

Conclusions The combination of transthoracic and contrast echocardiography can diagnose and classify URCS accurately and may be the first choice or screening method in diagnosis.

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ECHOCARDIOGRAPHIC DIAGNOSIS OF UNROOFED CORONARY SINUS SYNDROME

Yang Yali Department of Ultrasound, Union Hospital, Tongji Medical College, Huazhong University

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Purpose Unroofed coronary sinus syndrome (URCS) is a rare cardiac abnormality in which a communication occurs between the coronary sinus and the left atrium as a result of the partial or complete absence of the roof of the coronary sinus. The diagnosis is often difficult because of non-specific clinical features. The aim of the study was to explore the value of transthoracic and contrast echocardiography in the diagnosis of URCS.

Methods The echocardiographic characteristics of 23 patients with URCS evaluated and operated at our hospital from October 1999 to June 2010 were analysed retrospectively and compared with surgery results.

Results Of these 23 patients, 60.9% were totally unroofed, 26.1% partially unroofed in mid-portion and 17.4% partially unroofed in terminal portion. Echocardiography diagnosed 13 cases correctly (56.5%). The 6 cases before 2002 were all missed or misdiagnosed. However, after 2002, 88% of the 17 cases were diagnosed correctly and the defects of 2 missed cases were totally unroofed without a persistent left superior vena cava (PLSVC). 13 cases were also evaluated by contrast echocardiography. 12 cases were diagnosed correctly, including 5 mid-portion (1 of them was mixed type) and 7 Raghbi's syndrome, and 1 terminal portion was misdiagnosed as Raghbi's syndrome. Raghbi's syndrome (type Ia) presented with complete invisibility of the coronary sinus and the PLSVC directly draining into the upper left corner of the left atrium, in some cases the connecting position was visualised clearly. The contrast echocardiography showed first effusion of the coronary sinus and then the bubbles shifting into the left atrium and the right atrium almost simultaneously in type- α a, while firstly effusion of the right atrium and then a few bubbles into the coronary sinus through the discontinuous wall on coughing or Valsalva maneuver in type α b. In cases with the terminal portion type, the orifice of the coronary sinus was found at the left atrium near the posteromedial commissure of the mitral valve and its position at the interatrial septum was detected. The contrast echocardiography showed the sequential effusion of the coronary sinus, the left atrium and the right atrium in 1 of type β a case.