Iatrogenic Gerbode-type defect after surgical correction of double-outlet-right ventricle

Marcin Dominiak¹, Łukasz Chrzanowski¹, Ludomir Stefańczyk², Jarosław Kasprzak¹

¹Chair and Department of Cardiology, Medical University of Łódź, Łódź, Poland
²Department of Radiology and Diagnostic Imaging, Medical University of Łódź, Łódź, Poland

Gerbode-type atroventricular septal defect is a unique variant of a shunt lesion representing a rare congenital entity [1] or acquired disease. Its specific feature is the communication between the left ventricle and the right atrium. We present a case of a 34-year-old man who underwent a total surgical correction of the double-outlet right ventricle in 2002 (37-mm patch repair of the intraventricular septum, pulmonary valvuloplasty with a patch inserted in the right ventricular outflow tract) referred to our Department for echocardiographic examination due to holosystolic murmur heard over the left sternal border. On the day of examination, the patient was stable in New York Heart Association class II, without history of preceding infection, chest injury, or myocardial infarction. Transthoracic echocardiography revealed a communication with left-to-right shunt on the upper border of the surgical patch closing the ventricular septal defect (VSD) (Fig. 1A) with peak velocity of 5 m/s at systemic blood pressure of 125/70 mmHg. In addition, a previously undiagnosed unusual flow between the left ventricle and the right atrium was recorded, consistent with Gerbode-type atroventricular defect (Fig. 1D). On transoesophageal echocardiography, a shunt from the left ventricle to the right atrium was confirmed (Fig. 1B, C) through a 3-mm defect, probably residual to the surgical correction procedure. Cardiac magnetic resonance imaging was performed, reconfirming the presence of communication between the left and the right ventricles on the edge of the surgical patch (Fig. 1E) and the presence of a shunt between the left ventricle and the right atrium through an iatrogenic Gerbode-type defect (Fig. 1F). Due to the patient preference and small shunt flow volume with normal pulmonary pressures, the management was conservative. Gerbode defect is a rare congenital lesion, first described in 1857 by Buhl. It may also emerge after cardiac surgery, chest trauma, or endocarditis. It represents an estimated 1% to 2% of VSDs undergoing surgery [2]. Atrioventricular septal defect may be direct (consistent with current definition and presented case) or indirect — where the abnormal flow passes from the left to the right ventricle and further to the right atrium across the perforated base of the septal tricuspid leaflet. According to the original description of case series by American surgeon Frank Leven Albert Gerbode, it may be treated with surgical closure [3, 4] or, more recently, by percutaneous occluder implantation. Only a few cases of an acquired Gerbode defect were reported as a complication of complex congenital surgery as suspected in our patient. Importantly, this lesion has potential clinical consequences and is associated with a four-fold higher risk for infective endocarditis in comparison to patients with VSD [5].

References

Figure 1. A. Echocardiographic apical view demonstrating a colour-coded shunt flow between the left ventricle (LV) and the right atrium (RA) through the Gerbode-type defect. The second turbulent flow within the right ventricle (RV) results from the residual ventricular septal defect. B. Transoesophageal echocardiographic image of Gerbode defect (arrow). C. Transoesophageal echocardiographic image of corresponding colour-coded flow. D. Continuous Doppler tracings showing estimated systolic pressure gradient of 80 mmHg between the LV and the RA recorded at the Gerbode defect. E. Cardiac magnetic resonance image of communication between the LV and RV (residual septal defect on the edge of the surgical patch). F. Cardiac magnetic resonance imaging demonstrating the flow between LV and RA through Gerbode-type defect; Ao — aorta; LA — left atrium; arrow — rapid shunt flow

Address for correspondence:
Dr. Marcin Dominia, Chair and Department of Cardiology, Medical University of Łódź, ul. Knaślewicza 1/5, 91–347 Łódź, Poland, e-mail: marcin.dominia@gmail.com

Conflict of interest: none declared

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