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CORONARY ARTERY ANOMALIES;
AN AFIC/NIHD EXPERIENCE

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ABSTRACT ... Introduction: Congenital anomalies of the coronary arteries occur in 0.2% to 1.2% of the general population[1]. The incidence of various coronary anomalies and associated clinical, angiographic and hemodynamic findings have been cited in several internationally published clinical series[4-8]. To compare our experience with previously reported studies, we have reviewed clinical and angiographic findings for 50 adult patients with coronary artery anomalies. **Patients and Methods:** We surveyed the records of 5050 consecutive adult patients who had undergone coronary angiography. **Setting:** Armed Forces Institute of Cardiology and National Institute of Heart Disease (AFIC/NIHD) Rawalpindi. **Period:** 1st Jan 2004 and 30th April 2005, and identified 50 adults with various coronary artery anomalies. **Results:** 5050 reports were reviewed and 50 (0.9%) coronary artery anomalies were identified in 50 patients. Different anomalies identified are; both coronary arteries from right sinus of Valsalva (RSV)-(n = 1), both coronary arteries arising from the left coronary sinus (n = 4), single coronary arteries (n = 2), LCx from RSV/RCA (n=6), anterior descending artery arising from the right coronary sinus (n = 1), coronary artery fistulae (n = 4), separated origin of anterior descending and left circumflex coronary arteries (n = 25), and separate origin of conus/ RV branch (n = 7). The initial course was retroaortic in all the circumflex arteries, interarterial in the right coronaries, and anterior in the anterior descending arteries. **Conclusions:** We conclude that adult congenital anomalies of the coronary arteries are not uncommon finding in a tertiary care cardiac center. Separate origin of LAD and LCx from LSV and left circumflex coronary artery arising from RSV/RCA are the most frequently diagnosed anomalies.

Key words: Anomalous coronary arteries, Single coronary artery, Left circumflex, LAD, RCA

INTRODUCTION

Congenital anomalies of the coronary arteries occur in 0.2% to 1.2% of the general population[1]. Depending upon the origin, course and termination of anomalous vessel, certain coronary anomalies may be associated with sudden death, syncope, other congenital heart disease, anginal syndromes, or they may be incidental findings without adverse prognosis[2-3]. Accurate recognition and documentation of coronary artery anomalies at the time of coronary angiography are essential to determine the significance of such findings and to avoid therapeutic complications. The incidence of various coronary anomalies and associated clinical, angiographic and hemodynamic findings have been cited in several internationally published clinical series[4-8]. To compare our experience with previously reported studies, we have reviewed clinical and angiographic findings for 50 adult patients with coronary artery anomalies. These anomalies, if associated with atherosclerotic disease, have been successfully tackled with angioplasty.

PATIENTS AND METHODS

We surveyed the records of 5050 consecutive adult patients who had undergone coronary angiography at the Armed Forces Institute of Cardiology and National Institute of Heart Disease (AFIC/NIHD) Rawalpindi between 1st Jan 2004 and 30th April 2005, and identified 50 adults with various coronary artery anomalies. Clinical characteristics of each patient had been recorded at the time of catheterization. We grouped the anomalies (Table-I) and defined the initial course of the anomaly.

Table-I.
Anomalies
Both coronary arteries arising from left sinus of valsalva
Both coronary arteries arising from right sinus of valsalva
Cx artery arising from right sinus of valsalva or RCA
Only one coronary artery exist
LAD from right sinus of valsalva or RCA
Coronary artery communicates aberrantly with a cardiac chamber or major thoracic vessel.
LAD and LCx arising separately (double barrel anatomy) from left sinus of valsalva
Conus/RV branch arising separately from right sinus

RESULTS

Five thousand and fifty angiography reports were reviewed and 50 adult patients were identified with 50 (0.9%) coronary artery anomalies (Table-II).

Table-II. Distribution of Anomalies and associated conditions in 50 patients with 50 coronary artery anomalies.

Anomalies	No. Of Anomalies	CAD in Anomalies Vessel	CAD in other Vessels	PCI in Anomalous Vessels
Both CA from RSV	01	-	-	-
Both CA from LSV	04	02	02	-
Cx from RSV or RCA	06	03	01	LCx
LAD from RCA	01	-	-	-
Single CA	01	01	-	LCx
CA Fistulas	04	01	-	-
Separate origin of LAD and LCx from LSV (double barrel anatomy)	26	-	10	-
Separate origin of conus/RV branch from RSV	07	-	-	-
Total	50	07	13	02

18 patients (36%) had associated coronary artery disease (defined as more than 50% luminal stenosis of 1 or more major epicardial coronary arteries). Out of these 18 patients, 5 had involvement of anomalous arteries, 2 had involvement of anomalous and other vessels and 11 had disease in other vessels only.

Origin of both Coronary Arteries from Right Sinus of Valsalva

The LCA originated from the right sinus of Valsalva in one female ptient who was 46 years old. Coronary angiography was carried out for evaluation of chest pain and positive exercise tolerance test. The initial course of the LCA in this patient was between the aorta and pulmonary artery (Fig 1 & 2 normal, Fig. 3 anomalous LCA). The course and distribution of the RCA was normal.

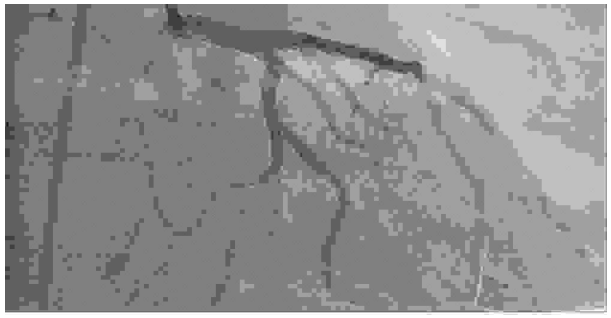

Fig-1. Normal coronary artery (Left)

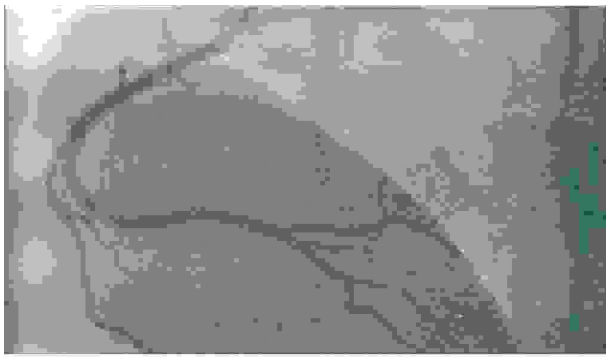

Fig-2. Normal coronary artery (Right)

Origin of both Coronary Arteries from the Left Sinus of Valsalva

The RCA arose from the left sinus of Valsalva in 4 patients, beginning anteriorly to the left main coronary artery (LM) and coursing anteriorly between the aorta and pulmonary artery (Fig. 4). All were male patients with age of 38- 55 years. These patients were evaluated for chest pain. Two patients were found to have coronary artery narrowing of 80% and 90% respectively in the anomalous portion of the proximal RCA. Both were successfully tackled with PCI.

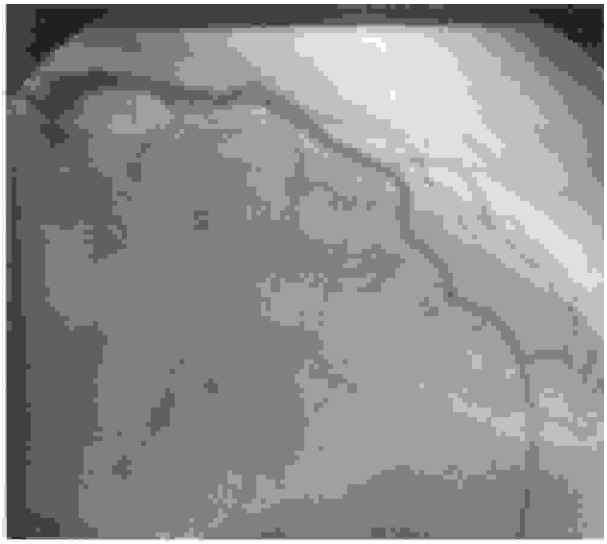

Fig-3. Anomalous left coronary artery

Anomalous Origin of the Circumflex Coronary Artery

Anomalous origin of the LCx from the right aortic sinus or the 1st portion of the RCA was observed in 6 patients (4 males and 2 females). In all cases, the initial course of the LCx was posterior to the aorta (Figs. 5). Coronary artery disease was found in 4 patients. Two patients had SVCAD involving proximal portion of anomalous LCx (70% and 80% stenosis respectively). Both were successfully opened up with PCI. One patient had TVCAD with moderate LV function. He was referred for CABG surgery. One patient had DVCAD involving LAD and RCA.

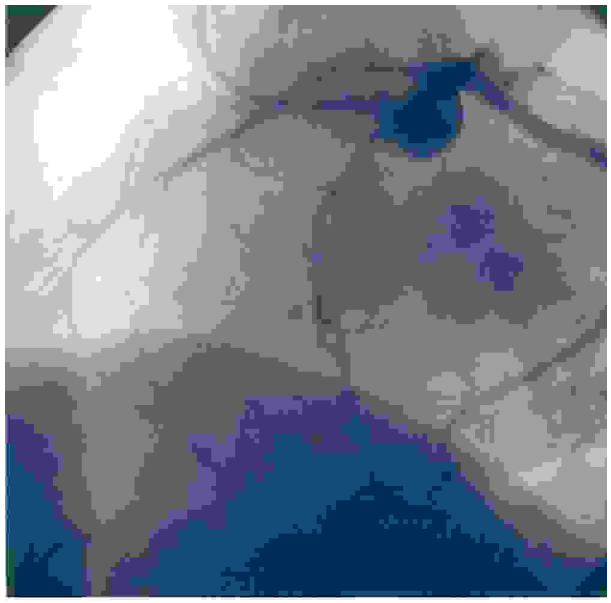

Fig-4. Origin of both coronary arteries from left coronary sinus of valsalva

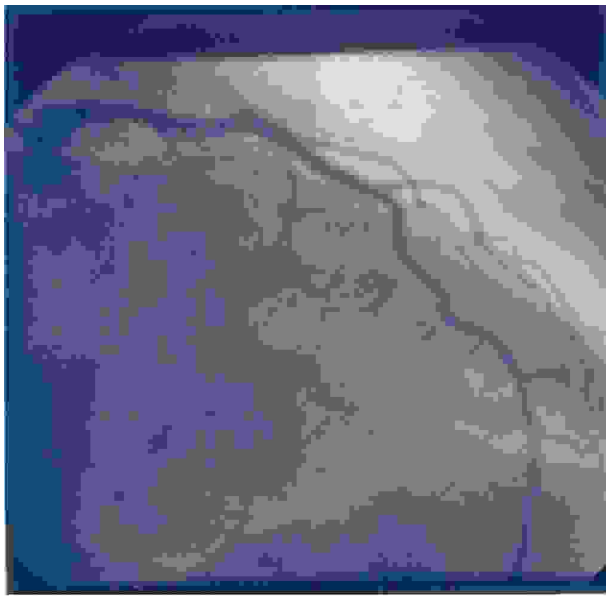

Fig-6. LAD from right aortic sinus

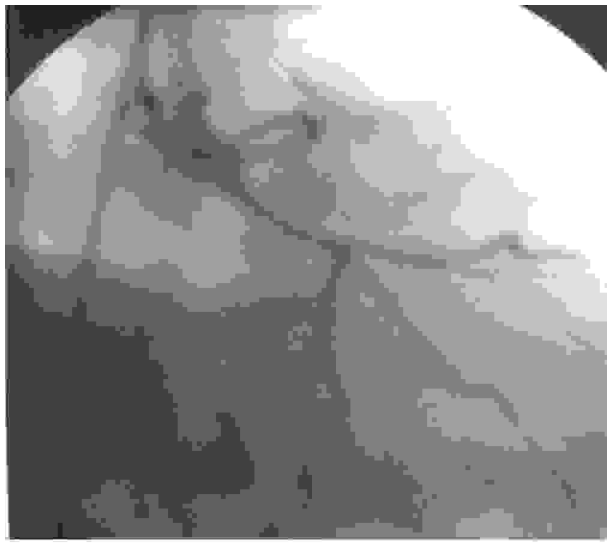

Fig-5. Lcx from right sinus of valsalva

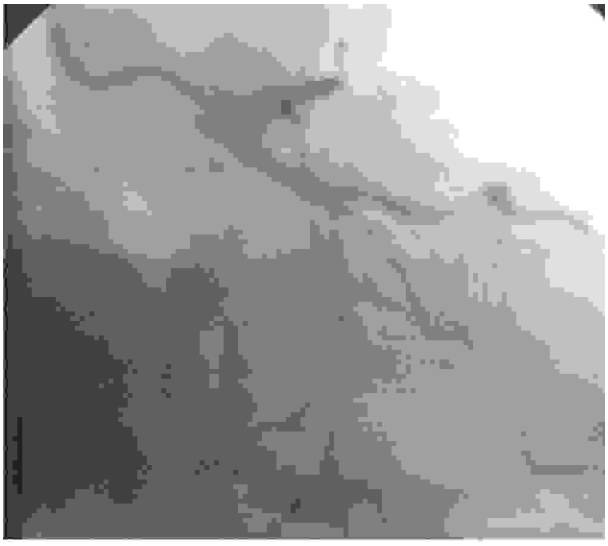

Fig-7. PCI to aberrant LCx

Anomalous Origin of the Left Anterior Descending Artery from the Right Aortic Sinus

The LAD originated from the right aortic sinus in one male patient of 46 years. The initial course of the LAD was anterior to the right ventricular outflow tract (Fig.6). Coronary arteries were disease free.

Single Coronary Artery

Single coronary artery was observed in two male patients of 57 and 52 years of age respectively. One of these patients had critical disease in the LCx, which was successfully dealt with PCI (Fig 7). Coronary arteries of other patient were normal. The initial portion of the single

coronary artery in both patients followed the path of a normal LCA but continued posteriorly in the atrioventricular groove to the area of the heart normally supplied by the RCA (L-1 anomaly).

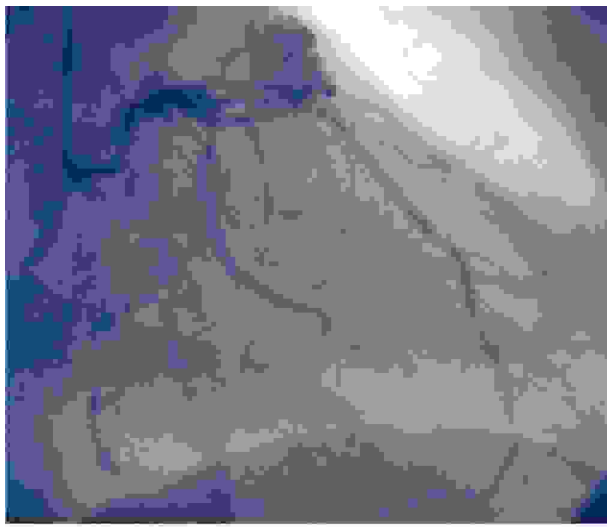

Fig-8. Fistula between LAD and pulmonary artery

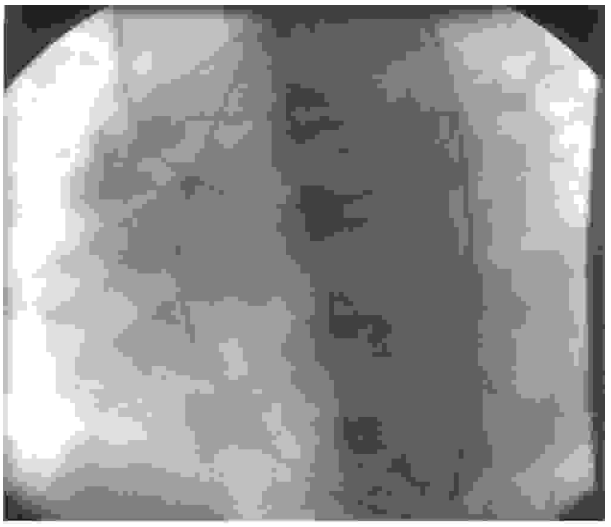

Fig-9. Fistula between LAD and pulmonary artery

Coronary artery Fistula

One patients had a big fistulous communication between left system and RV. Other three had small fistulas between LAD and pul artery/ LA (Fig 8 & 9).

Separate origin of LAD & LCx (Double barrel anatomy)

Double barrel anatomy of the left system was found in 25 patients (fig 10). Ten patients had significant narrowing of proximal LAD (`~70% stenosis) and were tackled with PCI successfully.

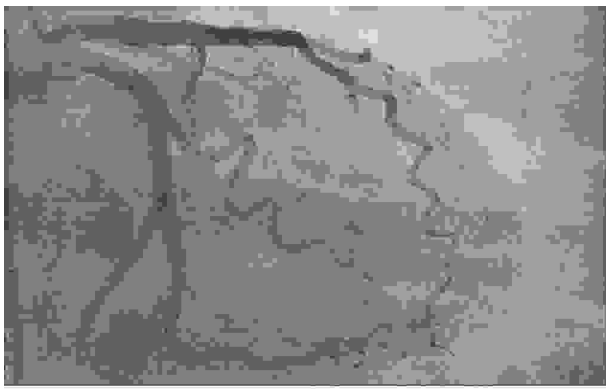

Fig-10. Normal coronary artery (Right)

Separate origin of RCA and conus branch or RV branch was found in 7 patients.

DISCUSSION

The incidence of coronary artery anomalies in our review is 0.9%, which compare well with the incidence from other studies of patients referred for coronary angiography[7,9] In our study, as in others[7,9] coronary artery anomalies appear to be more common in men than in women (41 males vs 9 females), although this finding may reflect the selective nature of referral for cardiac catheterization and coronary angiography. We found 36% of patients with anomalous coronary arteries associated with atherosclerotic CAD; whereas Wilkin et al[9] have described 68% incidence of atheromatous heart disease in patients with anomalous coronary arteries. We attribute this difference to relatively younger patients and a relatively higher bias towards angiography in view of strict fitness criteria of the armed forces personnel in our study. Anomalous origin of both coronary arteries from

the left sinus of Valsalva was found in 4 patients (.08%) while in other studies it has been reported around .28%[9]. The incidence of this anomaly in the general population is unknown. This anomaly has been associated with acute myocardial infarction, angina pectoris, syncope, ventricular tachycardia, ventricular fibrillation, and sudden death, in the absence of atherosclerotic or other cardiac disease. On the basis of angiographic studies, it has been estimated that approximately one-third of all patients with this anomaly will have symptoms of myocardial ischemia or dysfunction[9]. Myocardial ischemia in association with this anomaly is thought to he caused by an abnormal slit-like RCA ostium, acute angulation of the RCA, and compression of the RCA between the aorta and pulmonary trunk during exercise [9-13]. The lower incidence of this anomaly in our population may be explained on account of study of only symptomatic patients. It is quite possible that many such patients succumb before reaching hospital. Origin of both coronary arteries from the right sinus of Valsalva is found in 0.06% to 0.19% of patients undergoing coronary angiography[7]. In our study, this anomaly was identified in only 1 patients, or .02%. Four possible courses of the left coronary artery have been reported: anterior to the pulmonary trunk, posterior to the aorta, within the intraventrictlar septum beneath the right ventricular outflow tract, and between the aorta and pulmonary trunk[14]. In our patient the course was between the aorta and pulmonary trunk. Sudden death has been described in patients with this anomaly either during or immediately after the exercise[8,15]. The significance of this anomaly in patients 20 years of age or older is not clear. Barth and Roberts[17], have described one case of fatal myocardial ischemia past the 2nd decade, due to this anomaly. Most young patients who die suddenly with this anomaly, have prodromal ischemic symptoms[15-16]. Ischemia is thought to be caused by compression of the LCA between the aorta and pulmonary trunk during exercise, a spasm or kinking of the LCA, acute angulation of the LCA, or an anatomic abnormality at the orifice of the ostium[8,10,15-16]. Noninvasive evaluation of these symptoms is usually unrewarding, with the exception of echocardiography, which can exclude aortic stenosis or hypertrophic cardiomyopathy. Roberts[10] recommends coronary angiography if the results of a stress electrocardiographic study are abnormal, or if symptoms persist despite a normal stress electrocardiogram. Anomalous origin of the LCx from the RCA or the right sinus of Valsalva is the most common major coronary anomaly reported in angiographic series and necropsy studies.[9-10] It was found in .23% of our patients. This anomaly is thought to be of little clinical significance unless valve surgery or coronary artery bypass surgery is performed without previous detection of the anomaly, or unless severe atherosclerotic narrowing is present in the RCA proximal to the origin of the LCx[17-19]. Three of our patients with this anomaly had significant stenosis of the proximal LCx. Two patients had SVCAD involving LCx which were tackled successfully with PCI. Third patient had severe TVCAD and was referred for CABG surgery. It appears that in patients with coronary artery disease, there is a predilection for the development of atherosclerosis in the posteriorly coursing anomalous vessel. Kimbiris and associates[7] observed similar findings in 2 patients with LCAs originating from the right sinus of Valsalva and coursing posteriorly. Anomalous origin of the LAD from the RCA or the right sinus of Valsalva is rare in the absence of other cardiac abnormalities[20]. It occurred in one of our patients. Kimbiris and colleagues[7] reported an incidence of 0.03%. The course of the LAD is either anterior to the right ventricular outflow tract or through the interventricular septum at the level of infundibulum. This anomaly is frequently associated with tetralogy of Fallot, and vessels have been divided inadvertently during corrective procedures[21].

Single coronary artery is a rare anomaly, occurring in two of our patients. The majority of patients younger than 20 years of age present with an associated abnormality-most frequently transposition of the great vessels or coronary artery fistula-while older patients have a low incidence of associated anomalies[10]. In the absence of significant coronary atherosclerosis , a single coronary artery may be a benign finding unassociated with functional or anatomic evidence of ischemia[22]. In our series of adult patients, we have not found anomalous

coronary artery arising from pulmonary artery. It is a rare finding in adults who are undergoing coronary angiography for suspected ischemic heart disease.[10] However, in our pediatric group one infant was found to have ALCAPA, which was surgically corrected. Seventy-five percent of patients with this anomaly develop symptoms of congestive heart failure or myocardial ischemia in the 1st 4 months of life, and most die within 2 years of the onset of symptoms[23].

The majority of fistulas originating from coronary arteries terminate in the right side of the heart, and are true left-to-right shunts. One of our patients had large fistulous communication between LCA and right ventricle. Minor fistulas are not uncommon and are of little clinical significance. Approximately one-half of the patients with large fistulas develop complications, which include congestive heart failure, sub-acute bacterial endocarditis, myocardial ischemia, and rupture of an aneurysmal fistula[18].

We have found that separate origin of LAD and Cx or conus/RV branch from their respective sinuses is quite common. PCI in cases of double barrel anatomy is convenient because of selective guide catheter engagement.

We conclude that adult congenital anomalies of the coronary arteries are not uncommon finding in a tertiary care cardiac center. Separate origin of LAD and LCx from LSV and left circumflex coronary artery arising from RSV/RCA are the most frequently diagnosed anomalies.

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EVERYBODY'S BUSINESS IS NOBODY'S BUSINESS

Sir Philip Sidney