

Original Research Article

Assessment of Congenital Coronary Artery Fistulas by Color Doppler Transthoracic Echocardiography: Report of 8 Cases

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Abstract: Coronary angiography is the ideal tool for imaging the coronary anomaly, but the relation of coronary artery fistulas to other structures, and their origin and course, may not be sensitive. We evaluated the ability of color Doppler transthoracic echocardiography in the assessment of patients with coronary artery fistulas. Eight patients with CT angiography confirmed coronary artery fistulas were investigated by transthoracic echocardiography at affiliated Hospital of Guilin Medical University between January 2009 and June 2015. The right coronary artery was involved in 4 cases and the left coronary artery in 4. Four fistulas drained to the right ventricle, 3 to the right atrium, and 1 to the left ventricle. Coronary artery aneurysms, aortic valve prolapse with severe aortic regurgitation were involved in one patient. The diagnosis of coronary fistulas can usually be made by echocardiography with color flow mapping and CT echocardiography. Color Doppler echocardiography is useful to visualize shunt flows originating from the opening or in the precise localization of coronary artery fistulas.

Keywords: Coronary anomaly; congenital heart disease; Echocardiography

INTRODUCTION

Coronary artery fistula is a rare congenital anomaly in which the involved coronary artery drainage into a cardiac chamber, pulmonary artery, or other structure. Elective closure has been recommended after some reports that patients with coronary artery fistula may develop complications such as congestive heart failure, myocardial ischemia, endocarditis or aneurysmal dilation during or after the second decade of life [1].

Coronary angiography is the gold standard for diagnosing coronary artery fistulas, but sometimes the exact proximal course of anomalous coronary arteries may be difficult to detect [2]. Coronary artery fistula can also be diagnosed by finding a dilated coronary artery by echocardiography, and its drainage can be detected by color flow mapping. We summarize our experience of coronary artery fistulas in 8 patients whose condition was prospectively diagnosed by color Doppler transthoracic echocardiography [3].

PATIENTS AND METHODS

Between January 2009 and June 2015, 8 patients with coronary artery fistula underwent surgery repair at affiliated hospital of Guilin Medical University. The study protocol was approved by the Institutional Review Board. Patient consent was not required because of the retrospective nature of the study.

Two-dimensional transthoracic echocardiography was performed to visualize a dilated coronary artery. Doppler color flow imaging was performed in all patients from the standard parasternal and apical approaches and nonstandard views when necessary. The luminal diameter of the main coronary arteries by two-dimensional echocardiography was assessed as the distance between two walls of the coronary arteries (brightest points of the two parallel echoes) just distal to the ostia. We tried to determine whether a mosaic-colored flow or turquoise which was suggestive of

disturbed flow) was present in any of the four cardiac chambers. When turbulent flow was visualized, conventional single-gated pulsed Doppler echocardiography was performed guided by Doppler color flow imaging to verify the characteristics of flow velocities on spectral and audio output. For examining the timing of flow, M-mode Doppler color imaging was also performed.

The diagnosis of coronary artery fistula was made on the basis of clinical, surgical and pathological criteria. Of these, 4 were men, 4 were women, and

mean age was 17 years. The right coronary artery was involved in 4 cases and the left coronary artery in 4. Four fistulas drained to the right ventricle, 3 to the right atrium (figure 1), and 1 to the left ventricle. Coronary artery aneurysm, aortic valve prolapse with severe aortic regurgitation was involved in one patient. A coronary artery fistula ligation procedure was done in 7 cases, a coronary artery fistula ligation + aortic valve replacement + coronary artery aneurysm resection in one case. The clinical data were collected from hospital records. Summary of patient's data see Table 1.

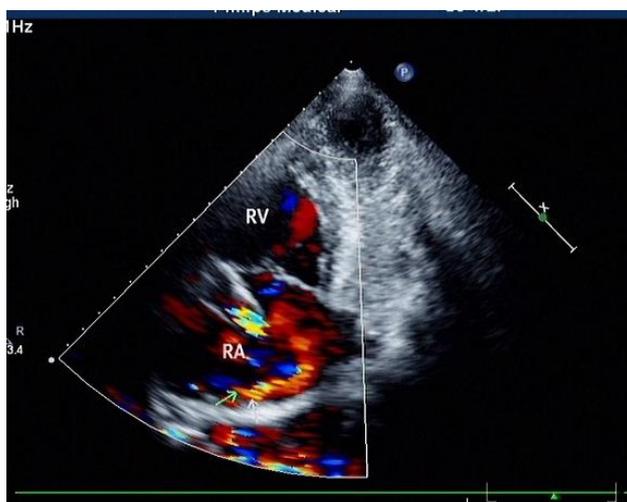


Fig 1: A fistula originating from the right coronary artery and terminating into the right atrium (arrows).

Table 1: Summary of patient's data

Case No.	Age	Gender	Origin of CAF	Drainage site	Coronary artery disease	Combined with cardiac malformations	Surgical procedure
1	3	male	right coronary artery	right atrium			ligation
2	10	female	left circumflex	right ventricle			ligation
3	41	male	right coronary artery	right atrium			ligation
4	54	male	right coronary artery	left ventricle	coronary artery aneurysm	aortic valve prolapse right coronary artery	Aortic valve replacement and coronary artery aneurysm resection
5	0.8	female	left circumflex	right ventricle			ligation
6	2	male	right coronary artery	right atrium			ligation
7	19	female	left coronary artery	right ventricle			ligation
8	6	female	left circumflex	right ventricle			ligation

RESULTS

All patients with coronary artery fistula had ligation or excision with the use of cardiopulmonary bypass. There were no intraoperative or postoperative complications. They were discharged home within 9 days after surgery. All patients are followed in the outpatient clinics no complaints and no recurrence of symptoms for more than 20 months. The first patient has completed to 7 years of follow up.

DISCUSSION

Congenital coronary fistula is a relatedly rare coronary anomaly. Coronary artery fistulas are present in 0.002% of the general population and are documented in nearly 0.25% of the patients undergoing coronary angiography [4]. Unless very large and hemodynamically significant, coronary artery fistulas are usually asymptomatic in younger patients. Coronary artery fistulas are incidental findings during physical examination or angiographic evaluation for coronary diseases. With increasing age, symptoms begin to appear as intracardiac shunts, endocarditis, myocardial infarction, coronary aneurysm and even sudden death [1].

Coronary arterial fistulas most originate from the right coronary artery and commonly open into one of the right heart chambers. It may occur isolated or along with other congenital heart diseases. Clinical presentation is dependent on the magnitude of the coronary artery fistula connection. If a large fistula is present, coronary artery fistula could result in a significant left-to-right shunt with congestive heart failure and cardiomegaly in young age [5]. Large, hemodynamically fistulas should be closed electively at the time of diagnosis. The differential diagnosis includes persistent ductus arteriosus, ruptured sinus of Valsalva aneurysm, aortopulmonary window, pulmonary arteriovenous fistula, prolapse of the right aortic cusp with a supracristal ventricular septal defect, and systemic arteriovenous fistula [6]. Imaging techniques, all of which have limitations, are utilized for diagnosis and for planning before surgical or percutaneous interventions if closure of the coronary fistula is indicated.

Coronary angiography is the gold standard for detection and visualization of coronary artery fistulas because of its accuracy in defining the artery of origin, as well as the recipient vascular structure [7]. However, the drainage site and course of the coronary artery fistula are visualized less clearly by coronary angiography because of its two-dimensional imaging pattern [2]. Kacmaz and his colleagues recommended that CT angiography may serve as an alternative non-

invasive tool to visualise coronary artery anomalies, especially coronary artery fistulas coursing between two vessels, to detect origin, course, and the drainage site of coronary artery fistula through its excellent spatial resolution and ability to show relationship of anatomic structures [8].

Transthoracic echocardiography can display a dilated coronary artery, where the abnormality artery arises, and even the fistula itself, including the drainage into a chamber or vessel, in addition to a continuous turbulent systolic and diastolic flow [9]. Due to better echocardiographic windows in this young age group [10], transthoracic echocardiography is a primary diagnostic tool in most patients under 20 years of age, but not ideal for the patients older than 20 years old. Intraoperative transesophageal echocardiography with colour flow Doppler was used for precise location of the coronary artery fistula communication, selective demonstration of vessels feeding the coronary artery fistulas and documentation of abolition of blood flow without the need for cardiopulmonary bypass. Furthermore the effect of shunt occlusion on regional wall motion was also documented which the successful ligation of the fistula [11]. Furthermore, different perioperative imaging may be required. Intraoperative ECG monitoring is a very useful tool to guide and detect perioperative ischemic changes. TEE is also helpful to demonstrate wall motion abnormalities and confirm the complete ligation of the fistula [6]. Transesophageal echocardiography allows imaging of the proximal coronary arteries, measurement of coronary flow reserve, identification of coronary artery anomalies, and observation of wall motion during cardiac surgery. Because of the better resolution of higher frequency transducers and the proximity of the coronary arteries to the esophageal window, transesophageal echocardiography is emerging as a valuable method for evaluation of coronary artery disease. The application of transesophageal echocardiography in evaluation of coronary artery disease will continue to grow as technology improves [12].

Treatment includes transcatheter or surgical closure, which can provide the best results early in the early course of the disease [13]. Very small coronary artery fistulas in asymptomatic patients should be monitored because fistulas tend to grow with age. Transthoracic echocardiography should be performed annually during the follow-up in these patients and decreasing the frequency in case of asymptomatic stable condition [14]. Nevertheless the growing potential of non-invasive techniques like transthoracic and

transesophageal echocardiography or cardiac imaging with magnetic resonance or computed tomography for identification and follow-up, transthoracic echocardiography is the best choice during the follow-up.

Having evaluated 8 patients with coronary artery fistulas, the current numbers are too low to determine the exact sensitivity of color Doppler transthoracic echocardiography compared with coronary CT angiography. But, color Doppler transthoracic echocardiography is still useful to visualize shunt flows originating from the opening or exit of a coronary artery fistula.

AUTHOR'S CONTRIBUTION

Haiyong Wang and Fang Lei wrote the paper. Yujing Yang, Feng Lin, Xianzhu Liang, Xingxing Peng, Fugui Ruan, Jiangbin Sun, Jianfei Song and Zhenzong Du supervised the composition of the paper. All authors read and approved the final paper.

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