Turkish Journal of Thoracic and Cardiovascular Surgery 2019;27(3):274-279
http://dx.doi.org/doi: 10.5606/tgkdc.dergisi.2019.16398

Original Article / Özgün Makale

Transcatheter occlusion of antegrade pulmonary blood flow in children with univentricular heart

Tek ventrikül hastalığı olan çocuklarda antegrad pulmoner kan akımının transkateter yöntemle kapatılması

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ABSTRACT

Background: This study aims to evaluate the results of transcatheter occlusion of antegrade pulmonary blood flow in children with univentricular heart.

Methods: Medical data of a total of seven patients (4 females, 3 males; median age 11.7 years; range, 1 to 24 years) who underwent transcatheter occlusion of the antegrade pulmonary blood flow following Glenn shunt or Fontan operation between September 2014 and January 2017 were retrospectively analyzed. Data including demographic and clinical characteristics of the patients, type of surgery, echocardiographic and cardiac catheterization findings were recorded.

Results: Four patients had a previous pulmonary artery banding operation, while three had pulmonary stenosis. Two patients had facial and upper extremity edema after Glenn operation, one had prolonged pleural effusion, one had prolonged pleural effusion after Fontan operation, and one developed dyspnea and effort intolerance several years after Fontan operation. In two patients, antegrade pulmonary blood flow was occluded to decrease systemic ventricular load before surgery. The Amplatzer Septal Occluder was used in five patients and the Amplatzer Vascular Plug-2 was used in two patients. Two patients developed transient, complete atrioventricular block during the procedure and the procedure was terminated early in one of these patients. Transient hemolysis was observed in one patient following the operation.

Conclusion: Transcatheter occlusion of antegrade pulmonary blood flow is an effective alternative to surgery in patients with hemodynamic compromise after Glenn shunt or Fontan operation.

Keywords: Fontan procedure, pediatric, pulmonary artery, vascular closure devices.

ÖZ

Amaç: Bu çalışmada tek ventrikül hastalığı olan çocuklarda antegrad pulmoner kan akımının transkateter yolla kapatılmasının sonuçları değerlendirildi.

Çalışma planı: Eylül 2014 - Ocak 2017 tarihleri arasında Glenn şant veya Fontan ameliyatı sonrası transkateter yöntemle antegrad pulmoner kan akımının kapatıldığı toplam yedi hastanın (4 kız, 3 erkek; medyan yaş 11.7 yıl; dağılım, 1-24 yıl) tıbbi verileri retrospektif olarak değerlendirildi. Hastaların demografik ve klinik özellikleri, cerrahi türü, ekokardiyografik ve kardiyak kateterizasyon bulguları kaydedildi.

Bulgular: Dört hastaya önceden pulmoner arter bantlama ameliyatı yapılmış iken, üç hastada pulmoner darlık mevcut idi. İki hastada Glenn ameliyatı sonrası yüzde ve üst ekstremitede ödem, bir hastada uzamış plevral efüzyon, bir hastada Fontan ameliyatı sonrası uzamış plevral efüzyon ve bir hastada da Fontan ameliyatından yıllar sonra gelişen solunum sıkıntısı ve efor intoleransı mevcuttu. İki hastada ise cerrahi öncesi sistemik ventrikül yükünü azaltmak için antegrad pulmoner kan akımı kapatıldı. Beş hastada Amplatzer Septal Occluder ve iki hastada Amplatzer Vascular Plug-2 kullanıldı. İki hastada işlem sırasında geçici tam atriyoventriküler blok gelişti ve bu hastalardan birinde işlem erken sonlandırıldı. Bir hastada işlem sonrasında geçici hemoliz gözlendi.

Sonuç: Hemodinamik bozukluğu olan hastalarda Glenn şantı veya Fontan ameliyatı sonrasında antegrad pulmoner kan akımının transkateter yolla kapatılması, cerrahiye etkili bir alternatiftir.

Anahtar sözcükler: Fontan işlemi, çocuk, pulmoner arter, vasküler kapatma cihazları.

Received: June 22, 2018 *Accepted*: April 12, 2019 *Published online*: June 19, 2019

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Cite this article as:

Şahin GT, Tanıdır İC, Gökalp S, Güzeltaş A. Transcatheter occlusion of antegrade pulmonary blood flow in children with univentricular heart. Turk Gogus Kalp Dama 2019;27(3):274-279

Conflicting evidence remains regarding whether antegrade pulmonary blood flow (APF) should be left open or closed during Glenn shunt operations in patients with univentricular heart. Antegrade pulmonary blood flow may contribute to pulmonary artery growth, improve oxygenation, and prevent arteriovenous fistula formation by providing pulsatile flow and transmission of hepatic factors to the lung.[1,2] However, APF which is unproblematic in the beginning, may result in various complications such as effort intolerance, atrioventricular (AV) valve regurgitation, ventricle dysfunction, increased pulmonary artery pressure leading to Glenn shunt dysfunction, recurrent pleural effusions, and protein losing enteropathy over time.[3,4] Antegrade pulmonary blood flow should be, therefore, closed either when complications develop or during the Fontan surgery.

In the present study, we aimed to evaluate the results of transcatheter occlusion of APF in children with univentricular heart.

PATIENTS AND METHODS

A total of seven patients (4 females, 3 males; median age 11.7 years; range, 1 to 24 years) who underwent transcatheter occlusion of the APF following Glenn shunt or Fontan operation between September 2014 and January 2017 were retrospectively analyzed. Data including demographic and clinical characteristics of the patients, type of surgery, echocardiographic and cardiac catheterization findings were retrieved from the hospital database. A written informed consent was obtained from each parent. The study protocol was approved by the Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital Ethics Committee. The study was conducted in accordance with the principles of the Declaration of Helsinki.

All cardiac catheterization procedures were performed under general anesthesia with a laryngeal mask. Routine anticoagulation with a 50 IU/kg loading dose of heparin was administered to reach a target activated clotting time (ACT) of 200 sec. The procedure was performed through either the internal jugular vein or the femoral vein. After blood samples were taken and pressure recordings were recorded for the hemodynamic assessments (from superior vena cava, inferior vena cava, right atrium, left atrium, aorta, pulmonary artery and ventricular end-diastolic pressures), angiograms were taken. Aorticopulmonary collateral arteries were evaluated by aortic injection and right and left subclavian artery injections, if necessary. The APF quantity, spatial relationship between the aorta and pulmonary artery, and aortic flow were evaluated by ventricular injections. Venovenous and arteriovenous fistulas were examined by superior vena cava, inferior vena cava, left innominate vein, and pulmonary artery angiograms. In the absence of any other pathology unveiling clinical findings, the evaluation process for APF closure was started. If the patient cannot proceed to Fontan surgery, APF closure would have a very unfavorable outcome; therefore, APF should be only closed in very selected cases in whom no other choice is left.

In all patients, test occlusion with appropriate size balloons was conducted before APF device occlusion. The balloon, which was suitable for the main pulmonary artery width (wide enough to completely close the antegrade flow), was inflated in the main pulmonary artery. Pulmonary artery pressure and oxygen saturation were re-evaluated after a 10-min test occlusion period. The APF occlusion was only performed in patients with an oxygen saturation of >80%.

Either Amplatzer Vascular Plug-II (AVP-II, St. Jude Medical, MN, USA) or Amplatzer Septal Occluder (ASO, St. Jude Medical, St. Paul, MN, USA) devices were chosen for occlusion, depending on the diameter and length of the main pulmonary artery. The occlusion procedure was performed either via the right internal jugular vein or femoral vein depending on the existing cardiac pathology. The device position was confirmed with transthoracic echocardiography during the procedure.

In patients who were scheduled to undergo a Fontan operation, APF occlusion was completed a few days before the planned surgery time.

Statistical analysis

Statistical analysis was performed using the PASW version 17.0 software (SPSS Inc., Chicago, IL, USA). Continuous data were expressed in median (min-max) values, while categorical variables were expressed in number and frequency.

RESULTS

Of the patients, four presented with a double inlet left ventricle (DILV) and ventriculoarterial discordance (VA). Two patients had additional subvalvular and valvular pulmonary stenosis (PS), while the other two previously underwent pulmonary banding operation. The remaining patients presented with tricuspid atresia with VA concordance (n=1), double outlet right ventricle (DORV), and pulmonary stenosis (n=1) and double inlet right ventricle (DIRV) with DORV (n=1).

Table 1. Demographic and clinical characteristics of patients

Patient	Age (year)	Diagnosis	Procedure timing	Indication	Type of APF	Saturation	Relation of great vessels
1	1	Tricuspid atresia	Post BCPA	Upper extremity edema	PAB	90	N
2	1	DORV, VSD, PS	Post BCPA	Upper extremity edema	NPV	93	D-malposition
3	2	DILV	Post BCPA	Pleural effusion	PAB	92	Anteroposterior
4	4	DILV	Pre Fontan	Pre Fontan	NPV	92	L-malposition
5	4	DILV	Pre Fontan	Pre Fontan	PAB	94	Anteroposterior
6	11	DIRV, DORV	Post Fontan	Pleural effusion	PAB	92	D-malposition
7	24	DILV	Post Fontan	Shortness of breath and effort intolerance	NPV	100	Anteroposterior

APF: Antegrade pulmonary blood flow; BCPA: Bidirectional cavopulmonary anastomosis; PAB: Pulmonary artery band; DORV: Double outlet right ventricle; VSD: Ventricular septal defect; PS: Pulmonary stenosis; NPV: Native pulmonary valve; DILV: Double inlet left ventricle; DIRV: Double inlet right ventricle.

In total, four of seven patients previously underwent pulmonary banding operation with three having PS. Demographic and clinical characteristics of patients are shown in Table 1. In six patients, the pulmonary artery was located posterior to the aorta (directly posteroanterior in three patients, D-malposition in two patients and L-malposition in one patient) and it was normally related in the remaining patient.

Prolonged pleural effusion (persisting chylothorax for two months after surgery) after a Glenn operation was observed in one patient. Upper extremity edema secondary to high pulmonary artery pressure, prolonged pleural effusion after the Fontan operation in the early postoperative period (12 days after the operation), dyspnea and effort intolerance in the years following the Fontan operation were reported in a patient with APF. In two patients, the procedure was performed a few days before the Fontan operation.

The preferred devices for occlusion were ASO in four patients and Amplatzer Vascular Plug-II (AVP-II) in two patients (Figure 1). In one patient, closure attempt was unsuccessful. Procedures were performed via the right internal jugular vein in three patients and the right femoral vein in the other three patients.

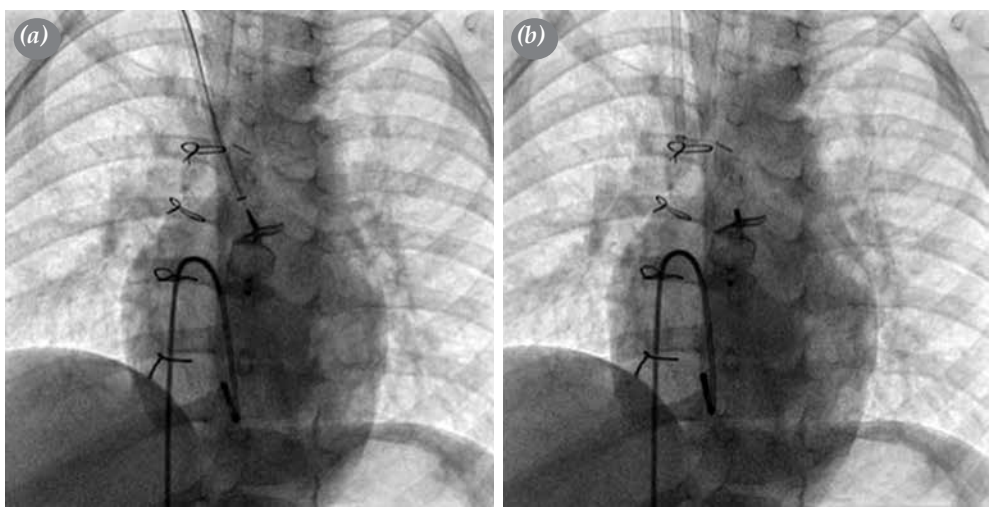

Figure 1. (a) Antegrade closure of native pulmonary valve APF with a 14-mm AVP II angiogram shows the device in good position prior to release. **(b)** An 14-mm AVP II device was successfully deployed from the right internal jugular vein.

APF: Antegrade pulmonary blood flow; AVP: Amplatzer vascular plug.

Table 2. Procedural data

Patient	APF closure device	Venous access	Mean PAP (baseline)	Mean PAP (after)	Outcome	Complication
1	ASO 14 mm	RIJ	18	16	Edema resolved	Temporary hemolysis
2	ASO 6 mm	RFV	24	18	Edema resolved	AV block (transient)
3	ASO 13 mm	RFV	10	8	Failed to process	AV block (transient)
4	AVP-II 14 mm	RIJ	19	13	Fontan completed	No
5	AVP-II 16 mm	RFV	16	13	Fontan completed	No
6	ASO 11 mm	RFV	33	26	Pleural effusion regression	No
7	ASO 15 mm	RIJ (AV loop)	9	9	Asymptomatic	No

APF: Antegrade pulmonary blood flow; PAP: Pulmonary artery pressure; ASO: Amplatzer septal occluder; RIJ: Right internal jugular; RFV: Right femoral vein; AVP: Amplatzer vascular plug; AV: Atrioventricular.

Complete AV block occurred in two patients during the procedure. In one patient, the sinus rhythm was established during the procedure and occlusion was performed, while the procedure was terminated early due to the sustained AV block in the other patient. In this case, normal sinus rhythm was established 36 hours after the procedure. Temporary hemolysis, which resolved within seven days, was observed in one patient. Procedural data are summarized in Table 2.

DISCUSSION

Permitting limited antegrade pulmonary blood flow after Glenn shunt operation has been shown to have several advantages. However, antegrade overflow or limited, but prolonged flow may cause complications, such as ventricular dysfunction or Glenn shunt dysfunction.[1,4-6] Therefore, the overflow must be prevented to avoid complications.

In our routine practice, APF is maintained after Glenn shunt operation, if the pulmonary artery pressure and oxygen saturation are suitable. Catheter angiography is recommended in case of pleural effusion and edema persisting for more than two weeks after Glenn shunt to identify the etiology and potential treatment.[7] We have also a standardized APF closure protocol in our clinic (Figure 2). After excluding AV valve regurgitation, PS, aortopulmonary collaterals, and stenosis between the superior vena cava and pulmonary artery as etiological factors in patients with clinical symptoms, we occlude APF. In addition, in eligible patients, we prefer to occlude the APF before the Fontan operation both to decrease volume load and reduce the diastolic dysfunction risk in the early postoperative period.

In the present study, the procedure was performed to resolve complications in two patients after Glenn shunt

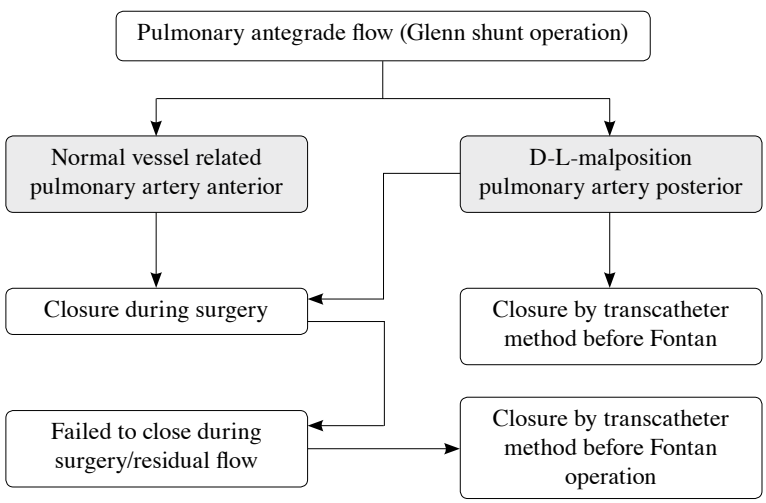

Figure 2. Our institutional standardized antegrade pulmonary blood flow closure protocol.

operation and in two patients after Fontan operation. Maintaining APF after Fontan operation is not a viable strategy, as it changes the pulmonary-to-systemic flow ratio (Qp/Qs). However, in patients where APF is believed to be occluded or in patients with a very limited flow, these complications may be overlooked. Due to very limited APF in two patients in our study, development of complications took some time and transcatheter occlusion requirement developed many years later.

In patients with transposition of the great arteries, successful location of the pulmonary artery during the operation is difficult and prolongs the operation time, thereby increasing the complexity of surgery. Therefore, in this group of patients, transcatheter APF occlusion has been suggested as an effective alternative to surgery (Figure 2).[8,9] The occlusion procedure was performed in our study either preoperatively (one week before the operation) or in the early postoperative period. Six of seven patients had transposition of the great arteries. In two of these patients, we occluded APF a week before the Fontan operation.

Device preferences for APF occlusion vary depending on the anatomy of an individual patient.[10] If the patient had a previous pulmonary banding operation, the band may be close to the pulmonary artery bifurcation. In this case, the preferred device should have a retention disk on one side, such as a duct occluder. If there is a wide space on both sides of the pulmonary valve in patients with native pulmonary artery stenosis, the chosen device should have a retention disk on both sides, such as a septal occluder.[7,11] Petko et al.[11] used an ASO, Ductal Occluder and AVP, while Desai et al.[8] preferred a Rashkind Umbrella Occluder, Amplatzer Septal and Ductal Occluder, and Shanghai Shape Memory Alloy (SHSMA) muscular ventricular septal defect occluder (Shanghai Shape Memory Alloy Co., Ltd. Shanghai, China) device was deployed at the ligation site of pulmonary trunk for APF occlusion by Li et al.[12] The aforementioned studies all reported successful results. In our study, we used an ASO in four patients and an AVP-II in two patients. In a patient in whom the ASO was used, complete AV block developed and remained unresolved; therefore, the procedure was completed without closure.

Pulmonary artery stenosis is frequently observed in patients with univentricular circulation. Stenosis in the pulmonary arteries or systemic veins, if noticed, should be corrected preoperatively. Stenosis may also develop postoperatively mainly at the anastomotic site. If the Fontan circulation fails due to APF and pulmonary artery stenosis, both pathologies can be treated with a single intervention by implanting a covered stent, as reported in the study of Yücel et al.[13]

Both antegrade and retrograde methods for APF occlusion have been previously defined in the literature.[11,14] Petko et al.[11] reported antegrade occlusion in four patients and retrograde occlusion in three patients. We preferred the antegrade method in three and retrograde method in four patients. In a patient where occlusion was unable to be performed using the antegrade method, we were obliged to make a loop for occlusion, as we were unable to reach the pulmonary artery from the femoral artery.

Closure of the source of APF using various occluding devices including Amplatzer ductal occluder, Amplatzer atrial septal defect occluder, and covered stents has been successfully described with no device-related complications.[8,15,16] However, long-term follow-up data are still lacking. Madan et al.[17] used AVP-II to occlude APF at an external center and reported residual antegrade flow probably due to the lack of occlusive material within the vascular plug and aortic valve perforation due to proximal disc impinging on the aortic wall. A patient whose right ventricle-pulmonary artery conduit (Sano shunt) was occluded by a vascular plaque died 10 days after the procedure due to cardiopulmonary failure and sepsis.[11] Torres et al.[7] reported left PS in a patient after a duct occluder device implantation for occlusion of APF. The stenosis was treated with a stent implanted in the left pulmonary artery in the same procedure. We observed complete AV block in two patients with D-transposition of the great arteries. In one patient, AV block was transient and reversed during the procedure, while the other continued and resulted in termination of the occlusion procedure. The patient's rhythm returned to sinus in the following 36 hours. We also observed transient hemolysis after occlusion in a patient with diagnosed tricuspid atresia (Glenn shunt and pulmonary banding). This patient required an erythrocyte transfusion and hemolysis did not recur during follow-up.

The main limitations of the present study include its single-center design and limited clinical experience in a small sample size.

In conclusion, transcatheter occlusion of antegrade pulmonary blood flow is an effective alternative to surgery, if *(i)* APF is left open in Glenn shunt operation, when Glenn dysfunction developes due to APF; *(ii)* malposition of the great arteries and adhesion of tissues complicates the occlusion procedure after

an elective Fontan operation; and *(iii)* when APF is unable to be occluded due to technical difficulties or there is a remaining residual flow. Nonetheless, further large-scale studies are needed to establish a definite conclusion.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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