

# Frequency, origins and courses of anomalous coronary arteries in 607 Turkish children with tetralogy of Fallot

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## Abstract

**Background:** *The aim of this study was to investigate the incidence, origins and courses of coronary artery anomalies using a combination of angiographic and surgical methods in Turkish children with tetralogy of Fallot (ToF).*

**Methods:** *Seventy-seven patients in whom coronary artery anomalies had been identified by angiography and/or at operation out of 549 ToF and 58 Fallot-type double outlet right ventricle (total 607) patients, were enrolled in the study.*

**Results:** *Coronary artery anomalies were identified in 12.7% of the patients. The incidence was 12.2% (67/549) in patients with aortic overriding  $\leq 50\%$ , and 17.2% (10/58) with aortic overriding  $> 50\%$  ( $p > 0.05$ ). The incidence of anomalous coronary arteries crossing the right ventricular outflow tract (RVOT) was 7.91%. The commonest anomaly was the left anterior descending artery (LAD) or accessory LAD arising from the right coronary artery (RCA;  $n = 25$ ). Other frequent anomalies were single coronary ostium ( $n = 21$ ) and enlarged conal branch of RCA ( $n = 18$ ). In 62.3% (48/77) of the patients with a coronary anomaly, the anomalous vessels were crossing the RVOT. The ratio of crossing the RVOT was 92.0% for LAD arising from the RCA, 66.7% for conal branch, and 42.9% for single coronary ostium.*

**Conclusions:** *Two thirds of the anomalous coronary arteries were crossing the RVOT, and had surgical importance. The most frequent coronary artery anomaly that crossed the RVOT was the LAD or the accessory LAD arising from the RCA. Also, an enlarged conus artery should be considered as an anomaly because of its surgical importance, given its high rate of crossing the RVOT. (Cardiol J 2011; 18, 5: 546–551)*

**Key words:** coronary variations, angiography, incidence, childhood

## Introduction

Tetralogy of Fallot (ToF) is one of the commonest cyanotic congenital heart diseases in childhood [1]. In angiographic, surgical, and autopsy series, coronary artery anomalies (CAA) have been reported in 2% to 14% of patients with ToF [2]. Anomalies

in the origin and course of the coronary arteries involving the anterior wall of the right ventricle are highly significant. These anomalies, if not detected pre-operatively, can lead to major problems at operation [3, 4]. In some cases, the anomalous coronary artery may be undetectable intraoperatively, especially when it is obscured by the overlying myo-

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Received: 18.02.2011

Accepted: 17.05.2011

cardium [5–8], by epicardial fat [8, 9], or by epicardial-pericardial adhesions [10, 11] due to previous palliative surgery. Pre-operative recognition of such arteries is therefore crucial in deciding the time and type of procedure to be performed [12].

The aim of this study was to investigate the incidence of abnormalities of the origin and course of the coronary arteries by a combination of angiographic and surgical methods in ToF and Fallot-type double outlet right ventricle (DORV) patients, and to ascertain the surgical importance of each coronary artery anomaly.

## Methods

Seventy-seven patients in whom CAA were identified by angiography and/or at operation out of 549 ToF and 58 Fallot-type DORV (total 607) patients in Baskent University Hospital between 1989 and 2003 were enrolled in the study. Fifty-one (66.2%) were male and 26 (33.8%) female; mean age was  $3.9 \pm 2.9$  years (4.5 months – 17 years) and mean body weight was  $15.5 \pm 9.6$  kg (3.4–54 kg).

In order to identify the origins and courses of the coronary arteries, we analyzed retrospectively the cineangiograms and the surgery reports of all patients in whom CAA were determined by angiography and/or at operation. All abnormalities of the origin and course of the coronary arteries were enrolled in the study. Also, an enlarged conal branch of the right coronary artery (RCA) was considered as an anomaly when it was remarkably developed, enlarged, and ran towards the right ventricular outflow tract (RVOT).

Angiograms were performed in Baskent University Hospital using the Siemens Bicor Top System and interpreted by two experienced pediatric cardiologists simultaneously. All surgical procedures were performed by two surgeons and their team in the same hospital.

Ages and body weights were presented as mean  $\pm$  standard deviation and range. For statistical analysis, we used the SPSS computer package (SPSS Inc., Chicago, USA). Chi-square test was used for analyzing the statistical significance of difference between two groups. A p value of less than 0.05 was considered as significant.

## Results

Coronary artery anomalies were identified in 77 of the 607 (12.7%) patients. The incidence was 12.2% (67/549) in patients with aortic overriding  $\leq 50\%$ , and 17.2% (10/58) in patients with Fallot-

-type DORV in whom aortic overriding  $> 50\%$ ; the difference between the two groups was not statistically significant ( $p = 0.37$ ).

In 62.3% (48/77) of the patients, the anomalous coronary arteries were crossing the RVOT. When only the anomalous vessels crossing the RVOT were considered, the incidence of coronary anomaly was 7.91% (48/607).

Anomalous coronary arteries and their rate of crossing the RVOT are set out in Table 1.

### Left anterior descending coronary artery arising from the right coronary artery or the right sinus of Valsalva

There were 25 (32.5%) patients in this group. In 12 (15.6%) cases, the left anterior descending artery (LAD) arose from the proximal portion of the RCA and all (100%) of them reached the anterior interventricular groove passing across the outflow tract of the right ventricle.

Twelve (15.6%) patients had two LADs. In these cases, an accessory LAD arose from the RCA. Ten (83.3%) of these accessory vessels reached the anterior interventricular groove crossing the RVOT and coursed parallel to the normal LAD. In the other two cases, the vessels reached the anterior interventricular groove without crossing the RVOT.

One (1.3%) patient had an anomalous origin of the LAD from the right sinus of Valsalva. In this case, the LAD crossed the outflow tract of the right ventricle.

In all cases in this group, the circumflex artery had a normal course, originating from the left sinus of Valsalva. In 23 (92.0%) of the 25 patients in this group, an anomalous coronary artery crossed the RVOT.

### Single coronary ostium

We defined single coronary ostium in 21 (27.3%) patients. In 16 (20.8%) cases, the coronary arteries were arising from the left sinus of Valsalva (LSV) with a single ostium. In these cases, the RCA arose from the proximal portion of the left coronary artery. The RCA passed behind the aorta in nine (56.3%) patients, and in seven (43.8%) passed in front of the aorta and crossed the outflow tract of the right ventricle.

Single right coronary ostium was seen in three (3.9%) patients. In one of these patients, the LAD passed across the RVOT.

In two (2.6%) cases, the coronary arteries arose from the non-coronary sinus of Valsalva, and one of these crossed the RVOT.

In nine (42.9%) of the 21 patients with single coronary ostium, there was a coronary artery crossing the outflow tract of the right ventricle.

**Table 1.** Incidences of coronary artery anomalies.

Coronary artery anomalies	CAA n (rate)	RVOT(+) n	RVOT(+)/CAA (rate)
LAD from RCA or RSV:	25 (32.5%)	23	23/25 (92.0%)
LAD from RCA	12 (15.6%)	12	12/12 (100%)
Accessory LAD from RCA	12 (15.6%)	10	10/12 (83.3%)
LAD from RSV	1 (1.3%)	1	1/1 (100%)
SCO:	21 (27.3%)	9	9/21 (42.9%)
SCO from LSV	16 (20.8%)	7	7/16 (43.8%)
SCO from RSV	3 (3.9%)	1	1/3 (33.3%)
SCO from NCSV	2 (2.6%)	1	1/2 (50.0%)
Enlarged conal branch of RCA:	18 (23.4%)	12	12/18 (66.7%)
Single conal branch	15 (19.5%)	9	9/15 (60.0%)
Double conal branch	3 (3.9%)	3	3/3 (100%)
RCA from LAD or LSV:	7 (9.1%)	3	3/7 (42.9%)
RCA from LSV	4 (5.2%)	0	0
RCA from LAD	3 (3.9%)	3	3/3 (100%)
Other CAA:	6 (7.8%)	1	1/6 (16.7%)
Coronary artery fistula	2 (2.6%)	0	0
RV branch from LAD	1 (1.3%)	1	1/1 (100%)
Narrowness of distance between LAD and RCA	1 (1.3%)	0	0
LCx from RCA	1 (1.3%)	0	0
LCx from RSV	1 (1.3%)	0	0
Total	77 (100%)	48	48/77 (62.3%)

CAA — coronary artery anomalies; RVOT(+) — CAA crossing the right ventricular outflow tract; LAD — left anterior descending artery; RCA — right coronary artery; RSV — right sinus of Valsalva; SCO — single coronary ostium; LSV — left sinus of Valsalva; NCSV — non-coronary sinus of Valsalva; RV — right ventricle; LCx — left circumflex artery

### Enlarged conal branch of the right coronary artery

In 18 (23.4%) patients, the conus artery arising from the proximal portion of the RCA was remarkably developed, enlarged and ran towards the RVOT. In 12 (66.7%) of the cases, these arteries crossed the RVOT. In 15 (19.5%) cases, one enlarged conus artery was seen, and in three (3.9%) cases, two enlarged conus arteries were seen. In nine of the cases with one, and in all of the cases with two, these arteries were crossing the outflow tract of the right ventricle.

### Right coronary artery arising from left anterior descending artery or left sinus of Valsalva

There were seven (9.1%) patients in this group. The RCA was arising from the LSV in four (5.2%), and from the LAD in three (3.9%) cases. In all cases with the RCA arising from the LSV, passing behind the aorta, the RCAs were not crossing the RVOT. However, all of the RCAs arising from the LAD were passing over the RVOT. In three (42.9%) of the seven patients in this group, the RCAs were crossing the RVOT.

### Circumflex artery arising from right coronary artery or right sinus of Valsalva

In one (1.3%) case, circumflex artery originated from the right sinus of Valsalva, and in another (1.3%) case it arose from the RCA. Both of these were passing behind the aorta.

### Other coronary artery anomalies

In one (1.3%) patient, there was a right ventricular branch arising from the LAD and crossing the RVOT. In one (1.3%) case, the distance between LAD and RCA was narrow. Coronary fistulas were seen in two (2.6%) cases. In one of these cases, the circumflex artery was connected to the left ventricle apex. In the other case, the RCA was connected to the apex of the right ventricle.

### Discussion

Although an anomalous coronary artery in itself may cause no recognized problems, it contributes significantly to morbidity and mortality rates during complete cardiac repair [13]. Any coronary artery that crosses the RVOT affects the type and

timing of surgical repair in patients with ToF [14–17]. Coronary vessels that would interfere with right ventriculotomy or resection of outflow obstruction must be identified pre-operatively, partly because a sternotomy may not be the approach of choice for palliation, and partly because the coronary arteries may be obscured by previous palliative surgery, or may pursue an intramural course [15].

The incidence of CAA varies according to the methods used by investigators. In angiographic [4, 12, 15, 18–21], surgical [10, 11, 13, 22] and autopsy [7, 23] series, coronary artery abnormalities have been reported in 2% to 14% of patients with ToF. In our study, we determined the coronary artery anomaly incidence as 12.7% overall using a combination of angiographic and surgical methods. The ratio was 12.2% in patients with aortic overriding  $\leq 50\%$ , and 17.2% in patients with Fallot-type DORV in whom aortic overriding was  $> 50\%$ ; the difference between the two groups was not statistically significant. Some studies have reported that the incidence of the LAD or the accessory LAD arising from the RCA and single coronary artery is increased according to the degree of aortic overriding, and it has been recommended that physicians be suspicious of CAA in all cases with DORV [24]. Since we included all abnormalities of the origin and course of the coronary arteries determined in the angiocardiograms and/or at operation, the incidence of coronary artery anomaly was high (12.7%) in our study. The presence of enlarged conal branch of RCA, as in our study, has been found to increase remarkably the incidence of anomaly in all studies where it has been included [4, 19].

The low incidence of coronary anomalies in ToF reported by surgical teams can be explained by the fact that many of these anomalies are difficult to detect intraoperatively [4, 5, 10, 11, 25]. Coronary artery distribution is not always definable by surgical observation. The anomalous vessel can have either an intramyocardial course, or, where there has been previous surgery involving pericardiotomy, may be obscured by epicardial adhesions [23]. Also, surgical teams have a tendency to enroll only surgically important coronary anomalies in their study. In our series, where only the anomalous vessels crossing the RVOT were considered, the incidence of coronary anomaly was 7.91%.

Post mortem examinations indicate a higher incidence, probably due to a greater possibility of carefully examining the coronary arteries [4, 6, 7]. In angiographic studies, the incidence is varied by the use of ventriculography, aortography or selective coronary angiography, as well as the positions

of the angiograms and their interpretations [4, 12, 18–20, 26, 27].

In our study, the commonest CAA, the LAD or the accessory LAD arising from the RCA or the right sinus of Valsalva, were seen in 32.5% of the patients with coronary anomalies. The incidence of these anomalies was 4.1% in all patients with ToF and Fallot-type DORV. In 23 out of 25 patients, the anomalous vessel reached the anterior interventricular groove passing across the outflow tract of the right ventricle. Our data was compatible with most previous studies, such as those by Fellows et al. (4%), Dabizzi et al. (4.9%) and Brizard et al. (3.4%) [4, 19, 20, 23, 26, 28].

The second commonest coronary artery anomaly in our study was single coronary ostium from one of the sinuses of Valsalva, as has been the case in most studies. In this situation, the anomalous vessels may cross over the proposed infundibular resection or travel around the RVOT and cause no surgical problems [13]. The incidence of this anomaly was 3.4% in our study. It was reported as 2.7% by Fellows et al. [20], 3.77% by Dabizzi et al. [19] and 1.5% by Brizard et al. [28]. We observed 21 (27.3%) single coronary ostium in the patients with coronary anomalies; 16 from the left, three from the right, and two from the non-coronary sinus of Valsalva. In nine (42.9%) of these patients, there was a vessel crossing the RVOT.

In 40% of patients with ToF, a large and remarkably long conus artery arising from the RCA supplied an important myocardial area on the subpulmonary infundibulum [7, 12]. Although a large conal branch of the RCA was not regarded in most previous studies to be an anomaly, when it is enlarged and courses over the right ventricular infundibulum it can present a serious hazard if unrecognized at the time of right ventriculotomy [4, 19]. We have taken an enlarged conus artery to be an anomaly because of its surgical importance. In 23.4% of our patients, the conus artery arising from the RCA was remarkably developed, enlarged and ran towards the RVOT. Twelve (66.7%) of these cases had surgical importance i.e. were crossing the RVOT.

In this study, although all of the RCAs arising from the LAD were passing over the RVOT, none of the RCAs arising from the LSV crossed the RVOT, because of passing behind the aorta. Other coronary anomalies we have defined were the circumflex artery arising from the RCA or the right sinus of Valsalva (two cases), the right ventricular branch arising from the LAD (one case), narrowness of the distance between the LAD and the RCA (one case), and coronary artery fistulas (two cases).

Small fistulas between coronary arteries and pulmonary arteries or right atrium in ToF have been reported in a study with selective coronary angiography [19]. Apart from these anomalies, some authors have reported rare cases such as the left coronary artery or the RCA arising from the pulmonary artery [29–31]. However, we did not encounter such an anomaly in our series. Also, in recent years, besides angiography, new imaging methods such as magnetic resonance imaging and especially multislice computed tomography have been used successfully to demonstrate coronary arterial anomalies [32, 33].

### Conclusions

1. The factors affecting the differences between reported coronary artery anomaly incidences were:
  - differences of the study groups as angiographic, surgical or autopsy series;
  - including or excluding the conal branch of the RCA;
  - including only surgically important anomalies, or all abnormalities of the origin or course of the coronary arteries;
  - the methods of angiography used, i.e. ventriculography, aortography or selective coronary angiography, and the position of the angiograms.
2. The incidence of coronary artery anomaly was higher in patients with aortic overriding > 50%, but the difference was not statistically significant.
3. The most frequent CAA were the LAD or the accessory LAD arising from the RCA, the single coronary ostium, and the enlarged conal branch of the RCA.
4. The most frequent coronary artery anomaly that crossed the RVOT (making it surgically important) was also the LAD or the accessory LAD arising from the RCA. The next most frequent was the enlarged conal branch of the RCA, followed by the single coronary ostium.
5. An enlarged conal branch of the RCA should be considered as an anomaly because of its surgical importance, given its high rate of crossing the RVOT.

### Acknowledgements

The authors thank Sait Aslamaci, MD and Sukru Mercan, MD for their surgical contributions.

The authors do not report any conflict of interest regarding this work.

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