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Original Article / Özgün Makale

Single-center outcomes after surgical creation of aortopulmonary window in pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries

Ventriküler septal defekt ve majör aortopulmoner kollateral arterleri olan pulmoner atrezi hastalarında cerrahi aortopulmoner pencere oluşturulması sonrası tek merkez sonuçları

Mehmet Akif Önalan[1], Murat Çiçek[1], Okan Yurdakök[1], Fatih Özdemir[1], Kaan Altunyuva[1], Ali Ertan Ulucan[1], Oktay Korun[1], Hüsnü Fırat Altın[1], Emine Hekim Yılmaz[2], Numan Ali Aydemir[1], Ahmet Şaşmazel[1]

[1]Department of Pediatric Cardiovascular Surgery, Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Türkiye
[2]Department of Pediatric Cardiology, Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Türkiye

ABSTRACT

Background: The aim of this study is to present our experience regarding the creation of an aortopulmonary window as the initial palliative procedure.

Methods: Between February 2016 and February 2021, a total of eight patients (3 males, 5 females; median age: 2 months; range, 0.7 to 6 months) who underwent aortopulmonary window creation were retrospectively analyzed. Data collection was performed by review from our institution.

Results: There was no occurrence of early or late mortality in any patient. The median postoperative duration of mechanical ventilation and length of hospital stay were five and eight days, respectively. No postoperative reperfusion injury or severe pulmonary overcirculation was observed in any of the patients. Four patients achieved complete repair with unifocalization of the major aortopulmonary collateral arteries, one patient had a second procedure, and the remaining three patients waited for complete repair. The median right ventricle-to-aortic pressure ratio after complete repair was 0.6 (range, 0.4 to 0.7). The median follow-up after complete repair was 1.4 (range, 0.9 to 2.8) years, and the median follow-up period for all survivors was 2.7 (range, 0.9-5.8) years.

Conclusion: Our study results suggest that aortopulmonary window operation can be safely performed in selected patients with good early and mid-term outcomes. Although the central pulmonary arteries are very small, half of the patients underwent complete repair and achieved acceptable right ventricle-to-aortic pressure ratios. Patient selection criteria and early postoperative interventions are of utmost importance to prevent postoperative pulmonary overcirculation.

Keywords: Aortopulmonary window, major aortopulmonary collateral arteries, pulmonary atresia.

ÖZ

Amaç: Bu çalışmada ilk palyatif işlem olarak aortopulmoner pencerenin oluşturulması ile ilişkili deneyimimiz sunuldu.

Çalışma planı: Şubat 2016-Şubat 2021 tarihleri arasında aortopulmoner pencere oluşturulan toplam sekiz hasta (3 erkek, 5 kız; medyan yaş: 2 ay; dağılım, 0.7-6 ay) retrospektif olarak incelendi. Veri toplama kurumumuzdan inceleme yapılarak gerçekleştirildi.

Bulgular: Hastaların hiçbirinde erken veya geç mortalite görülmedi. Medyan ameliyat sonrası mekanik ventilasyon ve hastanede kalış süresi sırasıyla beş ve sekiz gün idi. Hiçbir hastada ameliyat sonrası reperfüzyon hasarı veya pulmoner aşırı dolaşım izlenmedi. Hastaların dördüne majör aortopulmoner kollateral arterlerin unifokalizasyonu ile tam düzeltme yapıldı, bir hastaya ikinci bir cerrahi işlem uygulandı ve geri kalan üç hasta tam düzeltme için bekletildi. Tam düzeltmeden sonra sağ ventrikülün aort basıncına oranı medyan 0.6 (dağılım, 0.4-0.7) idi. Tam düzeltme sonrasında medyan takip süresi 1.4 (dağılım, 0.9-2.8) yıl olup, sağkalanlar için medyan takip süresi 2.7 (dağılım, 0.9-5.8) yıl idi.

Sonuç: Çalışma sonuçlarımız aortopulmoner pencere ameliyatının, seçilmiş hastalarda iyi erken ve orta dönem sonuçları ile güvenli bir şekilde uygulanabileceğini göstermektedir. Santral pulmoner arterler çok küçük olmasına rağmen, hastaların yarısında kabul edilebilir sağ ventrikül-aortik basınç oranları ile tam düzeltme sağlandı. Ameliyat sonrası pulmoner aşırı dolaşımı önlemek için hasta seçim kriterleri ve ameliyat sonrası erken müdahaleler çok önemlidir.

Anahtar sözcükler: Aortopulmoner pencere, majör aortopulmoner kollateral arterler, pulmoner atrezi.

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Correspondence: Mehmet Akif Önalan, MD. Dr. Siyami Ersek Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Çocuk Kalp ve Damar Cerrahisi Kliniği, 34668 Üsküdar, İstanbul, Türkiye.
Tel: +90 554 - 301 59 08 e-mail: mehmetakifonalan@gmail.com

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Pulmonary atresia (PA) with ventricular septal defects (VSDs) and major aortopulmonary collateral arteries (MAPCAs) is an uncommon and complex manifestation of congenital heart disease.[1] Improvement in the prognosis of PA/VSD/MAPCAs is due to the progress of treatment in these patients over the past two decades.[1,2] Single-stage midline unifocalization can be used in most patients with PA/VSD/MAPCAs and is, therefore, the preferred method of treatment.[3,4]

There is a small subgroup of patients with MAPCAs who have hypoplastic and confluent intrapericardial pulmonary arteries, and normal arborization; therefore, it is of utmost importance to ensure dual blood supply to the pulmonary segments that have blood flow from MAPCAs. Collateral arterial circulation in these cases is usually composed of multiple small vessels, most of which communicate with the native pulmonary artery circulation.[5] If clinical cyanosis occurs, then this group of patients is appropriate for the creation of an aortopulmonary (AP) window by means of direct anastomosis between the aorta and small main pulmonary artery trunk. This palliative procedure often results in growth of the native pulmonary artery system.[6] Although we have attempt to apply the midline unifocalization strategy for the majority of patients with MAPCAs, we also know that there is a small subgroup of patients who are appropriate for the creation of an AP window without touching the MAPCAs during the initial operation.[7]

Currently, only few centers report the findings in the literature regarding the effect of specific anatomical features on outcomes.[8,9] In the present study, we aimed to present the early and mid-term results of surgical creation of the AP window as the initial palliative procedure.

PATIENTS AND METHODS

This single-center, retrospective study was conducted at Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, Department of Department of Pediatric Cardiovascular Surgery between February 2016 and February 2021. The medical records of consecutive patients who underwent AP window creation due to PA/VSD/MAPCAs were reviewed. Patients with PA/VSD/MAPCAs undergoing shunt operation and single-ventricle patients were excluded from the study. Inclusion criteria for this procedure were multiple hypoplastic aorticopulmonary collateral arteries, dual supply collateral arteries, confluent and small (<2.5 mm) branch pulmonary arteries with a good arborization pattern and cyanosis. During this period, a total of 62 patients underwent operation for VSD/PA/MAPCA, and 48 of them underwent an unifocalization procedure. A total of eight patients (3 males, 5 females; median age: 2 months; range, 0.7 to 6 months) who met these criteria were included.

Preoperative cardiac catheterization was performed to identify the central branch PAs and MAPCA anatomy in all the patients. The patients had a median of 3.5 (range, 2 to 4) MAPCAs. The patients had a median of two MAPCAs supplying the right lung (range, 1 to 3) and a median of 1.5 MAPCAs supplying the left lung (range, 1 to 2). The median preoperative oxygen saturation was 79% (range, 75 to 85%), and the median preoperative fractional shortening (FS) was 36% (range, 33 to 39%).

Surgical technique

All operations were performed through median sternotomy with cardiopulmonary bypass (CPB) on standby. The main pulmonary artery and the right and left branch pulmonary arteries were carefully mobilized, and temporary neurovascular clips were placed on the branch pulmonary arteries. The atretic main pulmonary artery was divided as close as possible to its right ventricular (RV) infundibular origin. The location for the anastomosis on the aorta is critical to avoid kinking or stenosis of the branch pulmonary arteries. Systemic heparinization (150 U/kg) was applied, a side-biting clamp was inserted on the left posterolateral aspect of the ascending aorta, taking care of the left main coronary artery, and a small button of the aorta was extracted with a No. 11 blade. The main pulmonary artery segment was incised for a distance of approximately 3 mm, and the anastomosis was performed with 8-0 polypropylene suture. After finalization of the AP anastomosis, the side-biting clamp was taken, and temporary neurovascular clips were removed. Heparin was reversed in two patients due to the CPB requirement.

When pulmonary patch plasty was required in the second surgical procedure, first, the right and left pulmonary arteries were divided, and then both pulmonary arteries were opened down to the lower branch. Pulmonary patch plasty was performed, and the right and left pulmonary arteries were rejoined. For pulmonary artery reconstruction, we preferred using fresh autologous pericardium first, but if not possible, we used a bovine pericardial patch. In the final stage, a systemic-pulmonary (S-P) shunt or RV-pulmonary artery conduit (RV-pulmonary artery) was applied to

the reconstructed pulmonary artery. We performed pulmonary artery patch plasty with a native pericardial patch, if the patient had their own pericardium; if not, we used a bovine pericardial patch.

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 21.0 software (IBM Corp., Armonk, NY, USA). Continuous variables were expressed in median (min-max) values, while categorical variables were expressed in number and frequency.

RESULTS

The median weight at the time of surgery was 5 (range, 3.3 to 7) kg. Surgical creation of the AP window was performed as the primary procedure in all the patients included in this study. Native tissue-to-tissue anastomosis was used in all patients, and a synthetic aorta-pulmonary artery shunt was not required in any patient. No takedown of the window or revision of the anastomosis was required in any patient. The postoperative period of all the patients was uneventful, reperfusion injury or severe pulmonary overcirculation was not observed in any of the patients, and their postoperative management was routine. The median duration of intensive care unit stay after operation was five (range, 1 to 24) days, and the median duration of hospital stay after operation was eight (range, 4 to 38) days. The median postoperative oxygen saturation was 87% (range, 82 to 90%). Postoperative early mortality, congestive heart failure, pulmonary edema, or prolonged mechanical ventilation were not observed in any of the patients, and all patients were uneventfully discharged from the hospital.

The median follow-up for all the survivors was 2.6 (range, 0.8 to 5.8) years. A total of four of the eight patients could not achieve complete repair (CR), and one of them underwent S-P shunt with pulmonary patch plasty after AP window creation. In this case, at the hilum of both pulmonary arteries, there was a tissue ring, and pulmonary arteries were reconstructed as described in the surgical technique. A S-P shunt was performed on the right side of the pulmonary artery. The other three patients did not undergo their second surgical procedure and were waiting for CR.

The median pulmonary artery diameter, as measured at the midportion of the branch pulmonary arteries, increased from 2.3 (range, 2.1 to 2.4) mm before the creation of the AP window to 4.6 (range, 2.9 to 6.3) mm after the operation, with a median interval to postoperative cardiac catheterization of 13 (range, 8 to 16) months. The perioperative characteristics of the patients are listed in Table 1.

Four of the eight patients achieved CR with MAPCA unifocalization at an average of 14.5 months after AP window creation. One out of four patients underwent RV-pulmonary artery conduit replacement with pulmonary artery patch plasty as a second operation before CR. The median age at the time of CR was two (range, 1.2 to 3.5) years, and the median weight at the time of CR was 10 (range, 8.5 to 12) kg. One MAPCA was closed by coil embolization in a patient due to dual supply and competitive flow before CR. During the CR operation, two MAPCAs were unifocalized in two patients, and one MAPCA was unifocalized in the

Table 1. Perioperative characteristics of the patients

Characteristics	n	%	Median	Range
Sex				
Male	3	37.5		
Median weight at surgery (kg)			5	3.3-7
Median age at surgery (month)			2	0.7-6
Median of preoperative O_2 saturation		79		75-85
The median of preoperative Pa diameter (mm)			2.3	2.1-2.4
Median of postoperative ICU stay (days)			5	1-24
Median of postoperative hospital stay (days)			8	4-38
Median of postoperative Pa diameter after the follow-up (mm)			4.6	2.9-6.3
Median of follow-up (years)			2.6	0.8-5.8
Overall mortality	0			

Pa: Pulmonary artery;. ICU: Intensive care unit.

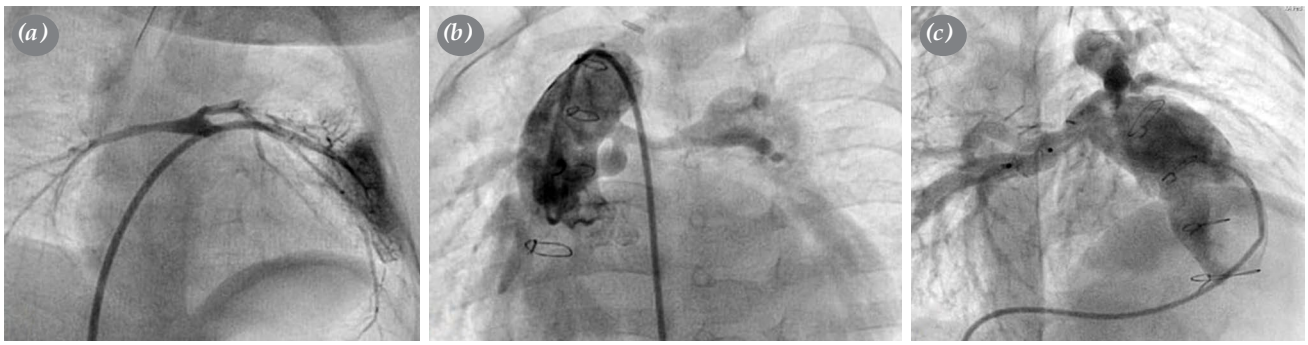

Figure 1. (a) Preoperative angiogram demonstrating diminutive central pulmonary arteries that are confluent and have normal arborization. **(b)** The angiogram of the same patient after the aortopulmonary window creation operation, demonstrating growth of the native pulmonary arterial system. **(c)** Angiogram of the same patient demonstrating the pulmonary arterial system after the complete repair operation.

other two patients. Additional pulmonary artery reconstruction was performed in all patients at the time of CR (Figure 1). A total of six MAPCAs were ligated during CR in four patients. The median RV/aortic peak systolic pressure ratio after repair was 0.6 (range, 0.4 to 0.7). After CR, pulmonary overcirculation occurred in only one patient, and one MAPCA was closed by coil embolization three days after the operation. The median duration of intensive care unit stay after CR was 10.5 (range, 6 to 26) days, and the median duration of hospital stay after CR was 19 (range, 8 to 27) days. Pulmonary artery reintervention was not performed in any of the patients who underwent CR; however, one of them was followed closely due to peripheral pulmonary stenosis. Repaired patients were followed for a median of 1.4 (range, 0.9 to 2.8) years after the repair. In our series, there was no late death during the follow-up period.

DISCUSSION

This report presents our surgical experience with eight patients who underwent an AP window creation operation as their initial palliative procedure for PA/VSD/MAPCAs. The primary goal of surgical management in patients with PA/VSD/MAPCA is single-stage midline unifocalization and CR. There is a specific subgroup of patients who have hypoplastic (<2.5 mm diameter) and confluent intrapericardial pulmonary arteries and have normal arborization with complete or nearly complete dual blood supply MAPCAs. When patients with this anatomical variation present with cyanosis in the neonatal period, our preference is to perform a surgical AP window to stimulate the growth of pulmonary arteries.[5]

In our study, there was no postoperative early mortality at the time of AP window creation. A total of 50% of patients eventually achieved CR, and 37.5% of patients achieved CR at the second surgery. These results indicate that surgical creation of an AP window is a very safe approach, in the early stage, for a specific subgroup of patients.

Pulmonary artery enlargement within two to six months allows many of these patients to become better candidates for subsequent single-stage or staged CR.[5] Bauser-Heaton et al.[8] conducted a study in 40 patients who underwent AP window creation due to VSD/PA/MAPCAs. In most patients in this study, pulmonary artery development was sufficient for CR at the second surgery, with a median of 6.2 months after AP window creation. In our study, half of the patients were able to undergo CR with unifocalization of MAPCAs, a median of 14 months after AP window creation. We believe that the patients who needed more time to achieve CR in our study compared to the literature is because most of our patients were living outside the city and they encountered certain obstacles that hindered follow-up during the pandemic. Survival after CR with an acceptable RV/aortic peak systolic pressure ratio (0.6) was good in our study, and there were no early- or mid-term deaths after APW creation or CR operations.

Watterson et al.[10] reported that postoperative pulmonary artery distortion or stenosis was common in older patient populations undergoing longer palliative procedures with an AP window; however, this finding was not observed in our study. Adequate mobilization of the pulmonary arteries and optimal placement of the window over the aorta are important to prevent

proximal branch pulmonary artery stenosis. In our study, none of the patients required surgical or catheter-based pulmonary artery reintervention after CR.

Mainwaring et al.[6] conducted a study in 35 patients who underwent AP window creation due to PA/VSD/MAPCA with hypoplastic and confluent pulmonary artery branches. Complete repair was achieved in 55% of the patients, consistent with our study. They suggested that hypoplastic pulmonary arteries and diminished pulmonary blood flow were markers for a less well-developed pulmonary vascular bed. Therefore, they hypothesized that the competitive flow related to a dual supply system might have opposite effects on the enlargement of the distal vascularity. Based on this view, MAPCA ligation was performed in 36.6% of the patients in their study, and in 50% of the patients in our study. In addition, while MAPCA unifocalization was performed in 49% of the patients in their study, MAPCA ligation was performed in 50% of the patients who underwent CR in our study.

The pulmonary artery reconstruction technique during the second surgical procedure is also important. Monge et al.[11] conducted a study of 16 patients who had peripheral pulmonary artery stenosis with Williams and Alagille syndromes. They separated the right and left pulmonary arteries and incised both lower branches toward pulmonary artery. After pulmonary artery patches were performed separately, they reconnected both pulmonary arteries and, finally, a S-P shunt or a RV-pulmonary artery conduit was placed on the reconstructed pulmonary arteries. They demonstrated a 55% decrease in RV to aortic peak systolic pressure ratios by this technique.[11,12] We used this surgical technique in our patients during the CR procedure or during interstage additional shunt operations, and we believe that this technique contributes to our acceptable RV/aortic peak systolic pressure ratios and in not requiring reinterventions of pulmonary arteries after CR operations.

During our study period, a total of 62 patients underwent operation for VSD/PA/MAPCA, and 12.9% (n=8) of them had an AP window. Mainwaring et al.[13] conducted a study in 307 patients who underwent any operation for VSD/PA/MAPCA, and 14.9% of them had an AP window. This study is the one of the largest series in the literature and the rates of AP window patients in our study are similar to this study.

The single-center, retrospective nature of the study and the small number of patients included in our study are the main limitations. Further multi-center, large-scale studies with long-term outcomes are needed to evaluate the surgical results of AP window creation.

In conclusion, our study results show that surgical creation of the aortopulmonary window is feasible in selected patients with early- and mid-term outcomes. However, it typically requires multiple operations to improve the development of pulmonary arteries to allow closure of the ventricular septal defects.

Ethics Committee Approval: The study protocol was approved by the Haydarpaşa Numune Training and Research Hospital Clinical Research Ethics Committee (27/12/2021/No: 2021/330-3127). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Patient Consent for Publication: A written informed consent was obtained from each parent.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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