Occlusion of the left superior vena cava-coronary sinus connection in a child with Glenn dysfunction by the transcatheter approach

Glenn anastomozu fonksiyon bozukluğu olan bir çocukta sol süperiyor vena kava-koroner sinüs bağlantısının kateter yoluyla kapatılması

Murat Saygı, M.D., Hasan Tahsin Tola, M.D., Alper Güzeltaş, M.D., Ender Ödemiş, M.D.

Department of Pediatric Cardiology, Istanbul Mehmet Akif Ersoy Cardiovascular Training and Research Hospital, Istanbul

Summary– A 14-year-old male patient presented with cyanosis and tiredness. The patient had undergone a Glenn procedure at age 12 following the echocardiographic determination of a double inlet left ventricle, ventriculoarterial discordance, moderate valvular-subvalvular pulmonary artery stenosis, non-restrictive inlet ventricular septal defect and right ventricle hypoplasia; his oxygen saturation was 70%. Echocardiography evaluation showed retrograde flow from the vena cava superior to the innominate vein and a left superior vena cava (LSVC) opening into the coronary sinus (CS). Here, we report the case of a patient evaluated for Glenn dysfunction in whom an increase in oxygen saturation was observed following transcatheter occlusion of the LSVC-CS connection using an Amplatzer septal occluder.

Özet– On dört yaşındaki erkek hasta, yorgunluk ve siyanoz şikayeti ile başvurdu. On iki yaşında iken Glenn prosedürü uygulanan hastanın ekokardiyografik incelemesinde, çift girişli sol ventrikül, ventriküloarteryel diskordans, orta derece subvalvüler-valvüler pulmoner arter darlığı, non-restriktif inlet ventriküler septal defekt ve sağ ventrikül hipoplazisi vardı. Oksijen satürasyonu %70 idi. Ekokardiyografik incelemede ayrıca, süperiyor vena kava'dan innominate vene geriye doğru bir akım ve koroner sinüse drene olan sol süperiyor vena kava gözlendi. Bu yazıda, Glenn anastomozu fonksiyon bozukluğu nedeni ile değerlendirdiğimiz ve sol süperiyor vena kava-koroner sinus bağlantısını kateter yoluyla Amplatzer septal occluder cihazı ile kapatarak oksijen satürasyon değerinde artış sağladığımız bir hastayı bildirdik.

The systemic veins that appear for the first time or increase in caliber following Fontan or Glenn procedures drain into the pulmonary venous circulation; therefore, cyanosis may be observed in these patients.[1,2] The detailed, exhaustive study of systemic venous structures both before and after the procedure is of major importance, especially in the case of heterotaxy syndrome and some other complicated cardiac disorders. Thanks to the development of interventional cardiac catheterization techniques, transcatheter occlusion is possible in several instances of cyanosis that develops as a result of systemic venous abnormalities. Although standard coil techniques are predominant in transcatheter occlusion, full occlusion devices are sometimes used, depending on the diameter of the vascular structure involved.[3]

Here, we report the case of a patient evaluated for Glenn dysfunction in whom an increase in oxygen saturation was observed following trans catheter occlusion of the left superior vena cava (LSVC)-coronary sinus (CS) connection using an Amplatzer septal occluder.

Abbreviations:

CS *Coronary sinus*
LSVC *Left superior vena cava*

CASE REPORT

A 14-year-old male patient presented with cyanosis and tiredness. The patient had undergone a Glenn procedure at age 12 following the echocardiographic determination of a double inlet left ventricle, ventriculoarterial discordance, moderate valvular-subvalvular pulmonary artery stenosis, non-restrictive inlet ven-

Received: March 16, 2014 *Accepted:* May 30, 2014

Correspondence: Dr. Murat Saygi. İstanbul Mehmet Akif Ersoy Göğüs Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hatanesi, Çocuk Kardiyoloji Kliniği, Halkalı, Küçükçekmece, 34303 İstanbul.
Tel: +90 212 - 692 20 00 / 1154 e-mail: saygimrt@gmail.com

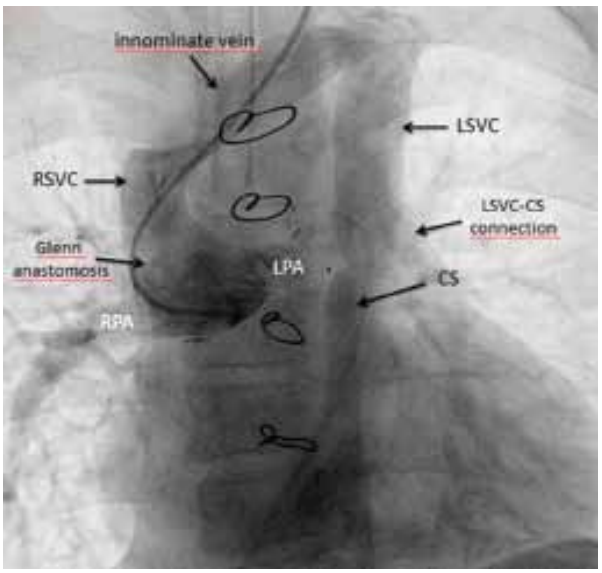

Figure 1. Retrograde flow from LSVC to CS observed after injection of contrast medium to the Glenn anastomosis through a left jugular access before transcatheter occlusion. CS: Coronary sinus; LPA: Left pulmonary artery; LSVC: Left superior vena cava; RPA: Right pulmonary artery; RSVC: Right superior vena cava.

tricular septal defect and right ventricle hypoplasia; his oxygen saturation was 70%. Echocardiography evaluation showed retrograde flow from the vena cava superior to the innominate vein and an LSVC opening into the CS. Heart catheterization to occlude the LSVC-CS communication was indicated. Using a left jugular vein access, a 5F NIH catheter was advanced into the innominate vein, and the injection of contrast medium showed retrograde flow from the LSVC to the CS through the innominate vein (Figure 1). The measured LSVC diameter was 16 mm, and the narrowest point of the LSVC-CS communication was 11 mm. Entering through the femoral vein, a 0.035″ hydrophilic guide wire and 5F Judkins right 4 catheter were advanced into the inferior vena cava, the right atrium, the CS, and finally, the LSCV. An injection of contrast medium confirmed angiographically the LSCV opening into the CS. This connection was occluded using a 25 mm x 30 mm sizing balloon. While applying the occlusion, it appeared that the retrograde flow shown earlier from the innominate vein was not present when contrast medium was injected into the catheter passing from the left jugular vein to the innominate vein; the contrast medium filled the entire pulmonary vascular bed (Figure 2a) in the absence of an elevation in the mean Glenn pressure. The patient's oxygen saturation rose from 70% to 87%. A 13 mm Amplatzer septal occluder was advanced through a 7F-long sheath in the left femoral vein to the LSVC-CS communication, where it was placed. After observing the absence of retrograde flow via the injection of contrast medium from the jugular venous catheter into the innominate vein (Figure 2b), the device was left in place. No complications were observed during the procedure. Following the inter-

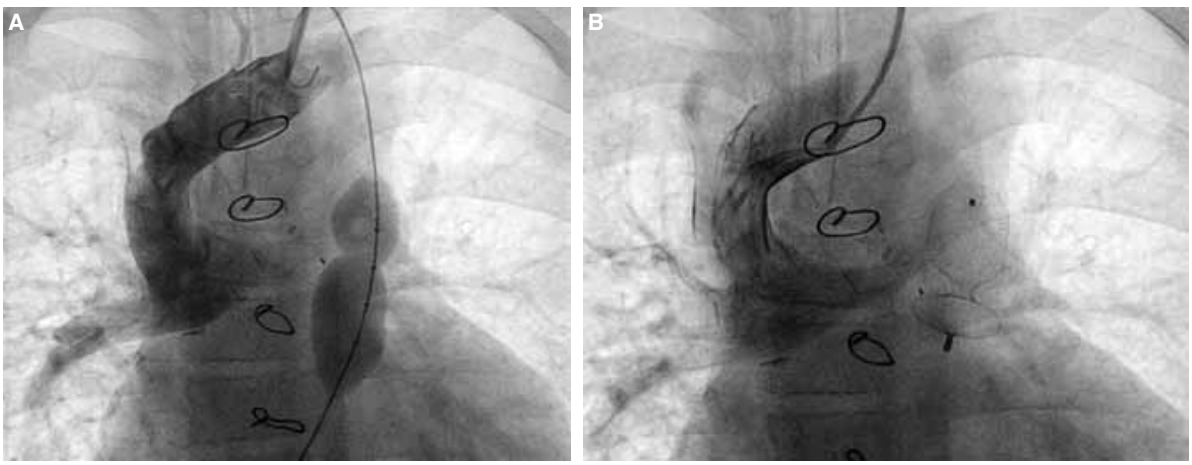

Figure 2. **(A)** Following occlusion of the LSVC-CS link by a sizing balloon, contrast medium injection into the innominate vein through the jugular shows absence of the earlier retrograde flow; the contrast medium fills the entire pulmonary vascular bed. **(B)** Following occlusion of the LSVC-CS link by an Amplatzer septal occluder, contrast medium injection into the innominate vein shows absence of the earlier retrograde flow; the contrast medium fills the entire pulmonary vascular bed.

vention, oxygen saturation values remained in the 85-90% range versus the pre-intervention 70% level.

DISCUSSION

Cardiopulmonary connecting procedures are part of palliative surgical treatment strategies applied in complex congenital cardiac malformations that do not offer an anatomic possibility of biventricular repair. A slow, progressive fall of oxygen saturation values due to various causes may develop in the postoperative course in these patients. The principal causes of the cyanosis observed in these patients are the development of collateral venous circulation between the superior and inferior vena cava systems, pulmonary arteriovenous malformations, veno-venous collaterals that develop between the systemic and pulmonary veins, and, following a Fontan procedure, the occurrence of baffle fenestration leaks.[1,2] The reopening of an LSCV or it's becoming symptomatic in the postoperative period, while hemodynamically insignificant before the procedure, may cause cyanosis in patients previously treated by cavopulmonary connecting procedures, due to the communicating systemic and pulmonary venous systems.[4,5] In these patients, systemic venous blood that should flow to the pulmonary artery reverses its course, directing it to the CS and causing desaturation. An exhaustive evaluation of the systemic venous anatomy in the preoperative evaluation of both Fontan and Glenn procedure patients is highly significant from the viewpoint of obtaining the desired oxygen saturation and reducing the risk of paradox embolization. In addition to the usual echocardiographic evaluation of communication between the systemic and pulmonary venous systems in patients subjected to Fontan or Glenn procedures, contrast echocardiography also has an important role to play. The filling of the left atrium earlier than expected by the contrast medium may be interpreted as suggesting the presence of a systemic venous communication draining into the pulmonary venous system.[6,7]

The Amplatzer vascular plug, Amplatzer duct occluder, and Gianturco-Grifka vascular occlusion device are frequently used by pediatric cardiologists to occlude abnormal vascular connections.[8] Abdullah et al.[9] reported on two patients in whom late cyanosis developed following a Fontan palliative procedure, leading to trans catheter occlusion of a dilated LSVC. A 10 mm x 12 mm Amplatzer duct occluder was used in one of the cases, and an Amplatzer patent foramen ovale occluder was used in the other case; desaturation was corrected in both. In a study of 45 consecutive patients subjected to a Fontan procedure and later developing cyanosis, Masura et al.[10] identified 51 unnecessary connections. The authors reported that all Fontan operations in their facility were using fenestration; 50 of these 51 redundant connections were treated by device implantation. Of these 50 interventions, 38 were performed for fenestration, six for collateral venous circulation, three for pulmonary arterial malformation, and another three for lateral tunnel leak. The study reported that follow-up showed that the Fontan procedure patients frequently developed cyanosis, most often due to a right-to-left shunt caused by the Fontan fenestration. Other important factors in developing cyanosis were identified as collateral venous circulation, pulmonary arterial malformation, and lateral tunnel leak. The report also indicated that unnecessary communications might be closed by trans catheter intervention using different Amplatzer occluders. In the case reported herein, we used an Amplatzer septal occluder device by the trans catheter route to occlude an LSVC-CS connection for desaturation connected to retrograde flow identified in a Glenn anastomosis; an important recovery of oxygen saturation was the result.

In conclusion, the presence of right and left vena cava superior flows should be extensively evaluated in patients in whom a cavopulmonary connection is planned. Characteristics that might be causing desaturation in the postoperative follow-up should also be carefully studied. A vena cava superior that does not participate in the Glenn anastomosis may become hemodynamically significant in the postoperative period, causing desaturation. These redundant connections that develop during the follow-up can be closed by transcatheter interventions using various occlusive devices.

Conflict-of-interest issues regarding the authorship or article: None declared.

REFERENCES

1. McElhinney DB, Reddy VM, Hanley FL, Moore P. Systemic venous collateral channels causing desaturation after bidirectional cavopulmonary anastomosis: evaluation and management. J Am Coll Cardiol 1997;30:817-24. CrossRef
2. Gatzoulis MA, Shinebourne EA, Redington AN, Rigby ML,

Ho SY, Shore DF. Increasing cyanosis early after cavopulmonary connection caused by abnormal systemic venous channels. Br Heart J 1995;73:182-6. CrossRef

3. Recto MR, Elbl F, Austin E. Transcatheter closure of large persistent left superior vena cava causing cyanosis in two patients post-Fontan operation utilizing the Gianturco Grifka vascular occlusion device. Catheter Cardiovasc Interv 2001;53:398-404. CrossRef
4. Hsu HS, Nykanen DG, Williams WG, Freedom RM, Benson LN. Right to left interatrial communications after the modified Fontan procedure: identification and management with transcatheter occlusion. Br Heart J 1995;74:548-52. CrossRef
5. Filippini LH, Ovaert C, Nykanen DG, Freedom RM. Reopening of persistent left superior caval vein after bidirectional cavopulmonary connections. Heart 1998;79:509-12.
6. Bernstein HS, Brook MM, Silverman NH, Bristow J. Development of pulmonary arteriovenous fistulae in children after cavopulmonary shunt. Circulation 1995;92(9 Suppl):II309-14.
7. Van Hare GF, Silverman NH. Contrast two-dimensional echocardiography in congenital heart disease: techniques, indications and clinical utility. J Am Coll Cardiol 1989;13:673-86.
8. Holzer RJ, Cheatham JP. Therapeutic cardiac catheterization. In: Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, editors. Moss and Adams' heart disease in infants, children and adolescents: including the fetus and young adult. 8th ed. Philadelphia: LWW; 2013. p. 288-347.
9. Abdullah AF, Menahem S. Transcatheter closure of dilated left superior vena cava for resolution of late cyanosis following fontan palliation. Heart Lung Circ 2006;15:393-6. Epub 2006 Jun 8. CrossRef
10. Masura J, Bordacova L, Tittel P, Berden P, Podnar T. Percutaneous management of cyanosis in Fontan patients using Amplatzer occluders. Catheter Cardiovasc Interv 2008;71:843-9.

Key words: Amplatzer septal occluder; Glenn procedure; heart catheterization; heart septal defects, atrial/therapy; septal occluder device; vena cava, superior.

Anahtar sözcükler: Amplatzer septal occluder; Glenn prosedürü; kalp kateterizasyonu; kalp septal defekti, atriyal/tedavi; septal tıkayıcı cihaz; vena kava, süperiyor.