An asymptomatic 29-year-old female was referred for evaluation of a cardiac murmur. A preappointment electrocardiogram revealed right axis deviation, right bundle-branch block, and “crochetage sign” with notched R waves in the inferior leads (Figure 1A, arrows). Physical examination revealed normal jugular venous examination and a 2+ parasternal lift. Cardiac auscultation at the left upper sternal border (Figure 1B, phonocardiogram; Audio file in the Data Supplement) revealed normal S1, grade 2/6 early-peaking systolic ejection murmur, and fixed splitting of the second heart sound (A2-P2). Chest radiography (Figure 1C) showed enlarged right heart border (arrow) and prominent pulmonary arteries (arrow head). Transthoracic echocardiogram showed moderate right ventricular enlargement and large (28×18 mm) ostium secundum atrial septal defect (Figure 1D, asterisk; Movie I in the Data Supplement) with left-to-right shunting by color Doppler (Figure 1E; Movie II in the Data Supplement), and estimated right ventricular systolic pressure was 26 mmHg.

The term “fixed” denotes absence of significant respiratory variation of A2-P2 and is a classic feature of ostium secundum atrial septal defect.1,2 Although the mechanism is incompletely understood, one hypothesis suggests that reciprocal respirophasic augmentation of systemic venous return and left-to-right shunting (ie, increased systemic venous return and decreased shunting with inspiration; vice versa with expiration) are key hemodynamic features. The cumulative effect is relatively static right ventricular stroke volume throughout the respiratory cycle.3 In the isolated right bundle-branch block, the A2-P2 interval is wide but still shows respirophasic changes. The systolic ejection murmur results from increased flow across the right ventricular outflow tract, and occasionally a soft diastolic rumble originating from the tricuspid valve is present.

The patient underwent successful percutaneous device closure with a 34-mm Amplatzer septal occluder. A preprocedure transesophageal echocardiogram with 3-dimensional imaging was performed; an oval shaped atrial septal defect was identified, with the major axis measuring 26 mm (Figure 2; Movie III in the Data Supplement). The calculated shunt fraction (Qp:Qs) was 3:1 before percutaneous device closure. There was no residual shunt postprocedure.

Disclosures
None.

References

Key Words: atrial septal defect ■ blood pressure ■ bundle-branch block ■ heart murmurs ■ stroke volume
Figure 1. A shows the electrocardiographic findings of right bundle branch block, right axis deviation, and “crochetage sign.” B is a phonocardiogram recorded at the bedside demonstrating soft systolic ejection murmur (SEM) and fixed splitting of the second heart sound. C highlights the chest X-ray findings of right atrial (arrow head) and right ventricular enlargement (arrow). Transthoracic echocardiogram with 4 chamber view showing a defect of the atrial septum (asterisk, D) with a large left-to-right shunt on color Doppler (E). A2 indicates aortic closure; LA, left atrium; LV, left ventricle; P2, pulmonic closure; RA, right atrium; RV, right ventricle; and SEM, systolic ejection murmur. See text for details.
Figure 2. Three-dimensional transesophageal echocardiogram demonstrating the oval shaped, ostium secundum atrial septal defect measuring 26 mm across the major axis.
Fixed…Or Needs Fixing?
D. Brian Newman, William R. Miranda and Heidi M. Connolly

doi: 10.1161/CIRCIMAGING.116.004877

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/9/5/e004877

Data Supplement (unedited) at:
http://circimaging.ahajournals.org/content/suppl/2016/05/06/CIRCIMAGING.116.004877.DC1
http://circimaging.ahajournals.org/content/suppl/2016/05/06/CIRCIMAGING.116.004877.DC2

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Cardiovascular Imaging 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: Cardiovascular Imaging is online at:
http://circimaging.ahajournals.org/subscriptions/
A 29-year-old female was referred for evaluation of a cardiac murmur. A preappointment electrocardiogram revealed right axis deviation, right bundle-branch block, and “crochetage sign” with notched R waves in the inferior leads (Figure 1A, arrows). Physical examination revealed normal jugular venous examination and a 2+ parasternal lift. Cardiac auscultation at the left upper sternal border (Figure 1B, phonocardiogram; Audio file in the Data Supplement) revealed normal S1, grade 2/6 early-peaking systolic ejection murmur, and fixed splitting of the second heart sound (A2-P2). Chest radiography (Figure 1C) showed enlarged right heart border (arrow) and prominent pulmonary arteries (arrow head). Transthoracic echocardiogram showed moderate right ventricular enlargement and large (28×18 mm) ostium secundum atrial septal defect (Figure 1D, asterisk; Movie I in the Data Supplement) with left-to-right shunting by color Doppler (Figure 1E; Movie II in the Data Supplement), and estimated right ventricular systolic pressure was 26 mm Hg.

The term “fixed” denotes absence of significant respiratory variation of A2-P2 and is a classic feature of ostium secundum atrial septal defect. Although the mechanism is incompletely understood, one hypothesis suggests that reciprocal respirophasic augmentation of systemic venous return and left-to-right shunting (ie, increased systemic venous return and decreased shunting with inspiration; vice versa with expiration) are key hemodynamic features. The cumulative effect is relatively static right ventricular stroke volume throughout the respiratory cycle. In the isolated right bundle-branch block, the A2-P2 interval is wide but still shows respirophasic changes. The systolic ejection murmur results from increased flow across the right ventricular outflow tract, and occasionally a soft diastolic rumble originating from the tricuspid valve is present.

The patient underwent successful percutaneous device closure with a 34-mm Amplatzer septal occluder. A preprocedure transesophageal echocardiogram with 3-dimensional imaging was performed; an oval shaped atrial septal defect was identified, with the major axis measuring 26 mm (Figure 2; Movie III in the Data Supplement). The calculated shunt fraction (Qp:Qs) was 3:1 before percutaneous device closure. There was no residual shunt postprocedure.

Disclosures

None.

References

Figure 1. A shows the electrocardiographic findings of right bundle branch block, right axis deviation, and “crochetage sign.” B is a phonocardiogram recorded at the bedside demonstrating soft systolic ejection murmur (SEM) and fixed splitting of the second heart sound. C highlights the chest X-ray findings of right atrial (arrow head) and right ventricular enlargement (arrow). Transthoracic echocardiogram with 4 chamber view showing a defect of the atrial septum (asterisk, D) with a large left-to-right shunt on color Doppler (E). A2 indicates aortic closure; LA, left atrium; LV, left ventricle; P2, pulmonic closure; RA, right atrium; RV, right ventricle; and SEM, systolic ejection murmur. See text for details.
Figure 2. Three-dimensional transesophageal echocardiogram demonstrating the oval shaped, ostium secundum atrial septal defect measuring 26 mm across the major axis.
Fixed... Or Needs Fixing?
D. Brian Newman, William R. Miranda and Heidi M. Connolly

doi: 10.1161/CIRCIMAGING.116.004877
*Circulation: Cardiovascular Imaging* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2016 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-9651. Online ISSN: 1942-0080

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/9/5/e004877

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in *Circulation: Cardiovascular Imaging* 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: Cardiovascular Imaging* is online at:
http://circimaging.ahajournals.org//subscriptions/

Downloaded from http://circimaging.ahajournals.org/ by Rose Brister on May 6, 2016
An asymptomatic 29-year-old female was referred for evaluation of a cardiac murmur. A preappointment electrocardiogram revealed right axis deviation, right bundle-branch block, and “crochetage sign” with notched R waves in the inferior leads (Figure 1A, arrows). Physical examination revealed normal jugular venous examination and a 2+ parasternal lift. Cardiac auscultation at the left upper sternal border (Figure 1B, phonocardiogram; Audio file in the Data Supplement) revealed normal S1, grade 2/6 early-peaking systolic ejection murmur, and fixed splitting of the second heart sound (A2-P2). Chest radiography (Figure 1C) showed enlarged right heart border (arrow) and prominent pulmonary arteries (arrow head). Transthoracic echocardiogram showed moderate right ventricular enlargement and large (28×18 mm) ostium secundum atrial septal defect (Figure 1D, asterisk; Movie I in the Data Supplement) with left-to-right shunting by color Doppler (Figure 1E; Movie II in the Data Supplement), and estimated right ventricular systolic pressure was 26 mmHg.

The term “fixed” denotes absence of significant respiratory variation of A2-P2 and is a classic feature of ostium secundum atrial septal defect.1,2 Although the mechanism is incompletely understood, one hypothesis suggests that reciprocal respirophasic augmentation of systemic venous return and left-to-right shunting (ie, increased systemic venous return and decreased shunting with inspiration; vice versa with expiration) are key hemodynamic features. The cumulative effect is relatively static right ventricular stroke volume throughout the respiratory cycle.3 In the isolated right bundle-branch block, the A2-P2 interval is wide but still shows respirophasic changes. The systolic ejection murmur results from increased flow across the right ventricular outflow tract, and occasionally a soft diastolic rumble originating from the tricuspid valve is present.

The patient underwent successful percutaneous device closure with a 34-mm Amplatzer septal occluder. A preprocedure transesophageal echocardiogram with 3-dimensional imaging was performed; an oval shaped atrial septal defect was identified, with the major axis measuring 26 mm (Figure 2; Movie III in the Data Supplement). The calculated shunt fraction (Qp:Qs) was 3:1 before percutaneous device closure. There was no residual shunt postprocedure.

Disclosures

None.

References


Key Words: atrial septal defect ■ blood pressure ■ bundle-branch block ■ heart murmurs ■ stroke volume
Figure 1. A shows the electrocardiographic findings of right bundle branch block, right axis deviation, and "crochetage sign." B is a phonocardiogram recorded at the bedside demonstrating soft systolic ejection murmur (SEM) and fixed splitting of the second heart sound. C highlights the chest X-ray findings of right atrial (arrow head) and right ventricular enlargement (arrow). Transthoracic echocardiogram with 4 chamber view showing a defect of the atrial septum (asterisk, D) with a large left-to-right shunt on color Doppler (E). A2 indicates aortic closure; LA, left atrium; LV, left ventricle; P2, pulmonic closure; RA, right atrium; RV, right ventricle; and SEM, systolic ejection murmur. See text for details.
Figure 2. Three-dimensional transesophageal echocardiogram demonstrating the oval shaped, ostium secundum atrial septal defect measuring 26 mm across the major axis.
Fixed...Or Needs Fixing?
D. Brian Newman, William R. Miranda and Heidi M. Connolly

doi: 10.1161/CIRCIMAGING.116.004877
Circulation: Cardiovascular Imaging is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2016 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-9651. Online ISSN: 1942-0080

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/9/5/e004877

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Cardiovascular Imaging 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: Cardiovascular Imaging is online at:
http://circimaging.ahajournals.org//subscriptions/