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Totally Anomalous Pulmonary Venous Connection Outcomes from A Single Center

Murat Çiçek[1], Oktay Korun[1], Okan Yurdakök[1], Hüsnü Fırat Altın[1], Mehmet Akif Önalan[1], Mustafa Orhan Bulut[2], Filiz İzgi Çoşkun[3], Ahmet Şaşmazel[1], Fatih Özdemir[1], Numan Ali Aydemir[1]

[1] Department of Pediatric Heart Surgery, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey

[2] Department of Pediatric Cardiology, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey

[3] Department of Anesthesiology and Reanimation, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey

ABSTRACT

Introduction: There is limited data on totally anomalous pulmonary venous connection (TAPVC) repair results in our country. The aim of this study is to evaluate the early postoperative results of a large series of TAPVC patients operated in our clinic.

Patients and Methods: The data of the patients who underwent TAPVC repair in our clinic between May 2005 and May 2021 were retrospectively reviewed using hospital records.

Results: A total of 150 TAPVC patients aged between 0 days and 39 years (median 3 months, IQR: 18 days-9 months), 92 males (61%) and 58 females (39%) underwent surgical intervention. The median length of hospital stay was 12 (IQR: 7-16) days. Overall mortality was 14% (21 patients). Seventeen out of 32 patients with additional cardiac anomalies (53%) and 4 out of 118 patients with isolated TAPVC (3%) died. The presence of additional cardiac anomalies was associated with mortality ($p< 0.00001$). Mortality rate in univentricular patients was 59% (10 patients). This rate was higher in comparison to the mortality rate of biventricular patients (8%; $p< 0.00001$). Three patients were reoperated due to postoperative pulmonary venous stenosis.

Conclusion: The TAPVC outcomes in isolated and biventricular patients were favorable with a low mortality and postoperative pulmonary venous stenosis in this cohort. Patients with a single ventricle physiology and/or heterotaxia had a significantly increased risk of mortality, which might be due to the intrinsic challenges of the univentricular physiology regarding the balance between the pulmonary and systemic circulations. Tendency for increased mortality in the cases with preoperative pulmonary venous obstruction is a potential target for improvement.

Key Words: Congenital heart disease; mortality; outcome; pulmonary vein.

Total Anormal Pulmoner Venöz Dönüş Tek Merkez Sonuçlarımız

ÖZ

Giriş: Ülkemizde total anormal pulmoner venöz dönüş (TAPVD) onarım sonuçlarına ilişkin sınırlı veri bulunmaktadır. Bu çalışmanın amacı, kliniğimizde ameliyat edilen geniş bir TAPVD hasta serisinin erken postoperatif sonuçlarını değerlendirmektir.

Hastalar ve Yöntem: Kliniğimizde Mayıs 2005-Mayıs 2021 tarihleri arasında TAPVD tamiri yapılan hastaların verileri hastane kayıtları kullanılarak geriye dönük olarak incelenmiştir.

Bulgular: Yaşları 0 gün ile 39 yaş arasında (medyan 3 ay, IQR: 18 gün-9 ay), 92 erkek (%61) ve 58 kadın (%39) toplam 150 TAPVD hastasına cerrahi girişim uygulanmıştır. Ortanca hastanede kalış süresi 12 (IQR: 7-16) gündü. Genel mortalite %14 (21 hasta) idi. Ek kardiyak anomalili 32 hastadan 17 (%53)'si ve izole TAPVD'li 118 hastadan 4 (%3)'ü kaybedilmiştir. Ek kardiyak anomalilerin varlığı mortalite ile ilişkiliydi ($p< 0.00001$). Univentriküler hastalarda ölüm oranı %59 (10 hasta) bulunmuştur. Bu oran iki ventrikül hastalarının ölüm oranına göre daha yüksekti (%8; $p< 0.00001$). Postoperatif pulmoner venöz darlık nedeniyle tekrar ameliyat olan üç hasta vardı.

Sonuç: Bu kohortta, izole ve biventriküler TAPVD hastalarında mortalite ve postoperatif pulmoner venöz stenoz oranları düşüktü. Tek ventrikül fizyolojisi ve/veya heterotaksisi olan hastalarda mortalite yüksekti. Bu durum pulmoner ve sistemik dolaşımlar arasındaki denge ile ilgili univentriküler fizyolojinin kendine özgü zorluklarından kaynaklanıyor olabilir. Preoperatif pulmoner venöz obstrüksiyonu olan olgularda artmış mortalite eğilimi, iyileştirilmesi gereken bir noktadır.

Anahtar Kelimeler: Konjenital kalp hastalığı; mortalite; sonuçlar; pulmoner ven.

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Correspondence

Murat Çiçek

E-mail: drmcicek@yahoo.com
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INTRODUCTION

Total abnormal pulmonary venous connection (TAPVC) accounts for approximately 1-3% of all congenital heart diseases[1,2]. Sufficient inteatrial flow is necessary for postnatal survival. Patients with insufficient right-left shunt face pulmonary edema and low cardiac output in the early stages of life and inevitably die. In the period from 1956 to the present, when Cooley and Ochsner successfully performed the first successful correction under cardiopulmonary bypass; surgical mortality gradually decreased in parallel with changes in surgical technique, intensive care follow-up procedures and technological developments and decreased from 85% to 5% in simple TAPVCs without additional congenital defects[3,4]. Despite current developments, isomeric patients, patients with single ventricular physiology and patients with diffuse pulmonary vein hypoplasia continue to present challenges in terms of mortality. Another potential problem is pulmonary venous stenosis (PVD) which might lead to early reoperation. Data on TAPVC repair results in our country is limited[5-8]. The aim of this study is to evaluate the early postoperative results of a large series of TAPVC patients operated in our clinic.

PATIENTS and METHODS

The data of the patients who underwent TAPVC repair in our clinic between May 2005 and May 2021 were retrospectively reviewed using hospital records.

All surgeries were performed through a median sternotomy under cardioplegic arrest. Cardiopulmonary bypass was run through aortobicaval cannulation and short-term low flow or deep hypothermia and circulatory arrest were applied in some stages of the anastomosis. Modified ultrafiltration was performed in all newborns and pulmonary hypertensive patients.

In cases with cardiac type TAPVC, where the pulmonary veins returned to the coronary sinus, the ceiling of the coronary sinus was opened and the atrial septal defect was enlarged. The atrial septal defect was closed with a pericardial patch after the pulmonary venous flow was directed to the left atrium without stenosis. In cases with supra and infra-cardiac type anomalies, the heart was lifted towards the right shoulder. A anastomosis as large as possible was created between the pulmonary venous confluence and the posterior wall of the left atrium using prolene sutures. The atrial septal defect was closed with a pericardial patch. In pulmonary hypertensive patients, a 4 mm fenestration was opened on the atrial patch. Vertical vein was not ligated in selected cases with preoperative obstructive pulmonary vein or advanced pulmonary hypertension. In all other cases, the vertical vein was ligated after the anastomosis. Alternatively, in some supra-cardiac cases, the heart was not lifted towards the right shoulder and the anastomosis was created through a transatrial-transseptal approach. Surgical repair was provided by combining the above-mentioned methods in the patients with mixed type abnormal connection.

In cases with postoperative severe pulmonary hypertension without residual gradient in the pulmonary venous connection, we mechanically hyperventilated the patient, started intravenous diuretic infusion, ilioprost infusion (2 ng/kg/min), and 20 parts per million (ppm) nitric-oxide (NO) inhalation therapy for 3 days to reduce pulmonary artery pressures.

Categorical variables were defined as the number of cases (%) and continuous variables as the median (interquartile range). Chi square test or Fisher's exact test were used to compare categorical variables. A p value below 0.05 was considered statistically significant.

RESULTS

A total of 150 TAPVC patients aged between 0 days to 39 years (median 3 months, IQR: 18 days-9 months), 92 males (61%) and 58 females (39%), underwent surgical intervention at Siyami Ersek Thoracic and Cardiovascular Surgery Education and Research Hospital in Turkey. Fifty-seven of all patients were neonatal (38%), and 56 were infant (38%). Seventy-four of the cases had supra-cardiac (49%) type, 40 had intra-cardiac (27%) type, 18 had infra-cardiac (12%) type, and 8 patients had mixed type (supra + intra-cardiac) (5%) TAPVC. In addition to preoperative echocardiography, 23 patients underwent catheter angiography, and 2 patients underwent computed tomography (CT) angiography. It was found that 20 (13%) patients were moderately or severely pulmonary hypertensive, 8 patients (0.5%) had obstructed pulmonary venous drainage (4 were infra-cardiac type, 2 were supra-cardiac type, 2 were mixed type), and 1 patient had diffuse pulmonary vein hypoplasia. One patient had a ventriculoperitoneal shunt due to hydrocephalus in the preoperative period. Additional cardiovascular anomalies were detected in 32 patients (21%) (Table 1).

The median length of hospital stay was 12 (IQR: 7-16) days. Six patients required extracorporeal membrane oxygenation (ECMO) support in the postoperative early period, two of them

Table 1. Additional cardiovascular anomalies

Additional cardiovascular anomaly	Number
Atrial isomerism (unbalance avcd, dorv, ps)	17
Pulmonary stenosis	4
Transposition of great arteries	2
Ventricular septal defect	2
Ventricular septal defect + Pulmonary stenosis	1
Coarctation	1
Coarctation + Pulmonary stenosis	1
Coarctation + Ventricular septal defect	1
Hypoplastic left heart	1
Absent pulmonary valve	1
Interruption inferior vena cava	1

could wean from ECMO and survived. In the postoperative period, 3 patients underwent tracheostomy, and 1 patient underwent left diaphragm plication. Three patients were reoperated due to postoperative pulmonary venous stenosis. One of these patients died due to sepsis, and the remaining two patients were discharged from hospital without any problem.

Seventeen out of 32 patients who had additional cardiac anomalies (53%), and 4 out of 118 patients with isolated TAPVC (3%) died in the postoperative early period, and the overall mortality rate was 14% (21 patients). The presence of additional cardiac anomalies was associated with mortality (p< 0.00001). Of the 4 mortalities who had isolated TAPVC, 1 had diffuse pulmonary hypoplasia and died in the early postoperative period. The other 3 patients were pulmonary hypertensive and died in the postoperative period due to low cardiac output.

Seventeen patients had heterotaxia and all of these patients went through a single-ventricle pathway. There was only 1 additional single ventricle patient with the diagnosis of hypoplastic left heart syndrome and the mortality rate in univentricular patients was 59% (10 patients). This rate was higher in comparison to the mortality rate of biventricular patients (8%; p< 0.00001). Out of these, 2 patients died intraoperatively, 4 patients died due to low cardiac output in the early postoperative period (despite ECMO support), 1 patient died due to sepsis 30 days after the operation.

There were 20 patients (13%) with preoperative obstructive venous drainage, and all of these patients were pulmonary hypertensive neonates. The mortality rate in this subgroup was %20 (4 patients) and all of them died due to low cardiac output in the early postoperative period. Preoperative obstructive venous drainage was not associated with hospital mortality (p= 0.60).

Twenty-four patients received inhale NO theraphy by the median of 3.5 days (range 3-5 days) due to postoperative severe pulmonary hypertension.

DISCUSSION

The results of TAPVC patients who were surgically treated in Siyami Ersek Thoracic and Cardiovascular Surgery Education and Research Hospital in the last two decades were analyzed. Although the operations were performed by many different surgeons, similar surgical techniques were used in accordance with the institutional strategy. In our series, it was observed that the mortality rate was low in patients with simple TAPVC. Additional cardiac anomalies and single ventricle pathway were associated with increased mortality. There was a tendency of increased mortality in patients with preoperative pulmonary venous obstruction. It is noteworthy that the rate of pulmonary venous stenosis that requires postoperative surgery is very low.

In our series, the mortality of univentricular patients was 59%, and most of these patients had heterotaxy. Both pathologies have been previously reported as risk factors for mortality. Although 79% survival has been reported in TAPVC patients with a single ventricle physiology in a study, reports documenting increased risk predominate[9]. In the Boston Children's Hospital series reported by Hancock Friesen and colleagues, 32% of the patients hada single ventricular anatomy. Mortality was 53% in 36 months in the single ventricle group, while it was 13% in patients with two ventricles[10]. This finding was supported by others too (Loca et al.). Increased risk of mortality in patients with TAPVC and heterotaxia was reported by Muhammad et al., in a 65-center study[11]. In our cohort, it was not possible to differentiate weather the increased risk was related with heteroxia or single ventricle physiology. The findings were comparable to the literature. It is plausible to think that even small imbalances in the systemic and pulmonary flow of postoperative single ventricle physiology might lead to bad outcomes in this patient group.

Preoperative pulmonary venous obstruction has been defined as intrinsic or extrinsic narrowing of the abnormal pulmonary venous canal or obstruction in the inter-atrial septum. The stenosis may occur in the connection region of the pulmonary venous sac to systemic circulation or in the form of pulmonary venous hypoplasia[12]. The incidence of obstructive TAPVC has been reported to be approximately 6%. Stenosis in preoperative pulmonary venous anatomy and diffuse pulmonary venous hypoplasia have been associated with mortality[13-15]. Similarly, in our series the mortality rate of the patients in this group was 20%. Presumably, the challenges in management of pulmonary hypertension and congestion contribute to this increased risk in these patients.

Postoperative pulmonary venous stenosis has been reported to be between 10% and 20% and is usually detected around 6 to 12 months post-op[16,17]. There are various hypotheses regarding the etiology of this complication. Pathologically, it is the fibrotic structure formed in the anastomosis line and it can be caused by intimal hyperplasia, which tends to progress along the pulmonary vein. Histologically, it has been reported that the absence of a muscular layer in the pulmonary vein wall and the fact that the left atrial wall is covered with sub-endothelial structure without endothelium in many cases may cause intimal hyperplasia[18]. It was thought that the presence of preoperative pulmonary vein stenosis may pose a risk in terms of postoperative stenosis and that the underlying mechanism is due to the already existing stenosis or hypoplasia being a progressive pathology[2,19,20]. It has been suggested that suture material used during surgery induces intimal hyperplasia and fibrosis[21]. It has been suggested that, the anastomosis performed by placing the apex of the heart on the right shoulder may cause distortion of the anastomosis and therefore performing the anastomosis without changing the position of the heart from the left atrial ceiling in superior type TAPVCs can be advantegous[22]. Primary sutureless technique has been advocated to reduce postoperative pulmonary stenosis but clear benefit could not be shown (R). In our own series, the incidence of postoperative

pulmonary venous stenosis was low (2%). Reoperation was required in 3 patients, two of whom were due to suture line stenosis and the remaining patient due to diffuse pulmonary vein hypoplasia. We attribute this to providing a wide anastomosis line by opening the surgical anastomosis towards each pulmonary vein in the pulmonary venous sac along the entire atrial free wall from the left atrial appendix to the inter atrial septum, expanding the inter atrial septum with the pericardium and preventing iatrogenic damage to the pulmonary veins during the surgical procedure. Similar outcomes were reported by Aytaç et al.[8].

CONCLUSION

The TAPVC outcomes in isolated and biventricular patients were favorable with a low mortality and postoperative pulmonary venous stenosis in this cohort. Patients with a single ventricle physiology and/or heterotaxia had a significantly increased risk of mortality, which might be due to the intrinsic challenges of the univentricular physiology regarding the balance between the pulmonary and systemic circulations. Tendency for increased mortality in the cases with preoperative pulmonary venous obstruction is a potential target for improvement.

Ethics Committee Approval: The Institutional Review Board of Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital approved the study protocol (Decision No: E-28001928-604.01.01, Date: 13.07.2021).

Informed Consent: Informed consent was obtained.

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