Introduction

Congenital coronary artery anomalies are usually incidental findings with a prevalence of 0.21-5.79% depending on study population, applied diagnostic methods and definitions. In 3-36% of cases those anomalies are associated with other congenital cardiovascular heart diseases. Even though congenital coronary artery abnormalities in the vast majority are asymptomatic, some of them might be clinically significant and demand treatment, either conservative or interventional.

Aim

The aim of the study was to assess the prevalence and anatomic characteristics of the selected congenital coronary artery anomalies: 1) single coronary artery (SCA), 2) anomalous coronary artery from the pulmonary artery (ACAPA), and 3) coronary artery fistula (CAF) in adults based on tomographic assessment and their clinical significance.

Methods

Between 2008 and 2020 45,817 coronary computed tomography (CT) examinations in 39,066 subjects were performed in a single high-volume hospital (National Institute of Cardiology, Warsaw, Poland). To select patients, the electronic database of all coronary CT reports was manually screened with the use of specific keywords. All selected CT reports were revalued with applied definitions as follows: 1) SCA – single coronary artery trunk originating from the right or left sinus of Valsalva and dividing further into the branches corresponding to three major coronary arteries: the right coronary artery (RCA), the left anterior descending (LAD) and the circumflex artery (Cx), supplying blood to the whole myocardium; 2) ACAPA – anomalous coronary artery from the pulmonary artery; and 3) CAF – an anomalous direct connection between $\geq$1 coronary arteries and a cardiac chamber or a vessel. Based on the electronic database all demographic and clinical data were collected. All information about current clinical conditions were obtained based on a telephone survey.

The presence of atherosclerotic lesions in coronary arteries was assessed in all patients in accordance with a dedicated classification for the assessment of atherosclerosis in CT, CAD-RADS Coronary Artery Disease – Reporting and Data System.

Coronary CT examinations of the patients with SCA were evaluated based on: 1) site of origin of SCA, 2) anatomical course of SCA according to Lipton's classification, including the site of origin from the sinus of Valsalva ("R" – right or "L" – left) and the course of SCA in relation to the ascending aorta and pulmonary artery (PA) ("A" – anterior to PA, "B" – between

PA and the ascending aorta, "P" – posterior to the ascending aorta). In group I the anatomical course of SCA corresponds to the normal course of RCA or the left coronary artery (LCA) (RII, LII). In group II SCA originating from the right or left sinus of Valsalva, crosses the base of the heart to access the proximity of the anatomically normal course (RII-A, RII-B, RII-P, LII-A, LII-B, LII-P). In group III RCA arising from the right sinus of Valsalva shares a common trunk with LAD and Cx (RIII). Within the SCA trunk length, minimal and maximal lumen area (LA) and lumen diameter (LD) were measured and compared with the measurements of the LCA trunk and proximal segment of RCA in the reference group with normal coronary artery anatomy and without atherosclerosis (n=199).

Coronary CT examinations of the patients with ACAPA were evaluated based on: 1) coronary artery originating from PA (right or left), 2) site of origin of coronary artery from PA, 3) presence of collateral circulation: collaterals between RCA and LCA, dilated septal vessels, dilated bronchial arteries, 4) presence of coronary steal phenomenon.

All CAFs were evaluated based on their site of origin and termination, and morphology: 1) number of CAFs, 2) complexity, 3) size, 4) tortuosity, 5) intramuscular course and 6) presence of aneurysms, calcifications, vegetations, thrombus in CAF or dissection of CAF. The minimal and maximal LD and LA of CAFs were measured at the site of origin and termination, as well as at the narrowest and widest segment of CAFs. CAF was defined as clinically significant if during follow-up, the patient required hospitalization and/or surgical treatment related to the confirmed congenital anomaly of the coronary arteries, or if it was the cause of myocardial infarction, infective endocarditis (IE), heart failure, or death.

Results

Among 39,066 patients SCA was diagnosed in 27 (0.069%), ACAPA in 6 (0.015%), and CAFs in 42 (0.1%) subjects. The mean age of the patients at which the CT examination was performed in the SCA group was 52.6 ± 19.5 years, in ACAPA group 33.2 ± 16.5 years, and in the CAF group 57.5 ± 13.8 years.

Single coronary artery

The most common site of origin of SCA was the right sinus of Valsalva (18/26; 69.2%), in 8 patients SCA was originated from the left sinus of Valsalva (8/26; 30.8%), whereas in 1 subject the sinus of Valsalva could not have been determined because of the presence of the common arterial trunk. According to Lipton's classification 26 patients were evaluated. Three types of SCA were predominantly observed: RII-A, RII-B and LI. A potentially malignant

course of SCA between PA and the aorta (type II-B) was detected in 8 subjects, in whom no impact of the anomaly on patients' clinical condition was observed. Congenital cardiovascular diseases coexisted in 10 patients with SCA (37.0%), who were younger and less frequently were diagnosed with diabetes and dyslipidemia ($p<0.001$, $0=0.026$, $p=0.018$, respectively). There was no correlation between the site of origin of SCA and its course and coexistence of congenital cardiovascular abnormalities. Moreover, no correlation between the localization of the site of origin, the interarterial course of SCA (type II-B) and the presence along with severity of atherosclerosis was seen. No statistically significant differences between the measurements of SCA trunks arising either from the right or left sinus of Valsalva were noticed.

It has been observed that the anatomy of the SCA trunks significantly differs from the anatomy of the trunks of normal coronary arteries. Compared to the reference group, the trunks of the patients with SCA were significantly shorter and had larger LD and LA ($p<0.001$, $p<0.001$, $p<0.001$, respectively). For the overall SCA group, LA of the proximal segment of SCA trunk was larger than the sum of LA of the LCA trunk and the proximal segment of RCA in the control group.

During the observation period (the mean observation time 62 ± 41 months) 5 patients underwent surgical interventions because of concomitant congenital heart diseases, and 2 patients with complex congenital cardiovascular diseases died.

Anomalous coronary artery from the pulmonary artery

The prevalence of ACAPA in the study group was 0.015% (6/39,066) of which: 1) anomalous left coronary artery from the pulmonary artery (ALCAPA) = 0.0077% (3/39,066); 2) anomalous right coronary artery from the pulmonary artery (ARCAPA) = 0.0051% (2/39,066); 3) anomalous origin of a single coronary artery from the pulmonary artery (ASCAPA) = 0.0026% (1/39,066), respectively. ACAPA was often present in women (5/6 subjects). Four of 6 patients (66.7%) required invasive treatment. Of the 5 female patients 2 gave birth twice.

In 3 patients (2 with ALCAPA, 1 with ARCAPA) numerous septal collaterals in ventricular septum were seen. The presence of numerous collaterals between RCA and LCA was observed in 2 patients (1 with ALCAPA, 1 with ARCAPA) as well as dilated bronchial arteries (1 with ALCAPA, 1 with ASCAPA).

The mean follow-up period was 76 ± 48 months with 100% survivability.

Coronary artery fistulas

The prevalence of CAFs was 0.1% (42/39,066). Forty-two patients with 56 CAFs were identified. Clinically significant CAFs were diagnosed in 7 of 42 patients (16.7%), of whom 3 died (42.9%) within the follow-up period. Clinically significant CAFs were more often detected in younger ($p=0.028$) and male patients ($p=0.041$). In addition, pulmonary hypertension was more frequently diagnosed in patients with clinically significant CAFs ($p=0.026$). The measurements of clinically significant CAFs were substantially larger at the site of origin and the widest segment of CAF ($p<0.05$). What is more, calcifications were more often seen in clinically significant CAFs ($p=0.003$).

The most common site of origin of CAFs was RCA ($p=0.021$). The site of origin did not have any impact on demographic characteristics, clinical presentation, or CAF morphology. Additionally, there was no significant difference between the site of origin and size of CAF.

CAFs were predominantly draining into the pulmonary artery (PA) (58.9%) ($p=0.023$). IE was often diagnosed in patients with CAFs draining into the right-side cardiac structures ($p=0.006$). CAFs terminating into PA as well as into the right and left pulmonary arteries were more often multiple, complex, bilateral, and tortuous ($p=0.001$, $p<0.001$, $p<0.001$, $p=0.001$, respectively). Among 42 patients, single CAF was found in 30 (71.4%), while in 28.6% of the cases multiple CAFs were revealed.

Atherosclerosis was detected in 22/42 (52.4%) subjects with CAFs, of whom in 6 (27.2%) lumen narrowing was $\geq$ 70%. No correlation between the site of origin as well as termination and the presence of atherosclerotic lesions was observed. In patients with atherosclerotic lumen narrowing $\geq$50% ($\geq$ CAD-RADS 3) CAFs significantly often were smaller ($p=0.021$).

Large CAFs (>10 mm) were not only more often found in younger patients, with pulmonary hypertension and/or IE, but also required surgical or percutaneous closure ($p=0.026$, $p=0.027$, $p=0.004$, $p=0.044$, respectively). Furthermore, CAFs >10 mm were often draining into the coronary sinus, as well as calcification was more frequently detected within their walls ($p<0.001$, $p<0.001$, respectively). However, CAFs <10 mm were often multiple and complex ($p=0.0014$, $p=0.012$, respectively).

Follow-up was available for 36 of 42 patients (85.7%) with CAFs, of whom 7 died (19.4%) during the observation period. The mean observation time was 52 ± 51 months.

Conclusions

Single coronary artery

1. Single coronary artery is a rare anomaly with the right sinus of Valsalva as the most common site of its origin, often coexisting with congenital cardiovascular diseases.
2. The trunk of single coronary artery is significantly shorter and wider compared to the trunk of the normal left coronary artery.
3. Single coronary artery is practically a clinically silent anomaly.

Anomalous coronary artery from the pulmonary artery

1. Anomalous coronary artery from the pulmonary artery is an extremely rare anomaly in adults.
2. The most common type is the left coronary artery anomalously originating from the pulmonary artery.
3. The anomaly is characterized by a usually severe clinical course and requires invasive treatment. However, even in females with uncorrected anomalous left coronary artery originating from the pulmonary trunk the uncomplicated pregnancy is possible.

Coronary artery fistulas

1. Coronary artery fistulas are the most common among chosen congenital coronary artery anomalies.
2. Clinically significant coronary artery fistulas are larger, simple, more often draining into the right-sided cardiac structures and are more frequently detected in younger and male patients.
3. The most common site of origin of coronary artery fistulas is the right coronary artery, while the most frequent site of their termination is the pulmonary artery.
4. Large coronary artery fistulas usually are single with calcifications within their walls and terminate in the coronary sinus.
5. Coronary artery fistulas draining into the pulmonary trunk are often multiple, tortuous, and complex.

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