

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

IMAGING VIGNETTE: CLINICAL VIGNETTE

Heart Block in Unpalliated Complex Adult Congenital Heart Disease



Massive Collateral Burden Precludes Epicardial Pacemaker

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ABSTRACT

Complete heart block is a common complication for adults with congenital heart disease (CHD). Epicardial pacing is preferred in patients with septal shunting due to risk of thromboembolism. Anatomic changes in complex CHD may preclude surgical epicardial lead placement. Thromboembolism risk reduction in such patients requiring endocardial pacing remains questionable. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:192-3) © 2021 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/>).

A 50-year-old woman with cyanotic, single-ventricle heart disease consisting of unbalanced atrioventricular septal defect, double outlet right ventricle, and functional pulmonary atresia, prior stroke, and secondary polycythemia presented with dyspnea and fatigue despite supplemental oxygen and sildenafil. Many years prior, she had received a late diagnosis of congenital heart disease (CHD) and did not receive palliative surgical correction. On presentation, she was cyanotic, afebrile, normotensive, and newly bradycardic. Electrocardiogram showed complete atrioventricular (AV) block with junctional and ventricular bigeminy and rate of 30 beats per minute. Echocardiogram found normal ventricular function and mild right AV valve regurgitation. Epicardial pacemaker placement was judged too high-risk because of innumerable thoracic collateral vessels (Video 1). Risks and benefits of an endocardial pacing system were discussed with the patient. A transvenous dual-chamber pacemaker was placed and warfarin started. At 1-week follow-up, dyspnea and fatigue were improved.

One month after discharge, she had witnessed sudden-onset loss-of-consciousness. While hemodynamically stable and not in cardiac arrest, she was persistently unresponsive. She was intubated, life-flighted, and admitted to the intensive care unit. Initial studies included hemoglobin 22.2 g/dl (baseline 20 to 21 g/dl), platelet count 174,000/ μ l, international normalized ratio 2.0, and arterial blood gas pH 7.06, pCO₂ 72, and pO₂ 41. Admission computed tomography (CT) of the brain did not show a new intracranial process. Magnetic resonance imaging of the brain was not performed. CT pulmonary embolism study was also negative. She remained unresponsive. Three days later, repeat CT brain showed evolving right middle and posterior cerebral artery territory strokes to which her acute presentation was attributed.

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Her hospital course was complicated by cardiogenic shock and subsequent liver and renal failure. She underwent continuous renal replacement therapy complicated by hypotension. Respiratory and metabolic acidosis worsened despite optimal medical treatment. Bronchoscopy was performed due to massive left lung atelectasis, during which she became hypotensive and difficult to ventilate. At this time, her family elected to withdraw aggressive measures.

This patient's substantial thoracic collateral vessel burden suggested substantial surgical bleeding risk and was a contraindication for epicardial pacemaker placement via sternotomy, subxiphoid, or thoracoscopic approach. Endocardial pacemakers with transvenous leads have been associated with increased thromboembolism risk (hazard ratio [HR]: 2.6); warfarin and/or aspirin use was not protective, although the study was not designed or powered for this comparison (1). Previous reports suggest a similar risk of thromboembolism and dislocation between leadless and transvenous pacemakers (2,3). In addition, in certain atrial tachyarrhythmia in CHD without transvenous pacemaker, risk of thromboembolism increased with CHD severity (HR: 3.47 on multivariable analysis) but not with anticoagulation use (HR: 1.07, not significant); major bleeding was a significant complication in patients treated with anticoagulation (4).

This patient's mixing single-ventricle lesion, loss of sinus rhythm, prior stroke, erythrocytosis, and 2 new transvenous pacemaker leads likely signaled significantly elevated thromboembolic risk. Anticoagulation merited consideration in the absence of evidence-based guidelines, but ultimately was not effective in preventing stroke in this patient. Sadly, patients with such disease have little cardiac reserve; complications can trigger significant decline, including multiorgan failure, as seen in this case. Additional studies are needed to better understand optimal device and anticoagulation therapy in patients with CHD requiring pacing therapy.

ABBREVIATIONS AND ACRONYMS

AV = atrioventricular
CHD = congenital heart disease
CT = computed tomography

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