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Additional Pathologies in Tetralogy of Fallot*

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Tetralogy of Fallot(TOF) is often associated with additional anomalies. These additional anomalies may worsen the patients' prognosis by causing deterioration of the clinical status during the preoperative period, prolongation of the duration of cardiopulmonary bypass and the frequent occurance of postoperative complications. Additional pathologies encountered in our series of 380 cases of TOF undergone total correction until June 1979 form the basis of this paper.

Clinical Presentation: Associated anomalies encountered in 380 cases of TOF are tabulated in Table 1. We also encountered poor physiological condition of the right ventricular outflow tract despite adequate anatomical relief of infundibular stenosis, which we called "spastic outflow tract" in 8 cases, causing high right ventricular pressure immediately after total correction.[1]

Discussion

Different kinds of congenital cardiovascular malformations may be present in tetralogy of Fallot (TOF). Goor and Lillehei[2] stated that the most important associated anomalies are right aortic arcus (in 25 % of the cases) and concomitant aberrant left subclavian artery, patent ductus arteriosus (PDA), atrial septal defect (ASD), coronary artery

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malformations, anomalies of the systemic and pulmonary venous return, pulmonary arterial branchial stenosis, intraarteriolar thrombi and complete absence of the pulmonary leaflets. Right aortic arcus and left aberrant subclavian artery are of importance in palliative shunting operations.[2] Chiariello et al.[3] tabulated the associated lesions in 403 cases and found that the ASD is the most common additional lesion in TOF (24 %), followed by pulmonary atresia (7 %), left persistant superior vena cava (LPSVC) (4.5 %), coronary artery anomalies (4 %), PDA (2.5 %) and double outlet right ventricle (2 %) in order of frequency. Cleland et al.[4] mentioned that right sided aortic arch, patent foramen ovale (PFO), ASD, peripheral pulmonary artery stenosis, absent left pulmonary artery, aortic valvar disease and pulmonary valvar regurgitation are the commonly found associated anomalies in TOF. Aortic valvar disease may consist of valvar or subvalvar stenosis, or aortic regurgitation. Aortic regurgitation may be due to the congenital prolapsus of the aortic cusps or subacute bacterial endocarditis.[3,4]

Pulmonary valvar regurgitation may result from either the infective endocarditis or the absence of the pulmonary valve.[4,5] Tricuspid pathology can also be encountered as the result of bacterial endocarditis or a congenital malformation.[4] Congenital mitral valve anomalies like stenosis or cleft mitral valve are also present in rare cases.[3] Other seldomly found additional anomalies in TOF have also been described like dextrocardia, corrected transposition, absence of the superior vena cava, left ventricular diverticulum, complete atrioventricular canal, anomalous left pulmonary artery arising from the aortic arch, absent right pulmonary artery, scimitar syndrome and cor triatriatum.[3,6,7,8,9] Some patients who live to their adulthood without total correction may even develop additional acquired pathologies like coronary arterio sclerosis.[10]

We have observed in our series that secundum type ASD, right aortic arcus and elongated aortic cusps are the most commonly found additional anomalies in TOF. The most common additional pathology is ASD in our series, with 56 cases out of 380 (14.7 %). However, there are only 2 cases of ASD in 31 adult cases (6.5 %) compared to 54 in pediatric age group of 349 patients (15.5 %), which may indicate that the presence of ASD shortens the life expectancy in patients with TOF.[11]

Some of the associated pathologies like abnormal distribution of the coronary arteries, presence of interatrial communications and abnormal systemic venous return are of practical importance during and after total correction. The most commonly encountered coronary artery malformation is the left anterior descending branch originating from

the right coronary artery, which can easily be injured during the right ventriculotomy.[12] Bland-White-Garland's syndrome (left coronary artery arising from the pulmonary artery) is not common in TOF.[12] We have observed 5 coronary artery anomalies in our cases of TOF, 3 of whom showed a left anterior descending branch originating from the right coronary artery, 1 absence of the right coronary artery, and the other one displaying a complex anomaly consisting of the left anterior descending branch arising from the pulmonary artery and left circumflex branch originating from the aorta.[13] This latter patient expired due to the low cardiac output syndrome and autopsy study revealed old and new areas of myocardial infarction.[13]

The presence of atrial septal defect or patent foramen ovale may not attrack attention during the preoperative catheterization. These interatrial communications result in decreased systemic arterial saturation and right-to-left shunt during the postoperative period if overlooked during the total correction. Levine et al.[14] observed 10 cases of right-to-left shunt at the atrial level in 50 cases of TOF studied hemodynamically in the postoperative period. Paradoxical systemic emboli may take place in these cases after total correction, as observed in one of our patients.[1, 14] Therefore, routine digital examination of the right atrium just before the insertion of caval cannulas is nesessary in every case of TOF subjected to total correction to detect a possibly unrecognized interatrial communication.[1, 14]

Anomalies of systemic venous return like the absence of superior venae cava, presence of left persistant superior vena cava and multiple venae cava are also of importance during open heart surgery owing to the cannulation problems in these cases. Left persistent superior vena cava should be cannulated when it drains the upper portion of the body in the absence of the superior vena cava and when the patient can not tolerate the temporary ligation of this aberrant vein in the presence of the superior vena cava. Permenant ligation of the left persistant superior venae cava is rarely performed, as done in only one of our cases. We also observed double inferior vena cava with two atrial orifices and cannulated both orifices during cardiopulmonary bypass in one of our patients.

Summary

Additional pathologies encountered in 380 cases of tetralogy of Fallot are presented. Atrial septal defect (secundum), right aortic arcus and elongated aortic cusps are the most commonly found concomitant anomalies in our series followed by other types of malformations as shown in Table I.

TABLE I

ADDITIONAL PATHOLOGIES IN 380 CASES OF TETRALOGY OF FALLOT*

Associated Pathology	No. Cases
Atrial septal defect (secundum)	56 (14.7 %)
Right aortic arcus	51 (13.4 %)
Elongated aortic cusps	15 (3.9 %)
Patent foramen ovale	10 (2.6 %)
Left persistant superior vena cava	10 (2.6 %)
Absence of crista supraventricularis	7 (1.8 %)
Double outlet right ventricle	7 (1.8 %)
Calcification of the pulmonary valve and outflow tract	7 (1.8 %)
Coronary artery anomalies	5 (1.3 %)
Pulmonary valvular atresia	4 (1.1 %)
Hypoplastic left ventricle	4 (1.1 %)
Patent ductus arteriosus	3 (0.7 %)
Calcification of the aortic valve	2
Absence of the pulmonary valve	2
Supravalvar pulmonary stenosis	1
Bicuspid pulmonary valve	1
Right pulmonary artery coarctation	1
Absence of superior vena cava	1
Double inferior venae cava	1
Partial anomalous pulmonary venous return	1
Pectus carinatum and excavatum	1
Apical cardiac diverticulum and epigastric hernia	1

* Infundibular pulmonary stenosis was associated with valvular pulmonary stenosis in 48 cases (12.6 %)

It is concluded that routine digital control of the right atrium should be done before the insertion of caval cannulas during cardiopulmonary bypass for total correction of Fallot's tetralogy to detect possible existence of preoperatively unrecognized interatrial communications (atrial septal defect and patent foramen ovale)which may lead to significant right-to-left shunt and /or paradoxical emboli during the postoperative period if overlooked during the total correction. The presence of the systemic venous drainage anomalies are also of importance owing to the cannula-

tion problems in these cases in the course of the cardiopulmonary bypass. Abnormal distribution of the coronary arteries can also cause some difficulties during the right ventriculotomy and abnormally branched coronary arteries may be injured leading to myocardial damage if not recognized in the course of the operation.

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