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IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Prominent Neck and Pinna Pulsations



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ABSTRACT

A young man presented with visible neck and pinna pulsations after a palliative bidirectional cavopulmonary (Glenn) shunt with open antegrade flow. This resulted in giant C-V waves in the jugular venous pulse. Giant C-V waves resulting from a Glenn procedure have not previously been described and represent a novel entity to recognize. (Level of Difficulty: Advanced.) (J Am Coll Cardiol Case Rep 2021;3:1780-1781) © 2021 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/).

19-year-old man presented with worsening facial swelling for the preceding 10 months. He also reported prominent neck pulsations over the last 6 months. His past history was significant for a palliative bidirectional cavopulmonary (bidirectional Glenn) shunt with antegrade flow through a patent pulmonary valve that was performed 10 years earlier for single-ventricle physiology and a double-outlet right ventricle with a large ventricular septal defect and severe pulmonary stenosis. Physical examination revealed atrial fibrillation with a mean heart rate of 98 beats/min and blood pressure of 96/60 mm Hg, pandigital clubbing, and central cyanosis. Also seen were bounding neck and pinna pulsations in conjunction with cardiac systole on the electrocardiogram (ECG) (Figure 1A [green arrows], Video 1). The pulsations initially appeared to be caused by prominent arterial pulsations arising from the carotid arteries. Cardiac examination revealed a loud holosystolic murmur in the left parasternal region suggestive of severe tricuspid regurgitation. Duplex ultrasonography of the neck confirmed a grossly dilated right internal jugular vein with significant retrograde flow during systole indicating giant C-V waves in the jugular venous pulse as the cause (Figure 1B, Video 2). The patient had a brief period of medical stabilization, following which he successfully underwent a Fontan procedure and was relieved of his symptoms.

Giant C-V waves have been described in severe tricuspid regurgitation (1). Interestingly, in patients who have undergone a Glenn procedure, the superior vena cava (SVC) is attached to the right pulmonary artery in an end-to-side fashion following interruption of the right atrial–SVC junction (2). Thus, prominent C-V waves in this case could not have resulted from tricuspid regurgitation because the right atrium was no longer in continuity with the SVC. Instead, they were likely caused by direct transmission of right ventricular systolic pressure waves through the cavopulmonary circuit by to-and-fro flow through the pulmonary valve. Furthermore, the pulsations were visible until they were above the angle of the mandible, a finding suggestive of grossly elevated jugular venous pressures and ventricularization. Giant C-V waves resulting from a Glenn procedure have not previously been described, making this a rare and novel entity to recognize.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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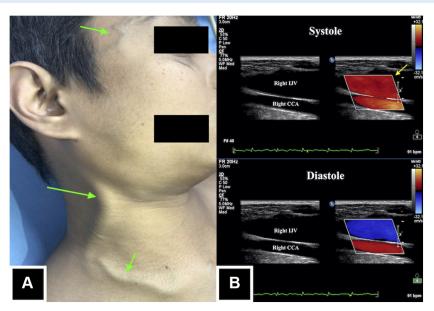
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ABBREVIATIONS AND ACRONYMS

ECG = electrocardiogram **SVC** = superior vena cava

FIGURE 1 Prominent Neck, Pinna, and Forehead Pulsations



(A) A representative still image depicting prominent pulsations and distended veins seen over the neck, pinna, and forehead (arrows). (B) Duplex ultrasonography confirmed a normal right common carotid artery (CCA) and a grossly dilated right internal jugular vein (IJV) with torrential retrograde (red) flow during systole (arrow) and antegrade (blue) flow during diastole.

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KEY WORDS congenital heart disease, heart failure, jugular venous pulsation

APPENDIX For supplemental videos, please see the online version of this paper.