MAJOR PRIMARY CONGENITAL CORONARY ARTERY ANOMALIES: AN ANGIOGRAPHIC STUDY

Priti Sinha1, Sanjeev Saxena2, S. L. Jethani3, Satyam Khare1, Shilpi Jain1, Namita Mehrotra4

1-Subharti Medical Coll. Meerut, 2-Metro heart institute Meerut, 3- HIHT University Dehradun,
4- Rama Medical Coll. Hapur, Gaziabad)

ABSTRACT

Congenital Coronary artery anomalies are found in about 1% population (0.6% to 1.5% in different geographic studies). Angiographic recognition of these anomalies is important because of their clinical significance (sudden death) and importance in patients undergoing coronary interventions or cardiac surgery. Our study included 3215 patients who underwent diagnostic coronary angiography during the 2 years period from 2008 to 2010. Twenty nine (0.90%) patients (22 male and 7 female) had major congenital coronary anomalies of which twenty seven (93.10%) patients had anomalies of origin and distribution while two (6.89%) had coronary artery fistulae. Right coronary artery was the commonest anomalous vessel, involved in 14 (48.27%) patients. It was originating from the left sinus of Valsalva in 10, from the non-facing aortic sinus in 2 and from left anterior descending artery in 2 patients. Anomalous origin of left circumflex artery was the second commonest anomaly, seen in 10 (34.48%) patients. Anomalous left anterior descending artery was found to be originating from right coronary artery in 2 (6.89%) and anomalous left coronary artery from right coronary artery was seen in one (3.4%) patient. Among patients with coronary fistulae, one (3.4%) had fistulae between the left anterior descending artery and the main pulmonary artery, one (3.4%) between the conal artery and the right atrium. The fistulae in our series were small & without significant shunt circulation. Atherosclerotic plaques in anomalous arteries were seen in only 8 (27.58%) patients, significantly less than the incidence (51.5%) of overall coronary artery disease in 3215 patients studied in this series. In two (6.89%) patients only the anomalous vessels were involved in coronary artery disease. Congenital coronary artery anomalies do not appear to be associated with an increased risk for development of coronary atherosclerosis. Recognition of coronary anomalies is important in patients undergoing coronary angiography, coronary interventions and cardiac surgery. Variations in the frequency of major congenital coronary anomalies may have a genetic background.

Keywords: Coronary anomalies, Congenital, Coronary angiography, Atherosclerotic plaque

INTRODUCTION

The most frequent cause of sudden cardiac death in young athletes is hypertrophic & right ventricular cardiomyopathy on autopsy based studies[1,2]. Congenital coronary artery anomalies are the second common cause of sudden young athletic death[3,4], specifically the anomalous coronary artery origin. Primary congenital coronary artery anomaly is one which is not associated with any other congenital structural heart anomalies. Congenital coronary artery anomalies owed to variations during early embryological development.

During embryological development, the primitive loosely packed myocardium is nourished via sinusoids, which communicate with the heart cavities. As the myocardium becomes more compact, the sinusoids disappear and give rise to a network of veins, arteries, and capillaries (at approximately 32 days of gestation) that may have connections with other mediastinal vessels. Persistence of these connections may lead to coronary artery fistulae. As the coronary artery network evolves, endothelial buds arise from the base of the truncus arteriosus. It is still unknown if initially there are only two buds, or buds from each potential cusp of the aortic and pulmonary sinuses (six buds) with later involution of all but two buds. These buds later grow and join the coronary artery network that develops from the sinusoids to establish the definitive coronary artery system. Abnormal involution (in the case of six initial buds), bud position, or septation of the truncus arteriosus may lead to the development of an abnormal origin of the coronary arteries. It is therefore expected that deviations in development may result in various ("abnormal") origins of the coronary arteries from the normal sinuses of Valsalva in the aorta or from the pulmonary artery.

Correspondence

Dr Priti Sinha

Astt. Professor , Deptt. Of Anatomy
Subharti medical college, Subharti university
Delhi haridwar bypass Road, Meerut (INDIA)
Mob: 91-9837017328
Email: drpritianatomy@yahoo.com

Although these coronary anomalies which are different from normal ones, persist since birth, they are incidentally encountered during selective angiography. Some of them may not be haemodynamically significant, but other major anomalies may predispose for sudden death, myocardial infarction and angina syndromes. Failure to recognize these major primary coronary anomalies may lead to misinterpretation and disastrous complications during coronary angiography, coronary interventions and cardiac surgery[5]. The incidence of congenital coronary anomalies in previous reports is 0.6 to 1.5 % (1-11). The aim of our analysis was to verify the incidence and to find out the more frequent types of these anatomical coronary variations.

MATERIAL AND METHODS

After taking the permission from ethical committee and pre-procedure consent from the patients for data analysis we reviewed the database of 3215 adult patients who underwent diagnostic coronary angiography in cardiac catheterization lab, during 2 year period from 2008 to 2010. Most of the selective Coronary angiography were done as per judkin's (femoral) method, although some were done as per sone's (brachial) method. Catheterization reports were analyzed, and those with coronary anomalies were selected for further assessment. The course of anomalous artery was defined according to the guidelines of Yamanaka and Hobbs[6] and the "eye-and-dot method"[7]. Patients with separate origin of conal branch from right sinus of valsalva, separate ostia for left anterior descending artery (LAD) and left circumflex artery (LCX) in left sinus of valsalva, high "take-off" of coronary arteries and coronary artery myocardial bridging were excluded. Patients with coronary anomalies associated with congenital structural heart disease were also excluded. Presence of atherosclerotic plaque causing ≥50% luminal narrowing was considered as significant coronary artery disease.

STATISTICAL ANALYSIS

Data presented as mean (± SD) and number (%). Chi-square test was used to assess any significant difference between two parameters where ever needed. p-value <0.05 were considered significant.

RESULTS:

This study included data of 3215 patients who underwent diagnostic coronary angiography during the study period. Twenty nine patients who had primary congenital coronary artery anomalies were entered into final data analysis. The indication of coronary angiography was evaluation of coronary artery disease in all the patients.

The overall incidence of congenital coronary artery anomalies was 9.02% (290 out of 3215 patients) in our angiographic population. 261 patients were later excluded as they had separate ostia for conal branch, separate origin of LAD and LCX, mayocardial-bridge, high "take-off" and coronary anomalies associated with other congenital structural heart disease. They were present in more than 1% of study population therefore considered as normal variations & not the anomalies. Thus the true prevalence of major primary congenital coronary anomalies was 0.90% (29 out of 3215 patients) of whom 22 were males (75.87%) and only 7 were females (24.13%). The mean age was 52.65 (± 15.7) years, with a range between26 and 84 years. Out of 29 patients 27 (93.1%) had anomalies related to origin of artery and distribution whole only 2 (6.89%) had coronary artery fistulae (table 1).

Anomalous right coronary artery (RCA) was the most common of the anomalies being present in 14 patients (48.27%) with angiographic incidence of 0.43% (out of 3215 patients). RCA originated from left sinus of valsalva in 10 patients (34.48%) and coursed between aorta and pulmonary artery, in 2 patients (6.89%) RCA originated from non-facing aortic sinus and coursed retroaortically. In remaining 2 patients (6.89%) it was originating from proximal segment of left anterior descending artery and coursed retroaortically (fig-1). The final distribution of RCA was normal in all these cases. More-over, the origin and distribution of the left coronary artery were also normal in 14 patients.

Anomalous origin of left circumflex artery (LCX) was the second commonest anomaly. Anomalous origin of LCX was present in 10 patients (34.48%) with angiographic incidence of 0.31% (out of 3215 patients). LCX originated from right sinus of valsalva (fig-2) in 4 patients (13.79%) and from proximal segment of RCA in 6 patients (20.68%). The initial course of left circumflex artery was retroaortic in all these cases. The distribution of LCX artery was normal in all these patients. The left anterior descending artery in all of them originated from separate ostium in left coronary sinus and had normal distribution.

Anomalous left anterior descending artery (LAD)

Coronary anomaly	No. of patients	Angiographic incidence (percent)	Anomaly incidence (percent)
Coronary artery fistulae	2	0.06	6.89
Anomalous origin of RCA from LCS	14	0.43	48.27
Anomalous origin of LCX from RCA/RCS	10	0.31	34.48
Anomalous origin of LAD from RCA	2	0.06	6.89
Anomalous origin of LMCA	1	0.03	3.44
All anomalous coronary arteries	29	0.90	100

Table-1

Incidence of different congenital coronary artery anomalies in angiographic population (total 3215 patients)

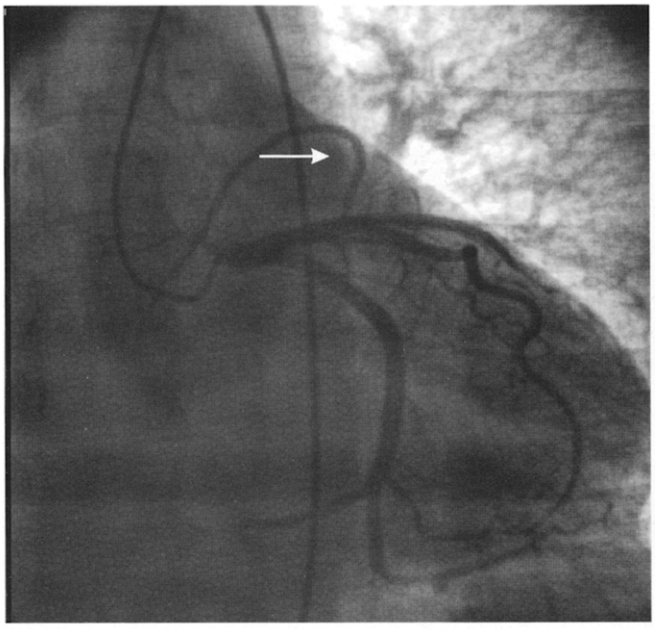

Fig-1: Anomalous Right coronary artery originating from Left anterior descending artery

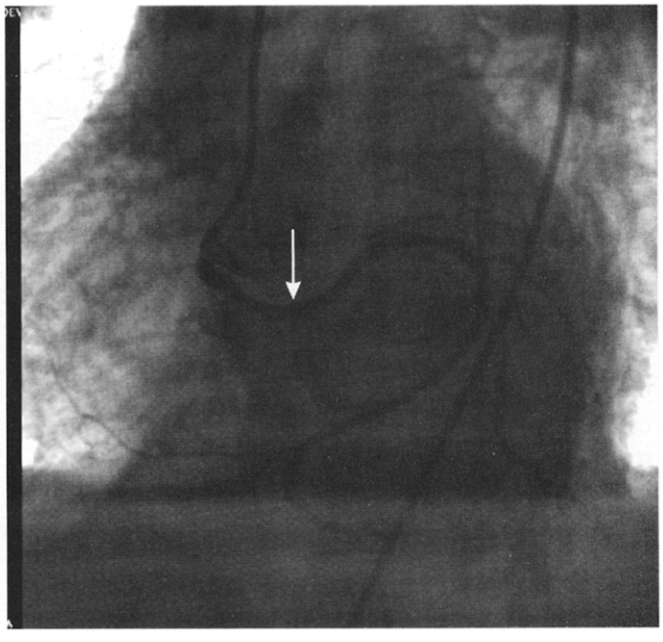

Fig-2: Left circumflex artery originating from right sinus of valsalva

originated from proximal segment of right coronary artery (RCA) in 2 patients (6.89%) with angiographic incidence of 0.06% (out of 3215 patients). These two anomalous left anterior descending coronary arteries were coursed anterior to the right ventricular out flow tract with normal peripheral distribution. The LCX artery was originated from the left coronary sinus through separate ostium with normal peripheral distribution in these two patients. Anomalous origin of left main coronary artery from right coronary artery was present in only one patient (3.4%) with angiographic incidence of 0.03% (out of 3215 patients).

In our angiographic population two patients (6.89%) had coronary artery fistulae with angiographic incidence of 0.06%. One patient (3.4%) had fistula between the conal branch of RCA and right atrium and one patient (3.4%) had fistula between left anterior descending artery and main pulmonary artery. Both the fistulae were small and not associated with haemodynamically significant shunt circulation.

The incidence of significant coronary artery stenosis due to atherosclerotic plaque (luminal narrowing ≥50%) among 3186 patients with non anomalous vessels was 51.5%. The overall incidence of significant coronary artery stenosis among 29 patients with anomalous coronary arteries was 48.27% (14 out of 29). But the significant coronary iea

stenosis in anomalous arteries was detected only in 2 patients (6.89%). Both the arteries were anomalous LCX. With this significantly lower (p value<0.05) incidence, coronary artery anomalies does not appears to be associated with increase risk of significant atherosclerotic plaque stenosis.

DISCUSSION:

The angiographic incidence of major congenital coronary artery anomalies in our study was 0.9% among patients with suspected coronary artery disease admitted for diagnostic coronary angiography. The largest angiographic review of Yamanaka and Hobbs, the incidence of congenital coronary artery anomalies among 126,595 American people was reported as 1.3% (6). The same range (0.6 to 1.5%) of incidence was reported previously in different studies with different angiographic population and with different exclusion criteria (2,5,6,11,13,15). As far as Indian population was concerned, Garg et al reported angiographic incidence of anomalous coronary artery 0.95% with the same exclusion criteria which we have opted[8].

We have excluded the patients with structural congenital heart disease, separate origin of conal branch, separate ostium of LAD and LCX, high take-off, and mayocardial-bridge. These "variations" (present in more than 1% of general population)[10] have been included in few studies[6,14] and excluded in others[8,10,15]. The most common coronary anomaly in our study was anomalous origin of RCA, however few study reported anomalous LCX as commonest anomaly[15]. But majority of angiographic study concluded anomalous RCA as most common anomaly[6,7,8,10]. Angiographic incidence of Anomalous RCA in our study was 0.43% which was in accordance with previous studies in which incidence was highest in Indian population (0.46%) & lowest in German (0.04%) populations[8]. Anomalous left coronary artery was one of the least common anomaly, being 0.03% in our angiographic population. Anomalous LAD was reported more in association with tetralogy of fellot[8].

Previous studies reported 0-1% incidence of anomalous LCX (6), being highest (1%) in central European population and lowest (0%) in Japan (8). In our study angiographic incidence of anomalous LCX was 0.31% and accounted for a 34.48% of overall incidence of major primary congenital coronary artery anomalies. Our angiographic incidence of anomalous LCX was more in accordance with Asian and Turkish population than to American populations, which might be due to ethnic and genetic factors. Anomalous LCX always coursed posterior to aorta (retroaortic). This anomaly alone causes no functional abnormality to the heart and considered benign (8). This anomaly should always be recognize during coronary angiography, especially in patients with aortic valve disease undergoing valve replacement surgery and atherosclerotic coronary artery disease[1,5,10].

Angiographic recognition of major coronary anomalies before cardiac surgery is always necessary and failure might lead to prolonged procedure and disastrous complications[8]. Surgical complication might arise if an anomalous artery was excluded from cardiopulmonary bypass surgery or surgeon incises this anomalous artery accidentally. Accurate identification of origin and course of anomalous coronary artery was mandatory before doing diagnostic and therapeutic coronary interventions to select correct catheters, wires and balloon systems.

In accordance to various previous studies we also found that the presence of anomalous artery did not increase the incidence of atherosclerotic coronary artery disease[6]. In fact the incidence was significantly less in anomalous vessel, suggesting that anomalous arteries were relatively protected from atherosclerotic stenotic disease[8]. The site of origin and its course might be important to determine how early and severe atherosclerotic lesion developed. Further studies were needed to determine relation between anomalous coronary arteries and occurrence of atherosclerotic disease.

CONCLUSION

The incidence of major primary congenital coronary artery anomalies in our angiographic population was similar as reported in other populations. The most common was RCA anomaly. Coronary anomaly did not increase the risk of developing atherosclerotic disease. In fact anomalous artery appeared less prone for stenotic lesions. Identification of anomalous vessels is important before coronary interventions and cardiac surgeries.

REFRENCES

1. Maron B.J., Roberts W.C., McAllister H.A.; et al. Sudden death in young athletes. Circulation, 1980, 62: 218-229.

2. Burke A.P., Farb V., Virmani R.; et al. Sports-related and non-sports-related sudden cardiac death in young adults, Am Heart J, 1991, 121: 568-575.
3. Taylor A.J., Byers J.P., Cheitlin M.D., Virmani R.; Anomalous right or left coronary artery from the contralateral coronary sinus. "high-risk" abnormalities in the initial coronary artery course and heterogeneous clinical outcomes, Am Heart J, 1997, 133: 428-435.
4. Cheitlin M.D.; Coronary anomalies as a cause of sudden death in the athlete, Estes N.A.M., Salem D.N., Wang P.J.; Sudden Cardiac Death in the Athlete 1998 Futura Publishing Armonk, NY 379-391
5. Sinha P., Mehrotra N., Jethani S.L., et al. Histo-morphological study of coronary artery. Journal of Anatomical Society of India, 2009, 58 (2): 166-168.
6. Yamanaka O, Hobbs RE: Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Catheterization and Cardiovascular Diagnosis 1990, 21(1):28-40.
7. Maron BJ: S Serota H, Barth CW, Seuc CA, Vandormael M, Aguirre F, Kern MJ: Rapid indentification of the course of anomalous coronary arteries in adults: the "dot and eye" method. The American Journal of Cardiology 1990,65(13):891-898.
8. Garg N, Tewari S, Kapoor A, Gupta DK, Sinha N: Primary congenital anomalies of the coronary arteries: a coronary arteriographic study. International Journal of Cardiology 2000, 74(1):39-46.
9. Makaryus AN, Orlando J, Katz s: Anomalous origin of the left coronary artery from the right cornary artery: a rare case of single coronary artery originating from the right sinus of Valsalva in a man with suspected coronary artery disease. The Journal of Invasive Cardiology 2005, 17(1):56-58.
10. Angelini P: Coronary artery anomalies: an entity in search of an identity. Circulation 2007, 115(10):1296-1305.
11. Dogan SM, Gursurer M, Aydin M, Gocer H, Cabuk M, Dursun A: Myocardial ischemia caused by a coronary anomaly left anterior descending coronary artery arising from right sinus of Valsalva. International Journal of Cardiology 2006, 112(3): e57-59.
12. Mirchandani S, Phoon CK: Management of anomalous coronary arteries from the contralateral sinus. International Journal of Cardiology 2005, 102(3):383-389.
13. Aydinlar A, Cicek D, Senturk T, Gemici K, Serdar OA, Kazazoglu AR, Kumbay E, Cordan J: Primary congenital anomalies of the coronary arteries: a coronary arteriographic study in western Turkey. International Heart Journal 2005, 46(1):97-103.
14. Pelliccia A: Congenital coronary artery anomalies in young patients:new perspectives for timely identification. Journal of the American College of Cardiology 2001,37(2):598-600.
15. Kardos A, Babai L, Rudas L, Gall T, Horvath T, Talosi L, Toth K, Sarvary L, Szasz K: Epidemiology of congenital coronary artery anomalies: a coronary arteriography study on a central European population. Catheterization and Cardiovascular Diagnosis 1997,42(3):270-275.