An asymptomatic right coronary ectasia due to a likely congenital fistula to the right cardiac chambers: an incidental finding detected in the course of investigation for the eligibility to serve the Army

The relevant role of the routine use of echocardiography combined with other diagnostic imaging methods

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Riassunto -Coronary artery fistulas are a rare congenital anomaly of termination of the coronary arterial tree due to an abnormal communication between one or more coronary arteries and great vessels or a cardiac chamber. The incidence of coronary artery fistulas is low but with steady increase detection with the broad application of Trans Thoracic Echocardiography. The correct diagnosis requires a multi-diagnostic approach: conventional coronary angiography, multi-slice detector computed tomography, Cardiac Magnetic Resonance. The treatment strategy of coronary artery fistulas is still a controversial issue. A likely coronary artery fistulas between the right coronary artery and the right chambers of the heart in a 22-year-old male examined at the Military Clinical Centre in Milan before being admitted as a recruit in the Italian Army is reported. Trans Thoracic Echocardiography is a recommended diagnostic tool in the regular investigation and follow up of the applicants to serve the Army.

Key words: congenital cardiac anomalies, coronary artery fistula, coronary –ventricular fistula, Doppler echocardiography, transthoracic echocardiography, cardiovascular screening.

Key messages:

- The diagnosis of congenital heart disease not detected until adulthood has decreased over the past several decades.
- Silent coronary artery anomalies can be considered uncommon cases of congenital cardiac anomalies in the ages over the adolescence.
- The very early diagnosis of silent coronary artery anomalies is desirable before they determine serious symptoms or complications mainly for those over 20 years of age.
- Trans thoracic echocardiography is considered a routinely diagnostic tool to suspect the presence of silent congenital cardiac anomalies in the applicants to serve the armed forces.

Introduction

A coronary artery fistula (CAF) or coronary arteriovenous fistula (CAVF) represents an anomalous communication between a coronary artery and any of the 4 chambers of the heart or any of the great vessels (superior vena cava, pulmonary artery, pulmonary veins, or coronary sinus). These connections are most often congenital (1). CAVFs were first reported by Krause (2) in 1865. Subsequent early reports by Cayla (1885), Abbott (1906), Trevor (1911), Blakeway (1918), Halpert (1930), and Harris

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(1937) are mentioned in literature (3). A CAVF of congenital origin is determined by the persistence of sinusoidal connections with the lumens of the primitive tubular heart that provide myocardial blood flow in the early embryologic period (4). In the primitive heart the coronary veins represented by the endothelial outgrowths initially forming the trabecular spaces of the myocardium terminate on the epicardial surface in the capillary network. In the meantime the capillary network is joined on the other side by the embryonic coronary arteries arising from the endothelial growth in the base of the aorta. Usually the previously mentioned intramyocardial sinusoids get thinner and consequently remain only as thebesian vessels in the adult. When the concealment of the intramyocardial trabecular sinusoids is inadequate a fistulous communication perseveres between the coronary arteries and a cardiac chamber (5). Similarly coronary-to-pulmonary artery collaterals are thought to be remnants of the splanchnic vascular system and commonly revert with the development of the pulmonary arterial system occurring later in fetal life (6). However, they may remain in case of incomplete development of the central pulmonary arterial system. Nonetheless CAVFs can occur as a result of trauma, infection, or iatrogenic injury (e.g. intracardiac congenital heart operations, transcutaneous catheter techniques for myocardial biopsy, coronary angioplasty, a complication of Kawasaki disease) (7).

Anatomic features

CAFs are recognized as a major coronary anomaly by Ogden's classification. CAF is a finding occurring in 0.002% of the general population and in 0.4% of all cardiac malformations (2,4). Fistulas originating from the right coronary artery predominate as they account for 50% up to 60% of all cases, while those ones originating from the left anterior descending artery 25% to 42%, from the circumflex artery about 18%, from the diagonal branch about 2%, and from the left main coronary artery or circumflex-marginal branch less than 1% (8). Single fistulas are most frequent, ranging from 74% to 90% (9,10), multiple fistulas comprise from 10.7% to 16% of all CAFs (9,10,11), and both coronary arteries are affected in 5% (2,6,11). CAFs drain more commonly into the right heart structures than into the left ones (19). In surgical findings, the drainage site is located into the pulmonary artery in 15% to 43% of cases, the right ventricle in 14% to 40%, the right atrium in 19% to 26%, the left ventricle in 2% to 19%, the coronary sinus in 7%, the superior vena cava in 1%, and the left atrium in 5% to 6% (2,8). Bilateral fistulas drain more often into the pulmonary artery (56%) than unilateral fistulas (17%) (12,13).

Cameral fistulas (coronary fistulas draining into the cardiac chambers; CCF) are uncommon congenital vascular anomalies reported in nearly 0.08% to 0.3% of non selected patients undergoing diagnostic coronary angiography (14,15). Cameral fistulas have been termed as arterioluminal, when a direct and focal communication with the cardiac chamber is demonstrated or arteriosinusoidal, when an arterial blood communication with the cardiac chambers is due to a sinusoidal network. An arteriocapillary variant has also been described (16). Cameral fistulas communicate in 90% of all cases with the right-sided chambers of the heart, while the rest drain to the left side of the heart, or to both (17). Isolated CAVF may be found in 55% to 80% of cases (18) or the associated anomalies with other congenital heart disease in 20% to 45% (tetralogy of Fallot, atrial septal defect, patent ductus arteriosus, ventricular septal defect, pulmonary atresia with intact ventricular septum, and superimposed coronary artery disease) (9).

Pathophysiology

The resistance of the fistulous link due to both its size, tortuosity, length and the site of termination is the main determinant of the physiologic derangement. As in a patent ductus arteriosus, flow from the coronary artery to a venous arrangement or right-sided cardiac chamber happens throughout the cardiac cycle. Blood deflects into the lower-resistance pathway through the fistula rather than passing through the smaller arterioles and capillaries of the myocardium. A "diastolic runoff" occurs with larger fistulas, unsheathing blood from the normal coronary route with an increased pulse pressure and a coronary steal. A left-to-right shunt is determined if the fistula empties into the systemic venous side of the circulation. Such a shunt volume loads both ventricles dissimilarly to the left-to-right shunt of atrial septal defect (right ventricular volume load) or ventricular septal defect/patent ductus arteriosus (left ventricular volume load). When the left atrium or pulmonary vein is the drainage site, a volume overloading the left heart only is the result of an effective left-to-left shunt. When the fistula penetrates a left-sided chamber, it determines a run-off from the aorta resembling aortic valve regurgitation (1).

Natural history, clinical presentation and complications

As most of the CAFs are so small that myocardial blood flow is not impaired patients are usually asymptomatic. 80% of patients aged younger than 20 years manifest no symptoms compared with only 40% of those ones aged older than 20 years (6). However small CAFs in children tend to expand with age and if untreated cause clinical symptoms in 19% of patients aged younger than 20 years and in 63% of older patients (18). In such conditions, early surgical correction is necessary due to the high prevalence of late symptoms and complications, particularly when the shunt between the left and right sections of the heart is relevant (Qp-to-Qs ratio >1.5) (19). Very uncommon (1% to 2% of cases) is the spontaneous closure of the fistula because of spontaneous thrombosis (20). Fatigue, exertional dyspnea are the most common clinical symptoms but even angina, and congestive heart failure are showed (6).

Complications of coronary fistula comprise:

1. Coronary complications— The increased flow is the cause of the enlargement of the coronary artery branches proximal to the shunt site. Symptoms and effects include chronic myocardial ischemia, angina, myocardial infarction, congestive heart failure and cardiomyopathy. Coronary steal due to diversion of blood produces a commonly expected myocardial ischemia (21,23). Massive dilatation and aneurysm formation is due to persistent high flow in coronaries (6, 24). Premature coronary atherosclerosis is also described (21);
2. Overloading of the cardiac chambers—wherever the site of fistula drainage is located in the left sections of the heart, CAFs determine an increased left ventricular end-diastolic pressure, left ventricular hypertrophy, and in older patients, congestive cardiac failure (6, 21). Congestive heart failure is prevalent in patients with a fistula to the coronary sinus (22). Atrial fibrillation and ventricular tachyarrhythmias have been reported in patients with CAVF (5, 21); the onset of atrial fibrillation is more frequent in older patients as a result of right atrial dilation from a fistula to the right atrium;
3. Valvular and endocardial complications—Functional valvular regurgitation due to papillary muscle dysfunction has been reported (23). The incidence of infective endocarditis ranges from 0% to 12% (6, 22–25) so that endocarditis prophylaxis is recommended as long as the flow continues across the fistula;
4. Extracardiac complications — Rupture of an associated aneurysm can result in hemopericardium (6). Pulmonary hypertension is elicited in case of a considerable left -to-right shunt, although it has been rarely reported (6, 26).

On examination, a continuous murmur may be hearable at the left lower sternal border (22). Cardiomegaly and ventricular hypertrophy may be manifest and a collapsing pulse may be uncovered with large fistulas to the left-sided heart chambers.

Diagnosis

In about 50% of the surgical patients the electrocardiogram (ECG) is normal while in case of volume overload it may show right or left ventricular hypertrophy and in older patients with fistulas to the right atrium atrial fibrillation may be present. An ischemic pattern is probable when the coronary steal involves a major branch of the left circumflex artery. Cardiomegaly and pulmonary plethora may be present in the chest X-ray. Two-dimensional transthoracic echocardiography or transesophageal echocardiography (TEE) may determine the diagnosis, displaying the origin and drainage site, or provide signs such as coronary dilation or chamber enlargement. A markedly expanded coronary artery can usually be recognized with both transthoracic echocardiography and TEE. Catheterization and coronary angiography are still generally necessary for a definitive diagnosis and planning management. Magnetic resonance imaging (MRI) and computed tomography (CT) cardiac coronary angiography are helpful, noninvasive, and reliable imaging techniques for the investigation of major coronary artery anomalies (27).

Case report

A 22-year-old male came to the Military Clinical Centre in Milan from the Army Centre of Recruitment in Milan for the cardiological examination before being admitted as a recruit in the Italian Army. All of the common cardiovascular risk factors were not manifested. He usually had no symptoms. Cardiac auscultation could hardly hear any murmur. He did not have a wide pulse pressure (blood presure 120/80 mmHg). His lungs were clear and there was no elevated jugular venous pressure. The rest physical examination was unremarkable. Electrocardiogram (ECG) showed normal sinus rhythm without significant ST-T wave changes and the

mean electrical axis was normally oriented (***Fig. 1***). Trans-Toracic Echocardiography (TTE) showed a noticeable pulsatile localized diastolic high turbulent speed blood flow on the lateral wall of the right heart close to the tricuspid valve annulus. The abnormal blood flow pointed from the atrioventricular groove toward the lateral wall of the right chambers in all the four chambers views (apical and subxiphoid positions); (***Figg. 2; 3; 4***). Reverse tracing of the abnormal blood flow was not possible so that its source was not immediately identifiable. In order to further characterize this vascular structure and to define its origin, course and draining site, as well as its relation with the coronary arteries further imaging investigation was followed with cardiac MRI (CMR). Atrial and right ventricular dimensions were enlarged while function as well as thickness of the right ventricular walls were within normality; a mild dilatation of the left ventricle with ejection fraction around 68% was registered. The left coronary artery (LCA) and its branches were normal in origin, course, and caliber. A right coronary normally originating from the right sinus of Valsalva with a serpiginous course and a dilatated diameter up to 19mm was identified. Its route was characterized by a proximal deflection anteriorly to the aortic root and a distal one located posteriorly at the level of the atrioventricular groove after the origin of the posterior interventricular artery. The measures of the blood flow across the pulmonary (net flow 134 ml/beat; Qp) and aortic valves (net flow 105 ml/beat; Qs) during the phase contrast velocity mapping of volume together with the calculation of the stroke volumes from both the right (135 ml) and left (147 ml) ventricles during the cine-balanced steady-state free precession sequences were also determined. The difference between the estimated Qp/Qs ratio (1.2) and the value of stroke volumes determined for both the ventricles was interpreted as a meaningful shunt between the left and

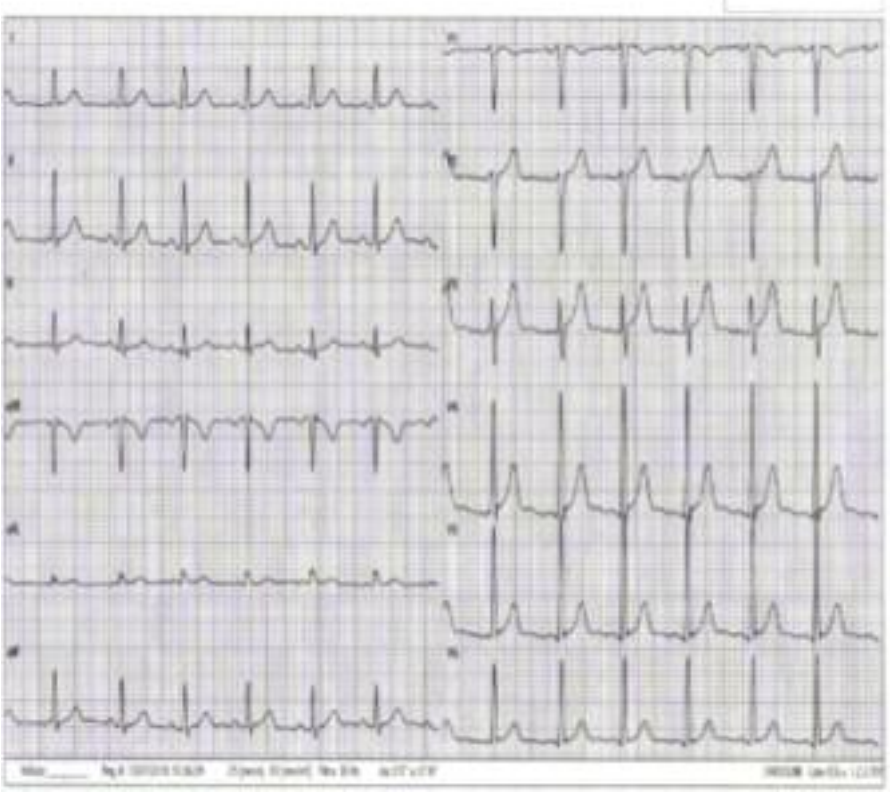

Fig. 1 - Electrocardiogram at rest.

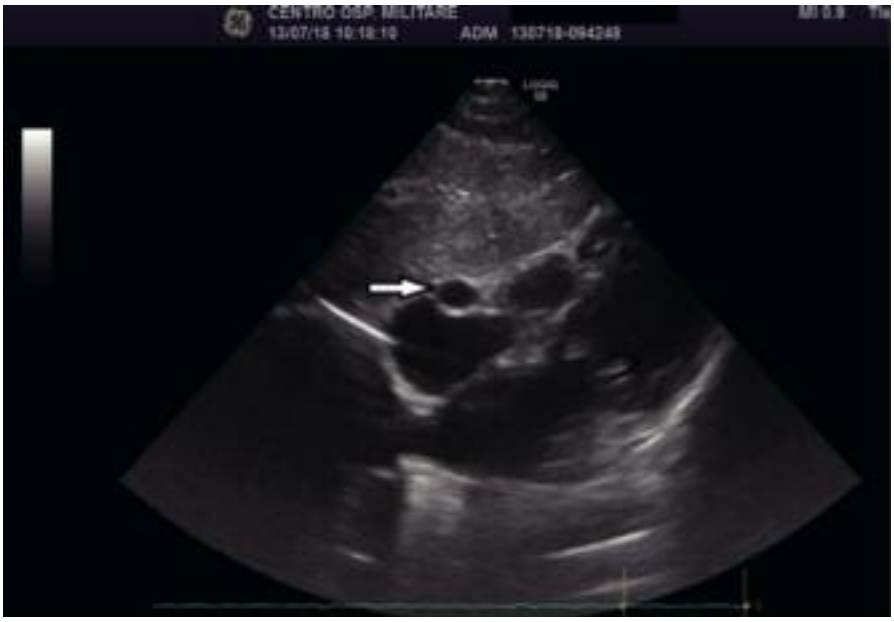

Fig. 2 - TTE showing a circular area proximal to the right lateral atrioventricular groove in the subxiphoid view (arrow).

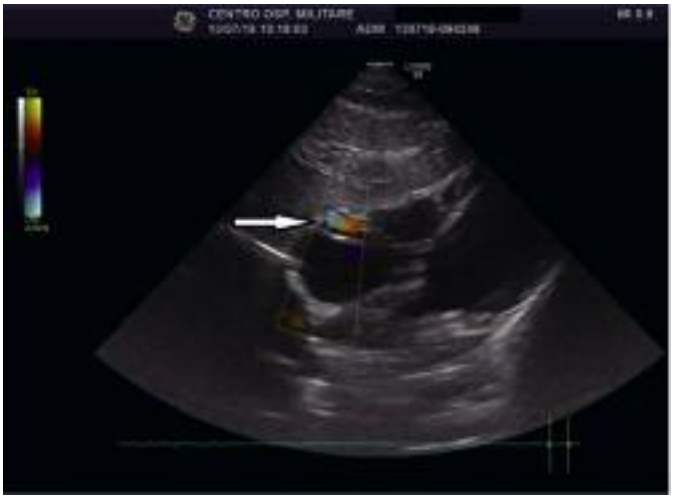

Fig. 3 - A continuous flow at color Doppler analysis in the same view of Fig. 2 (arrow).

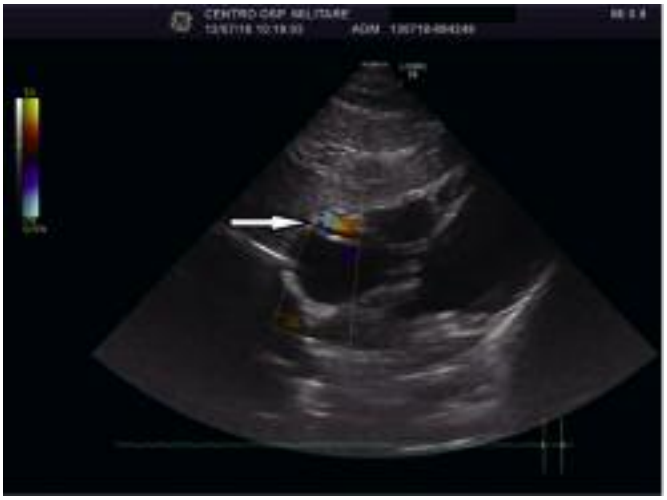

Fig. 4 - A continuous flow at color Doppler analysis of the area proximal to the right lateral atrioventricular groove in the apical four chambers view (arrow).

the right chambers of the heart. No perfusion defect was recognized during first pass perfusion sequence and the Late Gadolinium Enhancement (LGE) sequences did not show any proof of prior myocardial infarction or myocardial fibrosis of the LV wall. Considered the above mentioned anatomic features of the right coronary artery and the analysis of the hemodynamic parameters the whole imaging examination after CMR appeared highly suggestive for a CCF draining to the right cardiac chambers (***Fig. 5***).

Conclusions

It has become increasingly rare to make the diagnosis of congenital heart disease "de novo" in adult populations. Because of the raised access to well-trained pediatricians, family practitioners and cardiologists most of the "significant" congenital heart lesions are identified in childhood and adolescence. Thus, the number and nature of lesions not detected until adulthood has decreased over the past several decades. The single most common congenital lesion to be revealed in adulthood, other than the bicuspid aortic valve, is the atrial septal defect. There are infrequent cases of ventricular septal defects and other anomalies that escape detection to adulthood (28). On the basis of the above described case report the diagnosis of silent coronary artery anomalies can be reasonably comprised among the uncommon cases of congenital cardiac anomalies in the ages over the adolescence. In fact the very early diagnosis in all patients with a CCF is desirable before they develop serious symptoms or complications, mainly for those over 20 years of age. Current treatment options are represented by conservation treat-

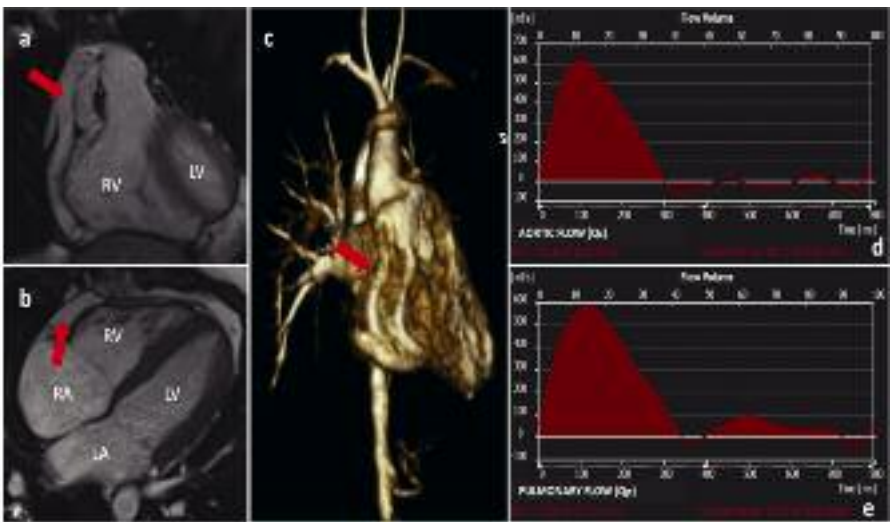

Fig. 5 - Cine images (a, b) and 3D reconstruction from angiographic images (c) showing a markedly dilated right coronary artery with tortuous course. Measurement of systemic and pulmonary flow from phase contrast images (d, e) demonstrates the presence of left to right shunt (Qp/Qs ratio 1.25) (Courtesy of Antonia Camporeale, MD, PhD,; Multimodality Cardiac Imaging Section I.R.C.C.S., Policlinico San Donato-Milan-Italy).

ment, surgery and transcatheter closure. Moreover the management of CAF is still a controversial matter, particularly in asymptomatic patients (6).

As with any diagnostic method, echocardiography has definite advantages and limitations. It is widely recognized that cardiac ultrasonography itself carries no risk to the patient, operator and that modern two-dimensional echocardiographic techniques are capable of visualizing all four cardiac chambers, cardiac valves and the great vessels. Moreover they provide immediate high-resolution tomographic views in unlimited planes, which is a comprehensive means for evaluating virtually all forms of anatomical and in particular of congenital cardiovascular disease found in both children and adults. The American College of Cardiology, American Heart Association, and American Society of Echocardiography have issued recommendations for appropriate training in echocardiography and likewise recommendations on its appropriate use (29). Moreover when further diagnostic evaluation is necessary to both better define the aetiology of a suspect congenital vascular lesion and sequentially to establish in as much as possible exhaustive way if an applicant is fit to serve the Army we need to recur to a second-level techniques of image investigation. As in our case CMR may provide supplemental information regarding pulmonary artery anatomy, complex venous and great artery connections. Due to the multiple capabilities for the evaluation of congenital heart disease CMR provide a morphological information by ECG-gated spin-echo and cine-sequences (e.g. ventricular volumes, mass and function). Moreover CMR allows both to visualize the major coronary arteries using the MR angiographic techniques and to assess the presence of intracardiac shunts.

In conclusion the number of clinical applications for which Color-Doppler echocardiography (CDE) can, or could, be used is substantial. We must acknowledge that in the Military Clinical Centre in Milan, the use of CDE has been routine. We wish the technique will become more widespread over the next few years because according to our experience it is very often regular part of the cardiac examination of all the applicants to serve the Army in order to achieve an early diagnose in those ones with never ascertained or asymptomatic valvular and/or congenital heart disease. Other potential applications, though only speculative at present, may well be realized as future technologic improvements occur. Thus, the future of CDE appears meaningful not only in the clinical field but also in the legal medicine one in regard to the access to the regular service in the Army.

Disclosures:

The Author declares that he has no relationships relevant to the contents of this paper to Disclose.

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