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Tetralogy of Fallot and Associated Coronary Anomalies

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Abstract

Background- Anomalous coronary artery disease is more common among patients with Tetralogy of Fallot (ToF).

Methods- In order to determine the coronary anomalies in patients with ToF, 135 patients (80 males) with ToF, 9 months to 40 years of age were studied over 7 years (1995-2002) at Modarres Hospital in Tehran, Iran.

Results- Eight out of the 135 patients (4 males) with ToF were associated with a surgically-relevant coronary artery anomaly (5.9%). The anomalous coronary arteries consisted of single ostium of the coronary artery (n = 5), the left anterior descending coronary artery originating from the right coronary artery (n = 2) or single right coronary artery (n = 2) and the right coronary artery arising from the left coronary artery (n = 1). Surgical technique employed in three ToF patients was transverse incision on the right ventricle, and commissurotomy via the pulmonary artery was the technique used for three other patients. In another patient, a composite graft between the right ventricle outflow tract and main pulmonary artery was inserted. In the remaining four patients with single ostium coronaries, routine repairs were done.

Conclusions-Anomalous coronary artery disease is more common among patients with ToF. Bearing these anomalies in mind during primary repair could decrease the risk of surgery in patients with ToF ***(Iranian Heart Journal 2004; 5(1,2):39-42).***

Key words: Tetralogy of Fallot ■ coronary artery ■ anomaly

Congenital coronary anomalies are a recognized cause of myocardial ischemia and sudden cardiac death, especially among children and young adults.[1] The prevalence of congenital coronary anomalies has been estimated at approximately 0.85%.[2,3,4] As might be anticipated, anomalous coronary artery disease is more common among patients with other forms of congenital heart disease[5], especially those with Tetralogy of Fallot (ToF), complete transposition of the great arteries, congenitally corrected transposition and bicuspid aortic valves.

Anomalous coronaries typically originate on the contralateral side to their destination (or less commonly from the noncoronary sinus or pulmonary artery) and 'cross' to the other side of the aorta. By far the most common (~50%) anomalous coronary artery condition in the adult is anomalous origin of the left circumflex (LCx) from the right sinus of valsalva or (right coronary artery) RCA.[2,3,6,7] The anomalous

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LCx may arise from a proximal branch of the RCA, or from a separate ostium.[7]
Coronary artery anomalies are encountered in up to 12% of patients with ToF. The most common variants include the (left anterior descending) LAD arising from the RCA or separately from the right sinus of Valsalva, the RCA arising from the (left coronary artery) LCA or LAD, the presence of a large conal branch from the RCA that supplies a portion of the anterior interventricular septum and a single coronary artery.[8]
Many of these variants result in a major coronary artery coursing anterior to the pulmonary outflow tract, which in turn complicates the ultimate surgical repair. In the past, the presence of a prominent artery crossing the infundibulum often led to either initial palliation (using a Blalock–Taussig or central shunt) rather than repair.[8] More recently, two reports regarding this difficult anatomy show that total repair may often be accomplished early in life without the use of a conduit.[9,10]
The objective of this study was to determine the coronary anomalies in patients with ToF.

Patients and Methods

This study was a retrospective study over seven years designed to ascertain the coronary anomalies in patients with tetralogy of Fallot at our department.
One hundred and thirty-five patients with ToF ranging in age from 9 months to 40 years underwent primary repair between 1995 and 2002 in our center.
Data analysis was done using SPSS statistical software package (version 10.0).

Results

The 135 patients with ToF who underwent surgery consisted of 80 men and 55 women (mean age: 8.3 ± 7.2 years). Associated congenital cardiac anomalies were seen in 47 patients (34.8%). Among these associated anomalies, right-sided aortic arch (12 patients) was the most common, followed by atrial septal defect (10 cases), patent ductus arteriosus (9 cases) and left superior vena cava (LSVC) (7 cases).
Eight of the135 patients with ToF (4 males and 4 females) were associated with a surgically-relevant coronary artery anomaly (5.9%). The mean age of these patients at repair was 7.8 ± 3.3 years (range: 4-13 years, Table I).
The anomalous coronary arteries included single ostium for the origin of coronary arteries (n = 5), the left anterior descending coronary artery arising from the right coronary artery (n = 2) and the right coronary artery originating from the left coronary artery (n = 1).

Table I. Characteristics of patients with Tetralogy of Fallot and associated coronary anomalies.

Patients	Age (year)	Sex	Abnormal origin	associated congenital anomalies
P1	7	Female	RCA from LCA	-
P2	9	Female	single ostium	-
P3	9	Male	LAD from RCA	Aneurysm of valsalva sinus
P4	4	Male	single ostium	LSVC
P5	13	Male	single ostium	-
P6	5	Female	LAD from RCA	-
P7	4	Female	single ostium	-
P8	11	Male	single ostium	-

LAD: left anterior descending
RCA: right coronary artery
LCA: left coronary artery
LSVC: left superior vena cava

LAD originated from RCA and involved RVOT (right ventricle outflow tract) in one patient (9-year-old boy). He underwent transverse incision on the right ventricle, resection of the moderator band, repair of ventricular septal defect with Gortex patch and commissurotomy via the pulmonary

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artery (PA). Ventriculotomy was closed with a pericardial patch. In a 9-year-old girl with single ostium coronaries, both arteries originated from a point in front of the aorta. LCA was in front of PA; LAD was branched arround PA for LCx developing. In another girl, 7 years old, RCA came off the left side and covered the infundibulum. The same technique was done for both of them.

In another patient (a 5-year-old girl), LAD originated from RCA; RVOT was opened beneath RCA and LAD and a composite graft was inserted between RVOT and main PA.

The remaining four patients had a single origin of coronaries but not involving the RVOT; as a result, the surgical techniques were routine repairs for them.

There have been no early deaths after surgery.

Associated congenital cardiac anomalies have been seen in 2 of 8 ToF patients with coronary artery anomaly: the aneurysm of sinus of valsalva in a 9-year-old boy with LAD from RCA, and the LSVC in a 4-year-old boy with single ostium of coronary arteries.

Discussion

In this study, 135 patients with ToF were operated on, 5.9% of whom had coronary artery anomaly. The prevalence of congenital coronary anomalies has been estimated at approximately 0.85% in other studies.[2,3,4] Our study shows that anomalous coronary artery disease is more common among patients with ToF. This data is in agreement with that in the study of Dabizzi, et al. However, the prevalence of these anomalies have been reported in up to 12% of patients with ToF.[8] The same rate of 5.9% is also reported in the study by Brizard, et al. on 611 consecutive repairs.[9] Also, aberrant origins of the coronary artery were found in 14% of 103 specimens with ToF in the study of Li, et al.[11]

Recently, the ability to repair ToF early in life, regardless of the anatomy of the coronary artery, has largely been the reason for many academic centers to no longer perform routine coronary artery angiography prior to surgery.[8]

In our study, anomalous coronary arteries were single ostium for coronary arteries (50%), LAD from the RCA (37.5%) and the RCA from the LCA (12.5%). In a study by Brizard, et al., anomalies included LAD from RCA or single RCA (61.1%), RCA from LCA or LAD (22.2%) and large RCA conal branch (16.6%).[9] In addition, in other studies, the most common variants of coronary arteries have been LAD arising from the RCA or separately from the right sinus of valsalva, the RCA arising from the LCA or LAD, the presence of a large conal branch from the RCA and a single coronary artery.[8]

Abnormal coronary arteries have been found in some studies to be a risk factor for surgically-related mortality.[13] While some of this risk can be assigned to the anomaly, often the relationship between the origin(s) of the pulmonary artery and coronary arteries is such that the coronaries are placed ‘at risk’ as a result of surgical repair.[13]

Taking account of these anomalies during primary repair could decrease the risk of surgery in patients with ToF.

In our study, surgical techniques in three ToF patients were transverse incision on the right ventricle, repair of ventricular septal defect and commissurotomy via PA in three patients (one single ostium coronaries, one LAD from the RCA and one RCA from the LCA). In another patient with LAD from the RCA, a composite graft between RVOT and main PA was inserted. In the remaining four patients with single ostium coronaries,

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routine repairs were done. There have been no early deaths.

Conclusion

Anomalous coronary artery disease is more common among patients with ToF. Taking these anomalies into account during primary repair could decrease the risk of surgery in patients with ToF. Primary repair of tetralogy of Fallot with anomalous coronary arteries can be done with excellent results.[10]

References

1. Cheitlin MD, De Castro MD, McAllister HA: Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva: a not-so-minor congenital anomaly. Circulation 1974; 50: 780–787.

2. Engel HJ, Torres C, Page HL: Major variations in anatomical origin of the coronary arteries: angiographic observations in 4250 patients without associated congenital heart disease. Cathet Cardiovasc Diagn 1975; 1: 157–169.

3. Kimbiris D, Iskandrian AS, Segal BL, Bermis CE: Anomalous aortic origin of coronary arteries. Circulation 1978; 58: 606–615.

4. Chaitman BR, Lesperance J, Saltiel J, Bourassa MG : Clinical angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. Circulation 1976; 53: 122–131.

5. Dabizzi RP, Teodori G, Barletta GA, Caprioli G, Baldrighi G, Baldrighi V: Associated coronary and cardiac anomalies in the tetralogy of Fallot. An angiographic study. Eur Heart J 1990; 11: 692–704.

6. Leberthson RR, Dinsmore RE, Bharati S, Rubenstein JJ, Caulfield J, Wheeler EO, et al: Aberrant coronary artery origin from the aorta: diagnosis and clinical significance. Circulation 1974; 50: 774–779.

7. Page HL Jr, Engel HJ, Campbell WB, Thomas CS Jr: Anomalous origin of the left circumflex coronary artery. Circulation 1974; 50: 768–773.

8. Burch GH, Sahn AB, David J: Congenital coronary artery anomalies: the pediatric perspective. Coronary Artery Disease. 2001; 12(8): 605-616.

9. Brizzard CPR, Mas C, Sohn YS, Cochrane AD, Karl TR. Transatrial–transpulmonary tetralogy of Fallot repair is effective in the presence of anomalous coronary arteries. J Thorac Cardiovasc Surg 1998; (116): 770–779.

10. Tchervenkov CI, Pelletier MP, Shum-Tim D, Beland MJ, Rohlicek C: Primary repair minimizing the use of conduits in neonates and infants with tetralogy or double-outlet right ventricle and anomalous coronary arteries. J Thorac Cardiovasc Surg 2000; 119: 314–323.

11. Li JS, Carvalho JS, Ho SY: Coronary arterial anatomy in tetralogy of Fallot: morphological and clinical correlations. Heart 1998; 80(2): 174-183.

12. Hanley FL, Heinemann MK, Jonas RA, Mayer JE Jr, Cook NR, Wessel DL: Repair of truncus arteriosus in the neonate. J Thorac Cardiovasc Surg 1993;105:1047–1056.

13. Bogers AJJC, Bartelings MM, Bokenkamp R, Stijnen T, van Suylen RJ, Poelmann RE, et al: Common arterial trunk, uncommon coronary arterial anatomy. J Thorac Cardiovasc Surg 1993;106:1133–1137.

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