

ANATOMIC VARIATIONS AND ANOMALIES OF THE CORONARY ARTERIES

Shajeev Jaikumar¹, Mullasari Ajit²

ABSTRACT

Background : Anomalies of the coronary arteries are a rare may be found incidentally in 0.3%–1% of healthy individuals and might be the cause of myocardial ischemia and sudden death. In this article we describe a pictorial anatomy of the coronary arteries of the heart and some of the anomalies with illustrations and CT-images. Cardiovascular computed tomography (CT) has emerged as the standard for identification and characterization of coronary artery anomalies.

Materials and methods: We present an account of incidentally diagnosed coronary artery variants and anomalies culled from a CT data of 1600 patients who underwent 64-slice CT angiography (CTA) (Siemens SOMATOM Sensation 64 CT Scanner) because of known or suspected coronary artery disease were retrospectively reviewed by two radiologists experienced in cardiovascular radiology from 2006 till 2012 at our Institute.

Results : Coronary artery anomalies (CAA) are uncommon congenital variations in coronary anatomy, occurring in 2% of the general population (as per our study)

Conclusion : Familiarity with the CT appearances of various coronary artery anomalies and an understanding of the clinical significance of these anomalies are essential in making a correct diagnosis and planning patient treatment.

Key words: coronary artery coronary anomalies coronary anatomy computed coronary angiography

INTRODUCTION

Anomalies of the coronary arteries may be found incidentally in 0.3%–1% of healthy individuals [1]. For several decades, premorbid diagnosis of coronary artery anomalies was made with angiography. However, it was

recently reported that, anomalous coronary arteries are better identified consensually with multi– detector row CT. Although coronary artery anomalies are far less common than acquired coronary artery disease, their impact on premature cardiac morbidity and mortality among young adults is not trivial. In a study by Eckart et al [2] of 126 nontraumatic sudden deaths in young adults, cardiac abnormality was found in 64 cases (51%), with coronary artery abnormalities being the most common cardiac abnormality (39 of 64 patients [61%]). The aim of this pictorial review is to illustrate the spectrum of MDCT findings that may be seen in subjects with spectrum of coronary artery anomalies, as familiarity with the CT appearances and an understanding of the clinical significance of these anomalies are essential in making a correct diagnosis and planning patient treatment

MATERIALS AND METHODS

We present an account of incidentally diagnosed coronary artery variants and anomalies culled from a CT data of 1600 patients who underwent 64-slice CT angiography (CTA) (Siemens SOMATOM Sensation 64 CT Scanner) because of known or suspected coronary artery disease were retrospectively reviewed by two radiologists experienced in cardiovascular radiology from 2006 till 2012 at our institute. *Anomalies of the coronary arteries may be found incidentally in 0.3%–1% of healthy individuals [1] but in our study the incidental cases were 2%, thus making it slightly higher than the previous reported data.*

Various imaging modalities are available for coronary artery assessment. Traditionally, catheter angiography has been used to evaluate the coronary arteries and their anomalies; however, it is an invasive technique, and angiography provides a 2-dimensional depiction of coronary anatomy, in contrast, cardiac computed

¹Consultant Radiologist, Department of Radiology. ²Director – Cardiology, Department of Cardiology, Madras Medical Mission, Chennai.

tomography (CCT), is noninvasive and has multiplanar capability.

RESULT

A total number of 1600 cases underwent 64 Slice CT Angiography in our Institute for symptomatic and asymptomatic reasons. The number of incidental findings of coronary artery anomalies in these patients is tabulated in Table 1.

From the Table 1 it is evident that, out of the 1600 patients, only 31 patients revealed congenital coronary anomalies accounting for 2 %. Among the 31 patients with coronary anomalies, the most common congenital anomaly was the origin of Right coronary artery from the Left coronary sinus, accounting to 15 patients (50%) of the 31 cases. The next common anomaly encountered was the anomalous origin of Left circumflex Artery from the right coronary sinus, accounting to 7 patients (25%) of the 31 cases. Rest of all the other anomalies as listed in the Table 1 are very rare and account for 1 % each.

TABLE 1: Coronary anomalies in our Institute

S. no.	TYPE OF CORONARY ANOMALIES	Number of Patients
1	Right coronary Artery from Left Coronary Sinus	15
2	Left Circumflex Artery from Right Coronary Sinus	7
3	Left Main Coronary Artery from Main Pulmonary Artery	1
4	Left Main Coronary Artery from Right Coronary Sinus	1
5	Left Anterior Descending Artery from Main Pulmonary Artery	1
6	Left Anterior Descending Artery from Right coronary Artery	1
7	Dual origin of Left Anterior Descending Artery	1
8	Left Anterior Descending Artery and Left Circumflex Artery from right Coronary Sinus	1
9	Separate origin of Left Anterior Descending Artery and Left Circumflex Artery from Left Coronary Sinus	1
10	Right coronary Artery from Right pulmonary Artery	1
11	Right coronary Artery from proximal Aorta	1
Total		31

Table 2: Classification of coronary arterial abnormalities

Anomalies of origin

1. Number of ostia: Multiple (>2), single
2. Anomalous location of ostium in the appropriate coronary sinus
3. Origin from opposite coronary sinus
4. Origin from noncoronary sinus
5. Origin from pulmonary artery
6. Abnormalities of angle of origin

Anomalies of course (normal origin)

Myocardial bridging

Duplication

Anomalies of termination

Coronary artery fistula

Coronary arcade

Extracardiac termination

Intrinsic coronary arterial abnormality

Coronary stenosis

Atresia

Ectasia/aneurysm

DISCUSSION

The four main coronary arteries evaluated at CT are the right coronary artery (RCA), the left main coronary artery (LCA), the left anterior descending (LAD) artery, and the left circumflex (LCx) artery. The coronary arteries are the first vessels that branch from the aorta, normally originating at the sinotubular junction.

LEFT MAIN CORONARY ARTERY

The left main coronary artery (LMCA) arises from the left posterior coronary sinus that branches into the left anterior descending (LAD) and left circumflex (LCX) coronary arteries. Occasionally, the LCA trifurcates to give the ramus intermedius.

LEFT ANTERIOR DESCENDING ARTERY

The LAD courses in the anterior interventricular groove towards the apex. It provides the diagonal branches to the anterior free wall of the left ventricle and the septal branches to the anterior interventricular septum.

CIRCUMFLEX ARTERY

The LCx artery courses in the left atrioventricular groove

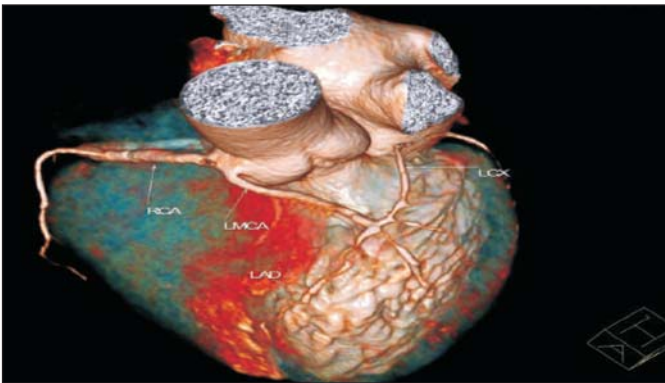


Figure 1: Single ostium - RCA and LMCA having common origin from the right coronary sinus.



Figure 2: Single Ostium –LAD arising from RCA and LCX having an interarterial malignant course

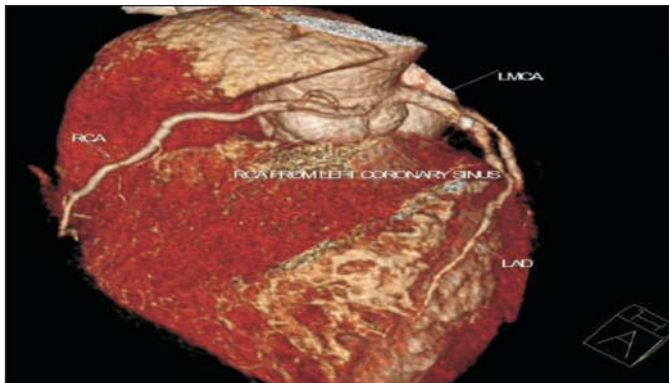


Figure 3 : RCA from the left coronary sinus.

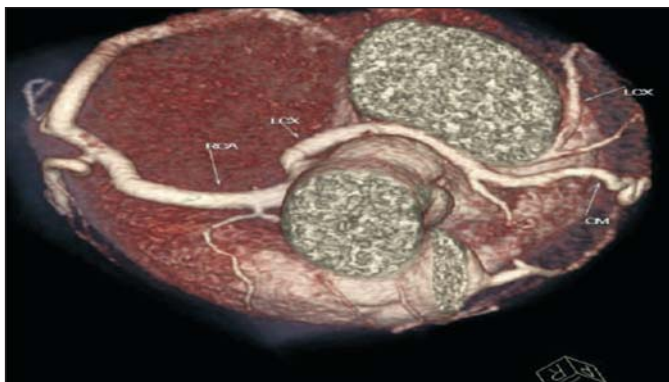


Figure 4: LCX from the right coronary sinus with an interarterial malignant course.

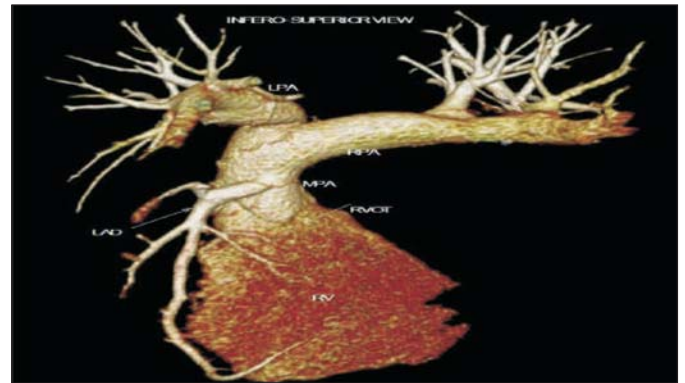


Figure 5: Left main coronary artery from main pulmonary artery.

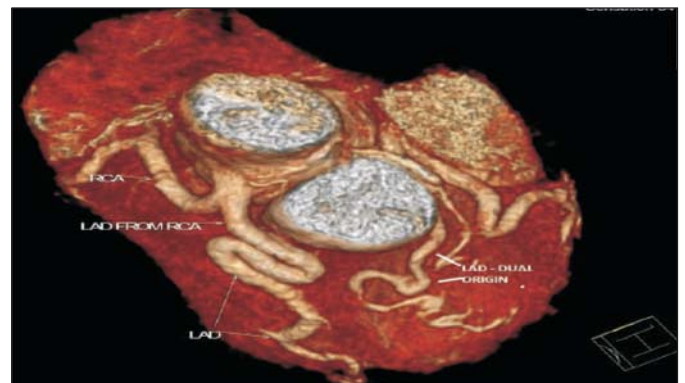


Figure 6: LAD duplication (Dual origin of LAD) one short LAD, terminates in the anterior interventricular groove without reaching the apex, and one long LAD, from the RCA/opposite sinus, enters the distal anterior interventricular groove.

and gives off obtuse marginal branches to the lateral left ventricle.

RIGHT CORONARY ARTERY

The RCA arises from the anterior right coronary sinus passes downward in the right atrioventricular groove towards the posterior interventricular septum. In more than 50% of individuals, the first branch of the RCA is the conus artery. The branch to the right ventricle at the junction of the middle and distal RCA is called the acute marginal branch. [3][4]

CORONARY DOMINANCE

Whichever artery crosses the crux of the heart and gives off the posterior descending branches is considered to be the dominant coronary artery. In approximately 85% of individuals, the RCA crosses the posterior interventricular groove and gives rise to the posterior

descending branches (right dominance); in 7%–8%, the LCx artery crosses the interventricular groove and gives rise to branches to the posterior right ventricular surface (left dominance); and in the remaining 7%–8%, the inferior interventricular septum is perfused by branches from both the distal RCA and the distal LCx artery (codominance) [3]

We have classified the coronary artery anomalies into anomalies of origin, anomalies of course, and anomalies of termination, with use of a modified version of a classification system developed by Greenberg et al [5] in Table 2

ANOMALIES OF ORIGIN

1. Number of coronary ostia. Normally there are 2 coronary ostia (one for the right main coronary artery and one for the left).

Multiple ostia. Three or more ostia are considered normal variants. This is most commonly due to the conus branch arising directly from the aorta, which is seen in 50% of subjects. The other common cause of multiple ostia is an absent left main artery with separate ostia for the LAD and LCX (estimated to be seen in 0.5% to 8% of population). [6]

Single coronary ostium. In this rare anomaly only one coronary artery arises from a single ostium (0.0024% to 0.044% of the population). [7]. It gives rise to the left main and RCA [FIG: 1] or courses directly to the LAD, LCX, and RCA [FIG: 2]. One or more arteries can have an anomalous course. This abnormality can have an adverse clinical outcome, particularly if one of the arteries takes an interarterial course (also termed a “malignant” course). [FIG:2]

2. Anomalous location of ostium in relation to the appropriate coronary sinus.

a. High ostium. This refers to a coronary ostium (either left or right) that is at least 1 cm above the sinotubular junction (instead of being at the aortic sinus) [8] [9]

b. Commissural ostium. When the ostium is located within 5 mm of the commissure between 2 sinuses, it is termed a commissural ostium. [8][9]

3. Anomalous origin of the coronary artery from opposite sinus. [FIG 3 & 4]

In this anatomy, the coronary artery arises anomalously from the opposite sinus and then takes one of the 4 paths described below. The precise path taken by the artery is important clinically. An interarterial course (previously called a malignant course) carries a high risk of sudden cardiac death, while the other 3 courses are considered nonmalignant or relatively benign

a. Interarterial. In this case, a coronary artery (such as the RCA, left main, LAD or LCX) arises from the opposite sinus and courses between the aortic root and pulmonary artery/right ventricular outflow tract [FIG 2]. This anomaly has been linked with sudden cardiac death. Several pathologic processes have been implicated; they include a narrow slit-like orifice, an acute angle of the ostium with a tangential proximal course of the ectopic coronary artery, and an intramural course where the coronary artery exits the aortic lumen and courses into the aortic wall before emerging on the surface. A consequence of this anomaly is lateral arterial compression, which worsens in systole.

b. Transseptal (subpulmonic). This refers to the left main or LAD taking a subpulmonic course. The artery traverses anteriorly and inferiorly through the interventricular septum and takes an intramyocardial course, giving off septal branches and finally emerging at its normal epicardial position. It is considered a relatively benign anomaly, though in some cases of sudden and unexpected cardiac death.

c. Retroaortic. This is the most common coronary artery anomaly, seen in 0.9% of the population. The ectopic coronary artery (more commonly the LCX) runs posteriorly between the aortic root and the left atrium [FIG 2 & 4]

d. Prepulmonic. The ectopic coronary artery runs anterior to the pulmonary artery or right ventricular outflow tract

4. Inverted coronary arteries. In this rare anomaly, the LCA arises from the right aortic sinus, and the RCA arises

from the left aortic sinus. The anomaly can become hemodynamically significant if the anomalous artery courses interarterially [10]

5. Anomalous origin of the coronary artery from noncoronary sinus. Either the RCA or the left main can arise from the noncoronary sinus. This is a rare anomaly and may have no clinical relevance. However, there are reports of cases that can be symptomatic, particularly if the proximal part of the artery has an intramural course inside the aortic wall and is hypoplastic. [11]

6. Anomalous origin of the coronary artery from pulmonary artery (ALCAPA). [FIG5] This is one of the most serious anomalies, with a 90% mortality rate in the first year of life. [11][12] Most patients are symptomatic in infancy and early childhood.

In the most common form of this anomaly, the LCA arises from the pulmonary trunk, and the RCA arises normally from the aorta (Bland-White-Garland syndrome). Extensive intercoronary collaterals develop that are often dilated and tortuous. Symptoms usually occur due to coronary steal phenomenon caused by the flow of blood from the higher pressure coronary arterial system to the lower pressure pulmonary arteries. Surgical treatment is usually recommended for these anomalies.

7. Anomalies involving the angle of origin. Normally the coronary artery arises perpendicularly to the aortic wall. However, the angle between the aortic wall and the proximal coronary artery can be quite variable. In its most extreme form, the origin can be slit-like or tangential, limiting coronary arterial flow. [8]

ANOMALIES OF COURSE

1. Myocardial bridge. In this anomaly, a portion of the coronary artery that is normally epicardial traverses through the myocardium. The myocardial tissue covering the artery is called a myocardial bridge, and the artery itself is called a tunneled segment. [13] The bridging can be superficial or deep and has been most commonly described in the mid LAD. Though myocardial bridging can be seen as a normal variant, without being clinically overt in a large proportion of cases, it has been infrequently linked with ischemia, tachycardia-induced

ischemia, conduction disturbances, myocardial infarctions, and even sudden cardiac death.[13][14] Additionally, the coronary segment proximal to the tunneled artery is vulnerable to atherosclerotic disease presumably due to low-wall shear stress, while the tunneled segment itself may be protected because of high-wall shear stress.[13]

2. Duplication. This anomaly refers to 2 LAD arteries: one short LAD, which terminates in the anterior interventricular groove without reaching the apex, and one long LAD, which originates from the LAD proper or anomalously from the RCA/opposite sinus, enters the distal anterior interventricular groove, and terminates at the apex. [FIG 6] Care should be taken to avoid mistaking a long LAD for a parallel diagonal branch. The differentiation is relatively straightforward; since a diagonal artery does not enter the anterior interventricular groove. Knowledge of this anomaly is important for planning surgical vascularization and to avoid mistaking a short LAD artery for a mid-LAD occlusion.

ANOMALIES OF TERMINATION

1. Coronary arteriovenous fistula. This is an abnormal communication between a coronary artery and a cardiac chamber, coronary sinus, superior vena cava, or pulmonary artery. The prevalence is reported to be 0.002%. [10] Various etiologies are known; they include traumatic, infectious, and iatrogenic causes. The involved fistulous artery is often dilated and tortuous. The most common site of drainage is the right ventricle creating a left to right shunt. There may be steal phenomenon with consequent myocardial ischemia. Drainage to a left-sided heart chamber causes a hemodynamic state similar to aortic regurgitation.

2. Coronary arcade. This is defined as angiographically evident connections between the RCA and LCA in the absence of coronary stenosis. Though small connections between the RCA and LCA are normal, these are not normally large enough to be visible on angiography. These connections differ from collaterals by virtue of straight connections between the coronaries in the absence of significant coronary artery disease. Coronary

arcades are mainly seen near the crux of the heart. [5] [15]

3. Extracardiac connections. Coronary arteries may have connections to extracardiac arteries, such as the bronchial, internal mammary, pericardial, superior and inferior phrenic, intercostals, and esophageal branches of the aorta. [5] These connections become significant with the development of coronary artery disease.

INTRINSIC CORONARY ARTERIAL ABNORMALITIES

1. Coronary stenosis. Though coronary stenosis is mostly acquired, congenital coronary stenosis has been described [16] and can be ostial (due to a valve-like ridge of the aortic wall or fusion of the aortic leaflets and aortic wall) or peripheral.

2. Congenital atresia of the left main coronary artery. In this condition there is complete atresia of the left coronary ostium, so the entire coronary arterial supply to the heart is derived from the RCA and its branches. The LAD and LCX are seen in their respective locations, but they receive blood from the RCA. This is an extremely rare condition and differs from a single RCA because some of the branches fill retrograde through the RCA. [16] The collateral circulation from the right to the left coronary system is usually not sufficient so almost all patients eventually develop myocardial ischemia.

3. Coronary artery ectasia or aneurysm. This lesion is defined as a coronary artery with a diameter of more than 1.5 times the adjacent normal segment and can be either focal or diffuse. Coronary aneurysms may be congenital or acquired; in the acquired group, Kawasaki disease is the most common cause of aneurysms worldwide [6] Congenital aneurysms are more commonly described in the RCA. [16] Possible complications include myocardial infarction from embolization of thrombus [6]

In this study, we have not given data on the dominance (such as right / left or co dominant) or anomalous branches (Ramus/ Conal) of the coronary arteries as these are classified as normal variants or minor anomalies These conditions result in no hemodynamic abnormality.

Conversely, our study discusses the hemodynamically significant coronary artery anomalies of origin, course, and termination that may be associated with an adverse outcome.

Anomalous origin of the coronary artery from the opposite sinus is the most potentially serious anomaly. Four patterns of anomalous origin can be observed with this anomaly: the LMCA arising from the right coronary sinus (0.09% to 0.11%); the RCA arising from the left coronary sinus (0.03% to 0.17%); the LAD or LCx arising from the right coronary sinus (0.32% to 0.67%); and the RCA, LMCA, or a branch of them arising from the non-coronary sinus [17-20]. In our study, anomalous origin of the coronary artery from the opposite sinus was observed in 25 (80%) of the 31 cases.

The Task Force on Sudden Death of the American Heart Association reported that coronary artery anomalies are the cause of 19% of deaths in athletes [21].

Complex anatomy of the coronary arteries in 3D can be accurately depicted by 64-slice CTA. It is a valid alternative to CCA in the diagnosis of the coronary artery anomalies. It also acts as a road map to the interventional cardiologist and cardiovascular surgeon, providing information about variations and anomalies in a patient before cardiac intervention or surgery to prevent possible complications of the procedure.

CONCLUSION : Familiarity with the CT appearances of various coronary artery anomalies and an understanding of the clinical significance of these anomalies are essential in making a correct diagnosis and planning patient treatment.

REFERENCES

- 1) Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation*. 2002;105:2449–2454.
- 2) Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. *Ann Intern Med*. 2004;141:829–834.
- 3) Reagan K, Boxt LM, Katz J. Introduction to coronary arteriography. *Radiol Clin North Am* 1994; 32:419–433.
- 4) Van Geuns RJ, Cademartiri F. Anatomy of the coronary arteries and vein in CT imaging. In: Schoepf UJ, ed. *CT of the heart*. Totowa, NJ: Humana, 2005; 219–228.

- 5) Greenberg MA, Fish BG, Spindola-Franco H. Congenital anomalies of coronary artery: classification and significance. *Radiol Clin North Am* 1989;27:1127-1146.
- 6) Patel S. Normal and anomalous anatomy of the coronary arteries. *Semin Roentgenol*. 2008;43:100-112.
- 7) Kim SY, Seo JB, Do KH, et al. Coronary artery anomalies: Classification and ECG-gated multi-detector row CT findings with angiographic correlation. *Radiographics*. 2006; 26:317-333.
- 8) Angelini P. Normal and anomalous coronary arteries: Definitions and classification. *Am Heart J*. 1989;117:418-434.
- 9) Angelini P, Velasco JA, Flamm S. Coronary anomalies: Incidence, pathophysiology, and clinical relevance. *Circulation*. 2002;105:2449-2454.
- 10) Dodge-Khatami A, Mavroudis C, Backer CL. Congenital heart surgery nomenclature and database project: Anomalies of the coronary arteries. *Ann Thorac Surg*. 2000;69:S270-297.
- 11) Angelini P, Walmsley RP, Libreros A, Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of valsalva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J*. 2006; 33:171-179.
- 12) Angeli E, Napoleone CP, Oppido G, et al. Anomalous origin of additional coronary artery arising from the pulmonary artery in complex congenital heart disease. *Cardiovasc Pathol*. 2008;17:190-191.
- 13) Leschka S, Koepfli P, Husmann L, et al. Myocardial bridging: Depiction rate and morphology at CT coronary angiography--comparison with conventional coronary angiography. *Radiology*. 2008;246:754-762.
- 14) Ko SM, Choi JS, Nam CW, Hur SH. Incidence and clinical significance of myocardial bridging with ECG-gated 16-row MDCT coronary angiography. *Int J Cardiovasc Imaging*. 2008;24:445-452.
- 15) Kruskal JB, Hartnell GG. Nonatherosclerotic coronary artery disease: More than just stenosis. *Radiographics*. 1995;15:383-396.
- 16) Montaudon M, Latrabe V, Iriart X, et al. Congenital coronary arteries anomalies: Review of the literature and multidetector computed tomography (MDCT)-appearance. *Surg Radiol Anat*. 2007; 29:343-355.
- 17) Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary angiography. *Cathet Cardiovasc Diagn*. 1990; 21:28-40.
- 18) Donaldson RM, Raphael M, Radley-Smith R, Yacoub MH, Ross DN. Angiographic identification of primary coronary anomalies causing impaired myocardial perfusion. *Cathet Cardiovasc Diagn*. 1983; 9:237-249.
- 19) Chaitman BR, Lesperance J, Saltiel J, Bourassa MG. Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. *Circulation*. 1976; 53:122-131.
- 20) Bunce NH, Lorenz CH, Keegan J, et al. Coronary artery anomalies: assessment with free-breathing three-dimensional coronary MR angiography. *Radiology*. 2003;227:201-208.
- 21) Maron BJ, Thompson PD, Puffer JC, et al. Cardiovascular preparticipation screening of competitive athletes. *Circulation*. 1996; 94:850-856.