect, aortic stenosis, coarctation and Ebstein's anomaly accompany right ventricular noncompaction. As echocardiographic evaluation of right ventricule hardly operator depended, may be some right ventricular noncompaction cases have been underestimated so far. Echocardiography of right ventricule is based on visual appearance. Magnetic resonance imaging may be accurately demonstrate trabeculations and deep recesses also segmental motions[2]. Color Doppler may be useful, if there is a remarkable deep perfusion in intertrabecular recesses[3].

Diagnosis of pulmonary hypertension may be a consequence of increased pulmonary venous pressures caused by systolic and diastolic ventricular dysfunction secondary to right ventricular noncompaction. Also, the other underlying mechanism may be associated with embolic events. Widespread usage of cardiac magnetic resonance imaging, may enhance visual quality and evaluation of ventricular morphology, probably this will provide prevalance increment and clinical outcome improvements. Early diagnosis would bring better results.

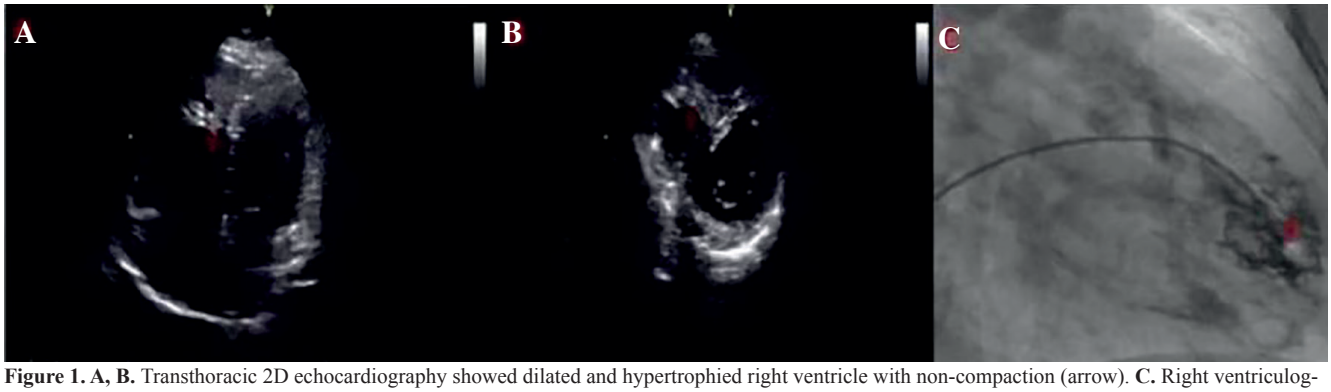


**Figure 1. A, B.** Transthoracic 2D echocardiography showed dilated and hypertrophied right ventricle with non-compaction (arrow). **C.** Right ventriculography showing the sponge-like appearance of the noncompacted ventricular Wall and marked retention of the contrast material in the trabecular recesses (arrows).

**Informed Consent:** Informed consent was obtained.

**Peer-review:** Externally peer-reviewed.

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