Retrospective Study

Congenital coronary artery fistulas complicated with pulmonary hypertension: Analysis of 211 cases

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Abstract

AIM
To compare the behavior of pulmonary hypertension (PHT) associated with coronary artery fistulas (CAFs) between the Asian and Caucasian subjects.

METHODS
CAFs may be complicated with PHT secondary to left-to-right shunt. Literature review limited to the English language. A total of 211 reviewed patients were collected. Of those, 111 were of Asian and 100 were of Caucasian ethnic origin. The mean age of the Asian and the Caucasian groups of patients were 48.9 (range 19-83) and 49.9 years (range 16-85), respectively. In both groups, right heart catheterization was the most commonly (95%) used method for determining pulmonary artery pressure.

RESULTS
From all of the reviewed subjects, PHT was found in 49 patients (23%), of which 15 were Asian and 34 were Caucasian. In 75% of PHT subjects, mild to moderate PHT was reported and 76% of the fistulas had a vascular mode of termination. Treatment was surgical in 61%, followed by percutaneous therapeutic embolization (27%) and finally conservative medical management in 12% of PHT subjects. PHT was associated with a slight female gender predominance. The majority demonstrated mild to moderate PHT. PHT was reported more frequent in the Caucasian compared with the Asian ethnicity group. The majority of fistulas in patients with PHT had a vascular mode of termination. The results of this review are intended to be indicative and require cautious interpretation.

CONCLUSION
The likelihood for a CAF patient to develop PHT is presented when possessing the following features, with a Caucasian female having a fistula with a vascular mode of termination.
Key words: Congenital coronary artery fistulas; Congenital anomaly; Pulmonary hypertension; Asian population; Caucasian population

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Core tip: Congenital coronary artery fistulas (CAFs) are infrequent but hemodynamically important anomalies which may evolve a myriad of complications, such as myocardial infarction, congestive heart failure, infective endocarditis, aneurysm, rupture, pericardial effusion, arrhythmias and sudden death. In addition, secondary pulmonary hypertension (PHT) may complicate the course of CAFs. Moreover, when monitoring CAF patients, the clinicians responsible for the management of patients with congenital CAFs should be aware of the development of PHT during the course of the disease.

INTRODUCTION

Congenital coronary artery fistulas (CAFs) are uncommon anomalies. Most CAFs are small and hemodynamically inconsequential with a negligible shunt. However, some can be sizeable and lead to shunting of blood from the coronary circulation to low-pressure pulmonary vascular bed, resulting in pulmonary hypertension (PHT)\(^1\). CAFs may be associated with normal\(^2-4\) pulmonary artery pressure (PAP) in unilateral\(^5-8\) or bilateral\(^9,10\) fistulas, or may sometimes be accompanied with elevated PAP\(^11-14\).

Rarely, in octogenarians with bilateral CAFs, PAP may remain normal\(^15\).

The hemodynamic consequences of CAFs varies, depending on their magnitude and the cardiac chamber or vascular site involved. Fistulas terminating into the right heart chambers may produce left-to-right shunt and volume overload of the pulmonary circulation, whereas fistulas to the left heart side cause left ventricular volume overload.

In a literature review, 211 subjects were included and a comparison was made between the Asian\((n = 111)\) and Caucasian\((n = 100)\) subjects regarding the behavior of PAP associated with CAFs.

RESULTS

Total group

A total of 211 (M: 87 = 41% and F: 124 = 59%) reviewed patients were collected from the world literature. The mean age was 49.4 years (range 16-85). The reported method of assessment of PAP was RHC\((n = 201, \text{Caucasian } n = 94 \text{ and Asian } n = 107)\) and Doppler echocardiography\((n = 10, \text{Caucasian } n = 6 \text{ and Asian } n = 4)\) in 95% and 5% of the subjects, respectively. The congenital CAFs were unilateral in 118 (56%), bilateral in 87 (41%) and multilateral in 6 (3%) of the subjects. The CAFs arose from the right\((133/268 = 49.6\%)\) and left\((135/268 = 50.4\%)\) coronary artery, respectively. The mode of termination was either vascular\((90/211 = 42.6\%)\) or vascular site involved. Fistulas terminating into the pulmonary or a cardiac disorder\(^16\).

Non-invasive method: In accordance with the European Society of Cardiology criteria for detecting the presence of PHT, based on the TR peak velocity and Doppler-calculated sPAP at rest (assuming a normal right atrial pressure of 5 mmHg), additional echocardiographic variables suggestive of PHT were used to determine the sPAP\(^16,18,20\). PHT was defined by an estimate of right ventricular systolic pressure of greater than 40 mmHg. sPAP is estimated using TR jet velocity based on the simplified Bernoulli’s equation\(4 \times (\text{TRV})^2 + \text{RA pressure})\(^19,21,22\) (TRV: TR velocity; RA: Right atrium). PHT was classified into three categories: Mild\((40-49 \text{ mmHg})\), moderate\((50-59 \text{ mmHg})\) and severe\((> 59 \text{ mmHg})\).

Statistical analysis

Values were expressed as means, averages, and percentages.
Said SAM. CAFs and PHT

Table 1 Reviewed Asian (n = 111) and Caucasian (n = 100) group of patients

| Gender | Total reviewed subjects | Asian group | Caucasian group |
|--------|-------------------------|-------------|----------------|
| n      |                         | 111 (53%)   | 100 (47%)      |
| F      | 124 (59%)               | F 63 (57%)  | F 61 (61%)     |
| M      | M 87 (41%)              | M 48 (43%)  | M 39 (39%)     |
| Mean age (range), yr | 49.4 (16-85) | M 48 (43%) | M 39 (39%) |
| Mean PAP | 56.8 (16-80) | M 48 (43%) | M 39 (39%) |

CAF characteristics
- Unilateral: 118 (56%)
- Bilateral: 47 (21%)
- Multilateral: 46 (21%)

Mode of termination
- CVFs: 90 (43%)
- CCFs: 121 (57%)
- RHC: 201 (95%)
- Average age: 35.6 ± 15.3 years
- Average PAP: 35.6 ± 15.3 mmHg

Management
- Surgical ligation: 124 (59%)
- Conservative medical management: 76 (76%)
- Percutaneous therapeutic embolization: 43 (57%)
- Right heart catheterization: 26 (25%)
- Watchful waiting: 2 (1%)

Table 2 Asian and Caucasian group of patients (n = 49) with pulmonary hypertension

| Gender | Total group | Asian group | Caucasian group |
|--------|-------------|-------------|----------------|
| n      | 49          | 15 (31%)    | 34 (69%)       |
| Age1   | 56 (16-80)  | 54.4 (24-77)| 56.8 (16-80)   |
| Gender | F 34 (69%)  | F 12 (80%)  | F 22 (65%)     |
|        | M 15 (31%)  | M 3 (20%)   | M 12 (35%)     |
| CAF    |             |             |                |
| Unilateral | 37 (76%)  | 9 (60%)     | 28 (82%)       |
| Bilateral | 12 (24%)   | 6 (40%)     | 6 (18%)        |
| PHT    |             |             |                |
| Mild   | 26 (53%)    | 8/15 (53%)  | 18/34 (33%)    |
| Moderate | 11 (22%)   | 2/15 (13%)  | 9/34 (26%)     |
| Severe | 12 (25%)    | 5/15 (33%)  | 7/34 (21%)     |
| Mean PAP | 35.6 (range 26-60) | 36.9 (range 27-54) | 34.3 (range 26-60) |

Mode of termination
- Mean Qp/Qs: 1.9 (range 1.12-1.27)
- RH: 43 (88%)
- SL: 6 (12%)
- PTE: 6 (12%)

Origin
- R 8, L 10, bilateral: 11
- R 6, L 22, bilateral: 5

Termination
- RH side 45: 19
- Side 2: 9
- Side 13: 12

Associated disorders
- M: 87 (41%)
- F: 63 (57%)
- RH side: 6 (12%)
- L: 6 (12%)

Subjects from ref. [35] were not included in calculation of mean age. Mean age was calculated from 170 (70 Asian and 100 Caucasian) subjects. One PTE failed (from ref. [147]) followed by SL treatment and another treated with hybrid procedures (from ref. [133]). CAF: Coronary artery fistula; CCFs: Coronary-cameral fistulas; CVFs: Coronary-vascular fistulas; CMM: Conservative medical management; F: Female; M: Male; PTE: Percutaneous therapeutic embolization; RH: Right heart catheterization; SL: Surgical ligation; PAP: Pulmonary artery pressure. PHT: Pulmonary hypertension; PTE: Percutaneous therapeutic embolization; RHC: Right heart catheterization; SL: Surgical ligation; S/P: Systolic pulmonary artery pressure.

Table 3

| Total reviewed subjects | Asian group | Caucasian group |
|-------------------------|-------------|----------------|
| n                       | 211         | 111 (53%)      | 100 (47%)       |
| Gender                  | F 124 (59%) | F 63 (57%)     | F 61 (61%)      |
|                        | M 87 (41%)  | M 48 (43%)     | M 39 (39%)      |
| Mean age (range), yr    | 49.4 (16-85)| 48.9 (18-93)   | 49.6 (16-85)    |
| Mean PAP                | 56.8 (16-80)| M 48 (43%)     | M 39 (39%)      |

CAF characteristics
- Unilateral: 118 (56%)
- Bilateral: 47 (21%)
- Multilateral: 46 (21%)

Mode of termination
- CVFs: 90 (43%)
- CCFs: 121 (57%)
- RHC: 201 (95%)
- Average age: 35.6 ± 15.3 years
- Average PAP: 35.6 ± 15.3 mmHg

Management
- Surgical ligation: 124 (59%)
- Conservative medical management: 76 (76%)
- Percutaneous therapeutic embolization: 43 (57%)
- Right heart catheterization: 26 (25%)
- Watchful waiting: 2 (1%)

PHT was found in 15 Asian (14%) (M, n = 3; F, n = 12) subjects with a mean age 54.4 years (range 24-77). Among the 15 subjects, mild, moderate and severe PHT was detected in 8, 2 and 5, respectively.

Caucasian population: n = 100

The review of 170 subjects (111 Male n = 48 (43%) and Female n = 63 (57%)) had a mean age of 48.9 years (range 19-83).

Between 1986 and 2014, papers published describing Asian population with congenital CAFs and reported data on PAP were included: from 1986-1990, 1994-1999, 2001-2004, 2005-2007, and 2009-2014. PAP was measured by RHC in 107 and by Doppler echocardiography in 4.

Ninety-six subjects (86%) had normal PAP. Among the CAFs, 42 were unilateral (38%), 63 bilateral (57%) and 6 multilateral (5%). The treatment modalities were SL [82 (74%)], CMM [20 (18%)] and PTE [9 (8%)]. No watchful waiting strategy was conducted and death did not occur in any of the subjects.
reported. Sixty-six subjects (66%) had normal PAP.

Treatment modalities included SL (42), PTE (20), CMM (18), and watchful waiting (2), and were not mentioned in 16 cases. There were 2 mortalities (2). PHT was found in 34 subjects (34%) [M: n = 12 (35%) and F: n = 22 (65%)], with a mean age of 56.8 years (range 16-80).

**PHT population: n = 49**

PHT was found in 49 patients (49/211 = 23%), with a mean age of 56 years (range 16-80). There were 34 females (69%) and 15 males (31%), with 15 Asian (mean age 54.4, range 24-77 years) and 34 (mean age 56.8, range 16-80 years) of Caucasian patients. The fistulas were unilateral in 37 (76%) and bilateral in 12 (24%) of the subjects. Measurement of PAP was achieved by RHC in 43 subjects (13 Asian and 30 Caucasian) and by Doppler echocardiography in 6 (2 Asian and 4 Caucasian) subjects. Mild, moderate and severe PHT was reported in 26 (53%), 11 (23%) and 12 (24%) subjects, respectively (Table 2).

The percentage of unilateral and CVFs was higher in the Caucasian group (82% and 82%) compared to the Asian group (60% and 60%), respectively (Table 3).

**DISCUSSION**

CAFs may remain silent, co-existing with longevity for years and emerging as a coincidental finding during non-invasive or invasive investigation for the analysis of suspected cardiac disorder.

CAFs are an uncommon congenital anomaly which may be associated with several complications (Table 4). These complications may have coronary vascular, pericardial or myocardial origin, they may have a valvular source or may originate from an atrial or ventricular arrhythmia. Such complications may include myocardial infarction (MI) (4%)[136,137], congestive heart failure (20%)[136], infective endocarditis (reported in 4%-12% in different series)[31,136], atrial[138] and ventricular[139] arrhythmias, aneurysm (reported in 20% of cases)[30,140], rarely ruptured aneurysm with hemopericardium[141] and unruptured aneurysm[139,142], pericardial effusion[143], syncope[142,144] and sudden death[145]. It has been postulated that fistula-related complications increase with age[138]. Secondary PHT is an infrequent complication of congenital CAFs. As early as 1955, Davison reported PHT in patients with CAFs[70].

Most CAFs are small and hemodynamically inconsequential with a negligible left-to-right shunt. However, some can be sizeable and lead to shunting of blood from the coronary circulation to low-pressure pulmonary vascular bed, resulting in PHT[14].

In congenital CAFs, although PHT may occur when sizeable left-to-right shunt exists; in the current review, the mean Qp:Qs was modest, with moderate magnitude 1.9:1.0.

It has been stated that severe PHT is not frequently observed in isolated CAFs[147]. Mild to moderate PHT[145] has sporadically been reported in unilateral[39,45,107,124,146,147] and bilateral fistulas[42,103,112,118]. Indeed, in the current literature review, only 25% were found to have severe PHT, with the majority (75%) having mild or moderate PHT. No reports of multilateral CAFs associated with PHT were found. It is noteworthy that CAFs may be associated with longevity[96] and PHT has been reported in septuagenarians[11] and octogenarians[187].

Although PAP can be measured on Doppler echocardiography, the gold standard for diagnosis is RHC. In the current review, 95% were direct calculation of PAP using RHC and only 5% as an estimate of right ventricular systolic pressure by Doppler echocardiography using TR jet velocity based on the simplified Bernoulli’s equation (Figure 1). It is widely accepted that pulmonary artery systolic pressure (sPAP) can be considered normal until 40 mmHg in the elderly and obese subjects. Moreover, tricuspid regurgitant jet velocity is a parameter that has been widely applied to estimate sPAP[22].

In comparison with the Caucasian group of patients (65%) with PHT, female gender accounted for 80% in the Asian group and was almost equally associated (35% vs 33%) with concomitant congenital and acquired coronary and valvular heart defects.

In the total group of patients (n = 49) with PHT, female gender accounted for (69%), unilateral fistulas was present in (76%) and mild to moderate PHT (75%) was predominant. RHC was performed in 88% of patients and in 12% Doppler echocardiography was used for estimation of the sPAP. Coronary vascular fistulas as a mode of termination were found in the overwhelming majority (76%) of patients. SL was performed in 61% of
patients with PHT.

In the present review of all 49 subjects, possible common features of CAFs associated with PHT were unilateral fistula (37/49 = 76%) originating from the left coronary artery (30/49 = 61%) with a vascular termination (76%) into the right heart side (45/49 = 92%). These findings have to be investigated in a future international survey or prospective study.

A significant difference was noted in the percentages of coronary-cameral fistulas between Asian (40%) and Caucasian (18%) groups of patients with PHT. There was no difference in associated cardiac defects, congenital or acquired, in both the Asian and Caucasian groups (33% and 35%, respectively).

Limitations of the study

Among the Asian population reported by Cheung et al.\(^{[35]}\) in 2001, among the 41 subjects, there were children included in their study. The time span for data collection spread from 1955 to 2014 due to period collection bias.

Publication bias, only subjects with abnormal findings are accepted for publication. Although the data were of high quality and were collected from the world literature, the results of this review are intended to be indicative and require cautious interpretation.

It is clear that more research and studies are warranted for the identification and registration of congenital CAFs associated with PHT; the cause seems to be more multi-factorial (gender, fistula origin and outflow) and dependent on the fistula characteristics itself. We are encouraged to initiate an international survey on CAFs (Euro-CAF.care).

In conclusion, among the whole population, 23% were found to have elevated PAP. In the Asian group of patients 14% demonstrated PHT compared to 34% among the Caucasian group. Among the patients (n = 49) with PHT, 69% were female. The majority of fistulas (76%) in patients (n = 49) with PHT were of CVFs type in contrast to CCFs who accounted for 24% of subjects. The likelihood for a CAF patient to develop PHT is presented when possessing the following features, with a Caucasian female having a fistula with a vascular mode of termination. The findings of this review need to be confirmed in a larger multicenter international registry, preferably with a longer follow-up.

### Table 3 Mode of termination coronary-vascular fistulas vs coronary-cameral fistulas in the pulmonary hypertension (n = 49) and all reviewed (n = 211) subjects

| Mode of termination | CVFs     | CCFs     | Mean age and range (yr) |
|---------------------|----------|----------|-------------------------|
| Total n = 211       | 90/211 (43%) | 121/211 (57%) | 38.3 (26-67) |
| Asian 15/111 (14%)  | 9/15 (60%)  | 6/15 (40%)  | 39.7 (27-67) |
| Caucasian 34/100 (34%)| 28/34 (82%) | 6/34 (18%) | 36.8 (26-60) |

CCFs: Coronary-cameral fistulas; CVFs: Coronary-vascular fistulas.

### Table 4 Possible complications of coronary artery fistulas

| Complication                  | Features                                    |
|-------------------------------|---------------------------------------------|
| Cardiovascular                | Myocardial infarction, stroke, aneurysm, rupture |
| Infectious                    | Bacterial endocarditis, septic pulmonary and septic renal embolism |
| Valvular                      | Incompetence, dysfunction, perforation       |
| Pericardial                   | Hemopericardium, pericardial effusion, tamponade |
| Myocardial                    | Congestive heart failure                     |
| Arrhythmic                    | Supraventricular arrhythmias, ventricular arrhythmias and sudden death |

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### COMMENTS

**Background**

Congenital coronary artery fistulas (CAFs) are uncommon anomalies. Most CAFs are small and hemodynamically inconsequential with a negligible shunt. However, some can be sizeable and lead to shunting of blood from the coronary circulation to low-pressure pulmonary vascular bed, resulting in pulmonary hypertension (PHT).

**Research frontiers**

CAFs may be associated with normal pulmonary artery pressure (PAP) in unilateral or bilateral fistulas, or may sometimes be accompanied with elevated PAP. Rarely, in octogenarians with bilateral CAFs, PAP may remain normal.

**Innovations and breakthroughs**

The likelihood for a CAF patient to develop PHT is presented when possessing the following features, with a Caucasian female having a fistula with a vascular mode of termination.

**Applications**

The findings of this research need to be confirmed in a larger multicenter international registry, preferably with a longer follow-up.

**Peer-review**

This paper is interesting review concerning association PAH and CAF. Therefore, this article should be published.

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October 26, 2016 | Volume 8 | Issue 10

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