An Unusual Equatorial Constriction of the Ventricular Cone

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ABSTRACT

A 25-year-old woman with a recent diagnosis of congenital heart disease and probable endocarditis was referred to our institution. During our evaluation we observed an unusual deformation of both ventricles. We discuss its possible origin as revealed by printing of a three-dimensional model. (Level of Difficulty: Advanced.) (J Am Coll Cardiol Case Rep 2022;4:1156–1159) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 25-year-old woman was referred for treatment at our institution with a recent diagnosis of pulmonary valvar disease and probable endocarditis. Over the past 3 months she had experienced progressive dyspnea, consistent with New York Heart Association functional class III heart failure, and a prolonged fever. On examination, systolic pulsations were palpable in the left parasternal area. Auscultation revealed the first heart sound to be normal, with a decrease in the pulmonary component of the second heart sound. A third heart sound was audible in the left parasternal fourth intercostal space, along with a middiastolic ejection murmur at the middle to upper left sternal border. A decrescendo diastolic murmur was heard in the second and third left intercostal spaces, which increased in intensity during inspiration and decreased during the Valsalva maneuver. During our evaluation of the imaging studies, we observed a finding in the ventricular walls that, to the best of our knowledge, is unique. Although we are sure that the finding was not the cause of the symptoms, nor of the findings at physical examination, the images are so unusual that we believe they are deserving of comment.

LEARNING OBJECTIVES

- To describe a unique case of deformation of the ventricular cone, produced by failed formation of papillary muscles.
- To show evidence in favor of remodeling of excessive trabeculation.
- To recognize the usefulness of 3D printing in the study of complex cases.

MEDICAL HISTORY

The patient had no additional significant medical history, had no cardiovascular risk factors, and did not describe episodes of either chest pain, edema, arrhythmia, or syncope.
**DIFFERENTIAL DIAGNOSIS**

Inasmuch as we have been unable to find comparable cases, we are unable to suggest a differential diagnosis for the rare finding in our patient.

**INVESTIGATIONS**

CT demonstrated valvar and subvalvar pulmonary stenosis, with the images compatible with the presence of a vegetation in the pulmonary trunk. The trunk and its branches were dilated, and the oval foramen was patent. The remarkable finding revealed by the images was the deformation at the level of the middle third of both ventricles. The images also showed that the left ventricular papillary muscles were not grouped in the anticipated fashion. Instead, they were located at the basal third of the ventricular cone, which also showed markedly excessive trabeculation (Figures 1A to 1E). The right ventricle was dilated, with hypertrophied walls interpreted as being secondary to the valvar disease. As in the left ventricle, we also observed deformation of its walls at the level of the papillary muscles (Figures 1A to 1C, Video 1).

The 2-dimensional images were confirmed by the volumetric reconstruction (Figure 1F). Owing to the

![Figure 1](image-url)
rarity of the findings, we decided to make a 3-dimensional printed model. The printer used was 3D Professional Industrial Creality Cr-5 Pro, and the material of the model was PLA BASIC 1.75-mm filament (Figure 2, Supplemental File). This confirmed the extent of the equatorial constriction of both ventricles, permitting us also to demonstrate the findings as would be seen in the frontal view of a chest radiograph. Despite the abnormal findings, however, the global, segmental mobility, and systolic functions were all normal, and there was no obstruction in the left ventricular outflow tract. Additional investigation with magnetic resonance imaging (Figure 3), ruled out any myocardial ischemia or infarction. No intraventricular gradients were found (Videos 2 and 3).

MANAGEMENT

Inasmuch the investigations indicated that the abnormal findings in the ventricular walls did not produce any hemodynamic alteration, we confined ourselves to treating the right ventricular problems. The patient underwent placement of a pulmonary valve prosthesis, enlargement of the right ventricular outflow tract, infundibulectomy, resection of the vegetation, and closure of the patent oval foramen.

DISCUSSION

We suggest that this deformation was produced by incomplete formation of the papillary muscles, with this process “pinching” the ventricular cone. It is significant that the constriction was found at the
junction between the ventricular inlets and the apical components. There was also evidence of excessive trabeculation. The images show that the compact layer was very well formed but that the trabeculations had themselves also compacted. We suggest that this implies abnormal remodeling of the trabecular layer, which normally would form only the papillary muscles. Supporting our interpretation is the presence of an anomalous muscle bundle in the right ventricle. There was no evidence to support the notion that the trabeculations coalesce to form the compact ventricular walls. However, it is becoming increasingly evident that they do compact to form the papillary muscles. We suggest that this process had gone astray in our patient, producing an equatorial constriction of the ventricular cone. However, we found no evidence that this anatomical constriction produced any alteration in the ventricular function.

FOLLOW-UP

Three years after the surgical treatment of the right-sided problems, the patient had reported no cardiovascular symptoms. Repeated magnetic resonance imaging revealed dysfunction of the pulmonary prosthesis, albeit without any evidence of abnormal hemodynamic behavior at the level of the deformation.

CONCLUSIONS

We report the previously undescribed finding of equatorial constriction of both ventricles, in the absence of any alteration in ventricular function.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS computed tomography, congenital heart defect, papillary muscles, three-dimensional printing