Prevalence of the Anomalous Aortic Origin of a Left Coronary Artery from the Right Coronary Sinus and Posterior Course: detection by Transthoracic Echocardiography in an Unselected Ambulatory Population

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Introduction
Coronary artery anomalies represent a group of relatively rare congenital malformations, that also include the anomalous aortic origin of the coronary arteries (AAOCA). Based upon large angiographic series the reported prevalence of AAOCA ranges from 0.45% to 1.3% in the normal population [1,2] while the incidence of all coronary anomalies appears to be much higher [3].
The majority of AAOCA are believed to have no clinical relevance but some have been related to sudden death, particularly during effort or sports activity [4-6].

Once suspected, coronary anomalies can be readily recognized by invasive and/or non-invasive means. At present usually anomalous vessels of the coronary circulation are detected incidentally, in the diagnostic work up of other diseases, or on the occasion of an invasive coronary angiography or CTCA performed for other purposes.

Transthoracic echocardiography (TTE) has been used to explore coronary anatomy [7-9] and found to be a valuable tool mainly for identifying coronary anomalies in pediatric patients [10]. Transesophageal echocardiography has been shown to accurately assess coronary anatomy in adults with an image quality suitable to detect and evaluate the anomalous course and/or origin of a coronary vessel [11-12]. In the past few years both Computed Tomography of the Coronary Arteries (CTCA) and Magnetic Resonance Coronary Angiography have gained a pivotal role for the identification and depiction of such malformations [13-14] and at present are considered the gold standard for the diagnostic evaluation of suspected coronary anomalies [15-16].

The main challenge in the evaluation of coronary anomalies is the identification of potential carriers of malignant variants, particularly in adult life. Usually, once an anomaly is suspected it is subsequently fully characterized by either invasive coronary angiography or by the newly available imaging modalities.

In this investigation we imaged prospectively by TTE an unselected population of ambulatory patients. The aim of this study was to evaluate the specificity and prevalence of this TTE pattern that we found associated to a left coronary artery vessel originating from the right coronary sinus (RCS) running posteriorly to the aortic root.

Methods
Population
We prospectively evaluated 3692 ambulatory patients (2146 males and 1546 females, mean age 64+9 yrs) by TTE with the aim of detecting a peculiar aspect that was incidentally found in one patient in our laboratory. This TTE pattern was found to be associated with the anomalous origin of a left circumflex artery (LCX) from the RCS, running posterior to the aortic root (Fig. 1-2). Following this original case we screened for this pattern all the subjects requiring a TTE examination from January 2013 to June 2015.

Indications for TTE performed at our Center included several common indications to a routine ambulatory examination (i.e. suspected heart failure, arterial hypertension, heart murmurs, hypertrophic and dilatative cardiomyopathy, coronary artery disease, post-myocardial infarction, mitral and aortic valve disease). Patients who were known to have a coronary anomaly were not included in the study. Patients showing an abnormal pattern suggestive for the AAOCA were asked to perform a CCTA to either confirm or better depict the coronary anatomy.

Informed consent for the imaging protocol, CCTA, and use of their clinical information was obtained from each patient. The study protocol conforms to the ethical guidelines of the 1975 Declaration of Helsinki as reflected in a priori approval by the institution’s human research committee

Transthoracic echocardiography
TTE was achieved by using either an Artida (Toshiba Medical System), or a Vivid 7 echo platforms (GE, Milwaukee, US). In all the cases a phase array ultrasound probe was used, usually operating at 3.5 MHz. Patients were imaged by using a standard
multiplane approach including a parasternal long axis view, a 2- and 4- chamber apical view and multiple short axis from the base to the apex of the left ventricle.

A modified 4-chamber apical view was added to the standard examination for the purpose of this investigation. Once the mitral annulus was identified, the imaging plane was shifted cranially in order to explore the area immediately below and behind the aortic root and above and behind the mitral leaflets. The identified pattern is a double rimmed structure with hyperechogenic borders perpendicular to the imaging plane and crossing the heart slightly above the level of the mitral ring (Fig. 3-5).

When the abnormal feature suggestive for the abnormal course of the coronary artery could be detected, the probe was moved in order to follow this vessel. The evaluation of patients was performed by experienced cardiologists with several years of activity in cardiovascular imaging. All the examinations belonging to the suspected cases were recorded and stored onto the cardiology PACS for subsequent review and analysis. All the patients exhibiting the described pattern were asked and accepted to perform a CTCA (Fig 2-3,6).

Coronary Computed Tomography of the Coronary Arteries
CTCA was performed by using a 64-slice scanner (GE Healthcare, Lightspeed) and a dose reduction protocol that included a “step-and-shoot” prospective scanning and a low voltage energy exposure (usually 80 mA). Patients received I.V. beta blockers to lower their heart rate to 50-55 bpm and were instructed and prepared to maintain an expiratory apnea during the iodinated contrast infusion and x-ray exposure. With such a protocol the radiation dose can be decreased by 80% compared to a standard helical examination and the effective dose ranged in this population from 1 to 4.5 mSv.

Results

Incidence of the abnormal pattern
In 3298 patients out of the initial 3692 subjects examined by two dimensional echocardiography (89.2% of TTEs) we obtained a good quality apical 4-chamber view thus allowing the search for the abnormal pattern above described. The anomalous aspect was identified in 16 cases out of the 3298 screened thus resulting in a prevalence of 0.48% of the whole group evaluated. In all these subjects the abnormality was very easily seen and documented.

Two dimensional echocardiographic features
The abnormal feature resembled a double track crossing the heart perpendicular to the probe. It had some distinctive features: it appeared as a double rimmed echo-free structure located just below and behind the aortic root above the mitral ring, had a route perpendicular to the scanning plane, it was a several centimetres long (usually 3-4cm), and it was neither attributable to the mitral valve nor to the coronary sinus. The initial aspect resembled a ghost artefact parallel to the chest wall, but once correctly identified it disclosed its vascular nature (Fig 1-5, see also the supplementary material).

Coronary anomalies
In all the patients the abnormal pattern was associated and could be attributed to an AAOCA with posterior course. By using CCTA
15 out of the 16 patients had a LCX originating from the RCS from a separate orifice. Once originated from the RCS the LCX showed a retroaortic course running behind and below the aortic root and finally reaching the left atrio-ventricular groove. In the remaining case the anomalous vessel was the left main coronary artery. In this patient (Pt N° 6) the left main coronary artery took origin from the right coronary sinus from a separate orifice and, following a retroartic course, reached its normal position later bifurcating in the left descending and LCX.

Clinical correlations
A thorough clinical history from the 16 patients did not provide any relevant information (Table I). One patient had a significant mitral regurgitation (Pt N° 6) and one had left ventricular dysfunction (Pt N°16). One patient had recovered from an uncomplicated myocardial infarction (Pt N° 11). In no case we could associate the individual clinical history or findings with the AAOCA observed. A few patients reported to suffer from atypical chest pain. All the patients were offered a test of ischemia. No patient showed objective signs of myocardial ischemia during stress ECG, nuclear stress test, or dobutamine echo stress test. The majority of patients were asymptomatic and the AAOCA was an incidental finding in all of them.

Discussion
We identified a novel echocardiographic sign able to detect an AAOCA in adult subjects undergoing a routine TTE. This novel sign, represents the echocardiographic aspect of the anomalous course of a left coronary vessel (either LCX or left main coronary artery) arising from the right coronary sinus and running behind the aortic root to reach its natural position. We identified 16 patients over a large series of otherwise normal subjects. In all the cases we confirmed the presence of an AAOCA by using CTCA. The observed echocardiographic pattern resulted to be pathognomonic for the anomalous course of a left coronary vessel arising from the RCS.

The underlying anomaly detected in the majority of patients should probably be considered a benign condition, with the only exception of one patient whose abnormal course was attributed to the left main coronary artery originating from the RCS. This latter AAOCA may not be harmless although the posterior course of AAOCA is usually benign and less dangerous than the interarterial course. Therefore, the herewith described novel sign identifies a left coronary vessel originating from the RCS without offering the ability to distinguish between the left main or the LCX.

Some Authors have used TTE to screen patients and athletes for AAOCA. Davis et Al. screened 2388 pediatric patients by using TEE and found evidence of AAOCA in 4 (0.17%) [10]. Also, Zeppilli used TTE to prospectively detect coronary anomalies in a large population of athletes [17]. In that study an AAOCA was identified only in 3 pts (0.09%) and the anomalous course was interarterial in all. By using TTE Labombarda ed Al [18]. have recently prospectively studied for major coronary anomalies 3502 subects, the majority of whom (84%) adults. They found 14 coronary anomalies (0.39% of the whole group). In this study the majority of the anomalies were detected in young or very young subjects. However, none of these studies described a posterior course of the AAOCA identified by TTE. Therefore the present study represents an original description never reported before.

With respect to the prevalence of the retroaortic course of the LCX originating from RCS there is evidence that this is the most common AAOCA in the adult population undergoing routine coronary angiography [19-20]. In such a population the incidence of AAOCA can be as high as 0.29% with a prevalence of the left
circumflex artery originating from the RCS of more than 50% of all the AAOCA (range 0.17-0.67%). Our data show a prevalence of 0.48% that is substantially similar to that expected based on previous data.

Our findings may have clinical relevance. Coronary angiography usually detects coronary anomalies but the assessment of both their origin and course may sometimes be difficult and the reported misdiagnosis can be as high as in 50% of patients [19]. TTE detection of the AAOCA may thus add significant information to a suspected angiographic finding possibly avoiding further imaging with CTCA or CMR. Also, the echocardiographic detection of a suspected AAOCA of the left coronary artery arising from the RCS may also prompt attention in candidates for cardiac surgery, percutaneous coronary interventions, or candidates for interatrial closure devices. In these patients a correct description of an AAOCA may avoid unnecessary long x-ray exposures during the interventions or avoid iatrogenic complications.

The LCX with a retroartic course is generally considered a benign anomaly. Unfortunately the observed TTE pattern although specific for an AAOCA cannot discriminate between the type of vessel involved. Our description about the possibility that the same TTE pattern may be present in only a minority of coronary anomalies. In this case the echocardiographic sign here described may represent a rarity rather than the rule. Further studies, on a larger population and of prospective nature may eventually clarify this aspect.

Although TTE may be inadequate to depict precisely the coronary anatomy or the extent of atherosclerosis the identification of even a small proportion of AAOCA may be relevant in clinical practice. Our experience indicates that a small supplement of time dedicated to the study of the coronary anatomy and the search for an abnormal pattern suggestive for a retroartic course of an AAOCA may help to identify a previously not detected coronary anomaly and that this anomaly may be clinically relevant. The need for a structured imaging protocol for detecting and describing the coronary anomalies, particularly in young subjects, has recently been discussed and there is evidence that this represents a major issue in this clinical field [22].

**Study Limitations**
The real incidence of the coronary anomalies has not been systematically assessed in the general as well as in our population. Also, we cannot establish whether the absence of the observed TTE pattern does exclude a coronary anomaly of the LCX or left main coronary artery. Technical and anatomical factors may indeed limit the detection of the observed sign. Although easily distinguished from other anatomical structures and artefacts the described TTE pattern may be present in only a minority of coronary anomalies. In this case the echocardiographic sign here described may represent a rarity rather than the rule. Further studies, on a larger population and of prospective nature may eventually clarify this aspect.

**Conclusions**
In conclusion, we identified for the first time a very specific sign for a specific group of AAOCA in adults who perform routine TTE. The described pattern is pathognomonic for a left coronary vessel originating the the RCS and with a posterior (retroartic) course. The most common anomaly associated with the observed pattern was the origin of the LCX from the RCS. However, the very same aspect was observed in a case of left main coronary artery originating from the RCS. The observed prevalence in this population was low (0.48%). Detection of this type of AAOCA
may represent useful clinical information.

Declaration of interest
The authors declare no conflicts of interest.

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References