Surgical treatment of tetralogy of Fallot with abnormal course of the coronary artery

Anormal seyreden koroner arter anomalisinde Fallot tetralojisinin cerrahi tedavisi

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Surgically important coronary artery anomalies may be seen uncommonly in tetralogy of Fallot. The most commonly encountered anomaly is the abnormal origin of the left anterior descending coronary artery from the right coronary artery crossing the right ventricular outflow tract. Many different techniques have been described for repair of tetralogy of Fallot with a coronary anomaly. We performed a single right ventriculotomy parallel and distal to the course of the left anterior descending coronary artery that crossed the right ventricular outflow tract in a seven-year-old boy, alleviating the right ventricular outflow tract obstruction without damaging the left anterior descending coronary artery. The pulmonary annulus was enlarged with a pericardial patch through the parallel ventriculotomy, and the patient was discharged without any problem. Our technique has the advantages of leaving the anomalous coronary artery intact throughout the operation, relieving right ventricular outflow tract obstruction easily and satisfactorily, allowing transannular patch application without any damage to the coronary artery, avoiding strecthing of the coronary artery along its course through linear closure of the right ventriculotomy, and avoiding any conduit or valve use.

Key words: Coronary angiography; follow-up; tetralogy of Fallot/complications/radiography/surgery.

Fallot tetralojisinde cerrahi olarak önemli koroner arter anomalilerine seyrek olarak rastlanabilir. En sık rastlanan anomali, sol ön inen koroner arterin anormal olarak sağ koroner arterden çıkıp sağ ventrikül çıkım yolunu çaprazlayarak seyretmesidir. Koroner anomalisi olan Fallot tetralojisinin onarımı için birçok farklı teknik tanımlanmıştır. Biz yedi yaşındaki erkek çocukta, sol ön inen koroner artere zarar vermeden sağ ventrikül çıkım yolu darlığını hafifletecek şekilde sağ ventrikül çıkım yolunu çaprazlayarak sol ön inen koroner arterin gidişinin distaline paralel tek bir sağ ventrikülotomi uyguladık. Paralel ventrikülotomi uygulanarak bir perikard yama ile pulmoner anulus genişletildi ve hasta sorunsuz olarak taburcu edildi. Bizim tekniğimizin, anormal koroner arterin ameliyat boyunca intakt tutulması, sağ ventrikül çıkım yolu darlığının kolay ve yeterli şekilde giderilmesi, koroner artere herhangi bir zarar vermeden transanüler yamanın uygulanması, sağ ventrikülotominin lineer şekilde kapatılması sonucunda koroner arterin trasesi boyunca herhangi bir çekilmenin olmaması ve herhangi bir konduitin veya kapağın kullanılmaması gibi avantajları vardır.

Anahtar sözcükler: Koroner anjiyografi; takip; Fallot tetralojisi/komplikasyon/radyografi/cerrahi.

Surgically important coronary artery anomalies may be seen uncommonly in tetralogy of Fallot. The incidence of a major coronary artery crossing the right ventricular outflow tract (RVOT) in tetralogy of Fallot is between 5 to 12%[1,2] The most commonly encountered anomaly is the abnormal origin of the left anterior descending coronary artery (LAD) from the right coronary artery (RCA).[3] This abnormally originated coronary artery may traverse the RVOT and therefore may be a challenging condition during surgery. Selection of surgical technique mainly depends on the exact location of the anomalous artery and the level and severity of the RVOT obstruction. Although the surgical procedures vary, all of the surgical techniques basically include a "tailored" right ventricular incision and outflow patch reconstruction or a placement of a conduit from the right ventricle to the main pulmonary artery.

Here we report a case of successful surgical repair of tetralogy of Fallot with a coexisting abnormally originated LAD from the RCA, using our simple technique.

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CASE REPORT

The patient was a seven-year-old boy who was found to have a cardiac murmur two years ago. He suffered from mild cyanosis and had frequent respiratory infections. Examination revealed a grade 3/6 systolic ejection murmur heard along the left sternal border. Chest roentgenograms revealed a mild cardiomegaly and decreased pulmonary vascularity. The electrocardiogram demonstrated sinus rhythm and evidence of right ventricular hypertrophy. Transthoracic two-dimensional echocardiography revealed a perimembranous ventricular septal defect and severe subvalvular and valvular pulmonary stenosis with transvalvular gradient of 86 mmHg. Echocardiographic examination also revealed the LAD to be arising from the proximal RCA and traversing the RVOT.

After opening the pericardium the anomalous LAD was seen to traverse the RVOT anteriorly. Following its course, the origin of the LAD from the RCA could be inspected on the anterior surface of the heart. The patient underwent complete repair using cardiopulmonary bypass and intermittent cold blood cardioplegia given in an antegrade fashion. After cardioplegic arrest, a vertical infundibulotomy incision was made parallel to the anomalous artery (Fig. 1). Hypertrophied infundibular muscle bundles were divided and resected through this incision (Fig. 2). The malalignment type ventricular septal defect was closed with a Dacron patch with interrupted pledgeted sutures (Fig. 2). Following ventricular septal defect closure, the pulmonary leaflets were inspected and annulus size was measured through the longitudinal incision on the main pulmonary artery. The pulmonary annulus was smaller than the mean normal diameter measured by a number 12 Hegar dilatator and the pulmonary arteriotomy was extended crossing the annulus to the right ventricle to a distance of about 5 mm from the anomalous artery. The pulmonary valve cusps were thick and had severe commissural fusion. The pulmonary valve leaflets were excised and the pulmonary annulus was enlarged to the size of a Hegar dilatator number 15 by using a patch of autologous pericardium treated with glutaraldehyde. Finally, the infundibular ventriculotomy was closed primarly with a prolene suture (Fig. 3a, b). The patient had an uneventful postoperative course. On the postoperative first month, echocardiography revealed an intact ventricular septal defect and a mean 20 mmHg gradient across the pulmonary outflow tract. At follow-up of six months, the patient is in well condition with the treatment of an angiotensin converting enyzme inhibitor and an oral diuretic twice a week.

DISCUSSION

Abnormal origin of a major coronary vessel transversing the RVOT poses special problems during repair of

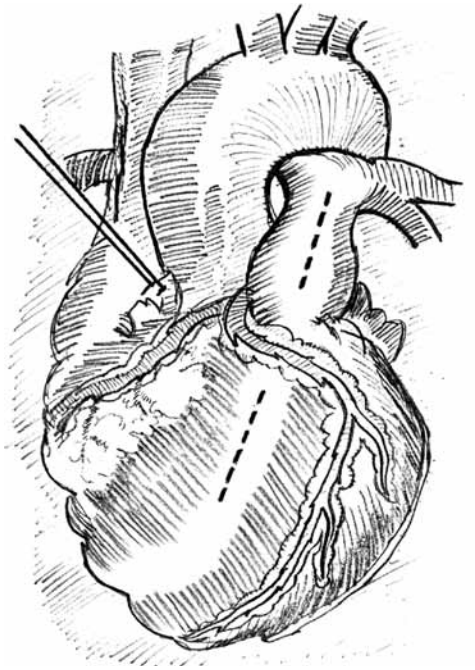

Fig. 1. Longitudinal incision was made in the right ventricular outflow tract distal to the anomalous left anterior descending and in the main pulmonary artery.

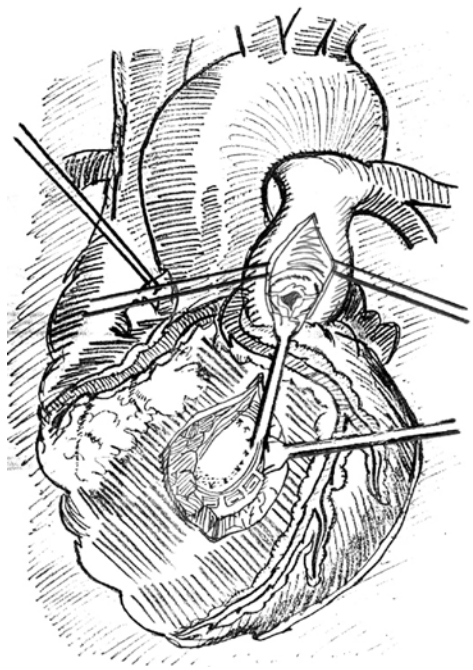

Fig. 2. The ventricular septal defect was closed through the ventriculotomy incision using interrupted pledged sutures as well as the right ventricular outflow tract obstruction relieving. Pulmonary leaflets were excised through the incision involving the main pulmonary artery.

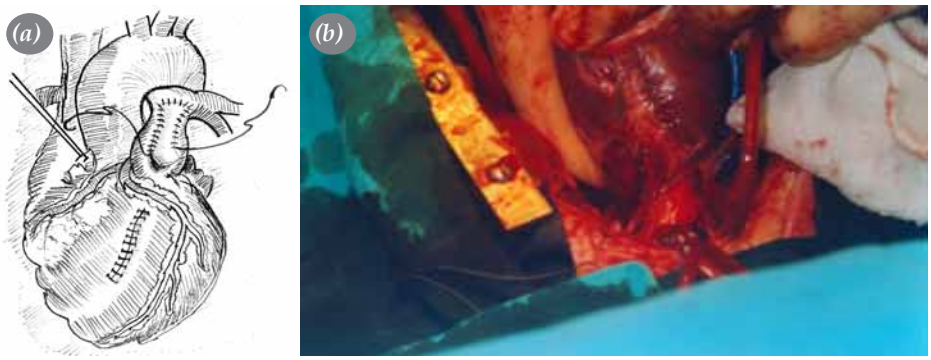

Fig. 3. (a) Intraoperative appearance of the transannular pericardial patch and the linear closure of the right ventriculotomy with intact anomalous left anterior descending coronary artery crossing the right ventricular outflow tract. **(b)** The transannular patch replacement was done using a glutaraldehyde-treated autologous pericardium. The ventriculotomy incision was closed primarly to avoid stretching of the anomalous left anterior descending.

tetralogy of Fallot. Although avoiding the residual leakage of the ventricular septal patch is the important point during surgery, residual gradient in the RVOT remains the major problem in these patients who have this rare condition. Standard methods of repair in tetralogy of Fallot without a coronary anomaly have been reported with good early and late results by many authors.[4-7] However, surgical treatment in tetralogy of Fallot with coexisting major coronary artery abnormality is still a challenging condition. Regarding the surgical technique, the most important factor affecting the residual pulmonary gradient after repair is the technical difficulty to repair a hard to reach RVOT, particularly in severe subpulmonary gradient. Residual significant pulmonary gradient may occur due to insufficient division and resection of the hypertrophied muscles. In such a condition, repair may be accomplished with a tailored right ventricular incision and outflow patch reconstruction or a primarily transatrial approach to relieve the right ventricular outflow tract obstruction. In addition, complete repair can be done by use of an extracardiac conduit. Conduit repairs may result a high reoperation rate especially if they are performed in childhood[8] or may lead to a conduit compressing the coronary artery.[9] Nonconduit methods include a transventricular approach with the ventriculotomy placed paralel to the abnormal coronary artery,[8] construction of a double barrel outlet in patients with a restrictive pulmonary annulus where the anterior wall of the main pulmonary artery is reflected down and sutured to the superior edge of the oblique ventriculotomy and the anterior defect patched with pericardium.[10,11] In addition, Cooley et al.[12] reported an infant with tetralogy of Fallot and a coronary artery anomaly who underwent an internal thoracic artery to left anterior descending anastomosis with successful early and long term result. Kalra et al.[13] reported their successful results with the transatrial-transpulmonary approach in this subgroup of tetralogy of Fallot. More recently, Özkara et al.[14] reported a case with tetralogy of Fallot and anomaly course of LAD who underwent LAD mobilization and RVOT patch enlargement. However, these methods have some drawbacks such as conduit compressing the anomalous coronary artery, residual significant pulmonary gradient and potential damage to the anomalous coronary artery.

Thus, we think that it is unnecessary to use a conduit or to perform other surgical options such as "double outflow technique" reported by van Son.[10] A parallel incision to the anomalous LAD may provide sufficient exposure to relieve the RVOT obstruction adequately. Our technique offers some advantages as follows: *(i)* the anomalous coronary artery is left intact throughout the operation. *(ii)* RVOT obstruction can be relieved more easily and satisfactorily than the other techniques. *(iii)* Transannular patch can be used if necessary without any damage to the coronary artery. *(iv)* Linear closure of the right ventriculotomy avoids strecthing of the coronary artery along its course. *(v)* It also avoids any conduit or valve use.

In summary, many different techniques are used for repair of tetralogy of Fallot with a coronary anomaly, but this method appears to be a viable alternative for relieving the RVOT obstruction, especially in instances of anomalous origin of the LAD from the RCA in cases of the appropriate anatomy between the anomalous coronary artery and pulmonary annulus.

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