What Is the Structural Abnormality in This Patient?

ECG CHALLENGE
A 59-year-old man presented to our hospital reporting 3 months of exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, and lower leg edema. On physical examination, he had jugular venous distention, bibasilar rales, large tender liver, and peripheral edema. On cardiac auscultation, he had a regular heart rate with premature ventricular contractions, systolic ejection grade II/VI cardiac murmur in pulmonic area, and louder P₂ than A₂. Chest x-ray showed cardiac enlargement, and an ECG (Figure 1) was recorded; because of premature heartbeats on admission, a 24-hour Holter ECG was obtained (Figure 2). On the basis of ECG findings, what is the rhythm of the patient and what is the structural abnormality? Please turn the page to read the diagnosis.

Figure 1. ECG obtained during emergency department admission.

Figure 2. Twenty-four–hour Holter ECG showing 1 premature ventricular contraction.
RESPONSE TO ECG CHALLENGE

To interpret the ECG and determine the anatomic abnormality in this case, it is necessary to take into account the clinical and ECG findings in combination, which are remarkable for right ventricle (RV) failure as demonstrated by the findings on physical examination of jugular venous distention, large tender liver, and peripheral lower leg edema. In addition, there is a systolic murmur, fixed splitting of S₂ and louder P₂, thus suggesting an interatrial septal defect.

The ECG shows a sinus rhythm with an extremely tall P wave in inferior and precordial leads and notable increased duration and depth of terminal-negative portion of P wave in V1 (mimicking rS morphology of a QRS). Other ECG findings include first-degree atrioventricular conduction block, RV hypertrophy, and nonspecific repolarization changes (Figure 3). The high-voltage P waves are observed in the 24-hour Holter ECG as well, where they can even generate confusion with the QRS morphology, suggesting ventricular bigeminy, but following them through the other channels clarifies that they represent P waves, which in second channel have even higher voltage than QRS (Figure 4).

Cardiac magnetic resonance imaging confirmed an ostium secundum atrial septal defect (ASD) of 13 mm, left-to-right shunt, Qp/Qs=2.1, giant right atrium (end-systole area of 63 cm²; normal reference, <18 cm²), and signs of volume and pressure overload to the RV with severe dilation (diastolic diameter, 59 mm), mild hypertrophy, TAPSE (Tricuspid Annular Plane Systolic Excursion): 6 mm, and a RV ejection fraction of 16% (Figure 5). The patient also had a severe impairment of left ventricle function with an ejection fraction of 15%; however, no explanation was found for this latest finding after the diagnostic workup.

ASDs are among the most common presentations of congenital heart diseases in adults. ECG characteristics of ASD include right bundle-branch block, which could be incomplete because of RV hypertrophy. Right QRS axis deviation is associated with ostium secundum ASD, the QRS is leftward in ostium primum defects, and inverted P waves in inferior leads may be seen in sinus venous ASD. Carmichael et al. described that, in patients with ASD, right bundle-branch block pattern and RV hypertrophy were seen in 66% and 25% of cases, respectively. Patients with normal pulmonary pressure more commonly have right bundle-branch block, whereas patients with higher pulmonary pressure develop QR morphology in V1 with incomplete right bundle-branch block. As an explanation to this, they argued that the higher the pressure in the RV, the higher the prob-

Figure 3. ECG findings.
Sinus rhythm, atrioventricular delay with a PR segment of 400 ms (A, solid line), high-voltage P waves are identified (dotted arrow in A, B, and C), and they precede every regular QRS complex (solid arrow in A and B), P waves have higher amplitude than QRS in V1 (B); QRS axis deviated to the right (121°, D), and morphology is compatible with incomplete RBBB because of QR in V1 (B) and right ventricle hypertrophy based on R/S <1 and S >7 mm in lead V6 (C). R in V1 + S in V6>10.5 mm and deep S waves in V4 to V6 (B and C). T waves are represented by arrow head in DII and V5 (A and C). RBBB indicates right bundle-branch block.
ability of developing RV hypertrophy, which generates an electric vector that counterbalances the left ventricle depolarization in V1; therefore, a predominantly positive deflection is seen in this lead. These findings are consistent with those in our patient (see Figure 3B); he had right-axis deviation and qR morphology in V1 that was consistent with his severe pulmonary artery hypertension, thus explaining clinical and ECG changes of RV hypertrophy and failure.

In summary, the ECG presented in this case shows changes consistent with secundum ASD, which include rightward axis deviation and incomplete right bundle-branch morphology. In addition, the ECG also showed extremely high-voltage P waves that correlated with a large dilated right atrium on cardiac MRI. The MRI fits criteria for giant atrium. The presence of such P waves is not commonly described on ECGs of patients with ASD, and this finding made the approach to our patient difficult at the beginning because the P waves were misinterpreted as bigeminy rhythm on ECG and 24-hour Holter monitoring.

ACKNOWLEDGMENTS
The authors thank Dr. Carlos José Jaramillo (Cardiologist, Departamento de Cardiología, Universidad de Antioquia) for advice regarding the ECG interpretation and Dr Pedro Abad (Radiologist, Instituto de Alta Tecnología Médica) for helping to interpret and organize cardiac magnetic resonance images.

DISCLOSURES
None.

FOOTNOTES
Circulation is available at http://circ.ahajournals.org.

REFERENCES
Figure 5. Cardiovascular magnetic resonance.
Axial (A) and 4-chamber (B) steady-state with free precession (SSFP) images correlating with physical and ECG findings. See the huge right atrium and the atrial septal defect (ASD) with black arrow heads pointing to the tips of the ASD. LA indicates left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle; and TV, tricuspid valve.

What Is the Structural Abnormality in This Patient?
Andrés F. Miranda-Arboleda, Jairo Gándara-Ricardo, Edwin F. Arévalo-Guerrero and Edison Muñoz-Ortiz

Circulation. 2017;135:808-811
doi: 10.1161/CIRCULATIONAHA.116.026903

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/135/8/808

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/