

## Successful stenting of systemic venous pathway stenosis after double switch repair for congenitally corrected transposition of great arteries in a child

### Doğuştan düzeltilmiş büyük damar transpozisyonu nedeniyle double switch ameliyatı yapılan bir çocukta gelişen sistemik venöz darlıkta başarılı stent işlemi

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**Summary**– An 8-year-old boy with previous shunt operation for corrected transposition of great arteries, ventricular septal defect, pulmonary stenosis and multiple aortopulmonary collateral arteries underwent corrective surgery. In the early post-operative period, there were clinical findings of superior vena cava obstruction. Cardiac catheterization at 72 h following surgery showed a systemic venous baffle stenosis between the vena cava and right atrium. A stent was successfully implanted in the vena cava percutaneously, and the stenosis was relieved. Her symptoms resolved in a short time period, and she was extubated rapidly. During the follow-up, excellent maintenance and patency of systemic venous baffle were observed.

**Özet**– Doğuştan düzeltilmiş büyük damar transpozisyonu, ventriküler septal defekt, pulmoner darlık ve çoklu aortopulmoner kollateral arter nedeni ile önce şant ameliyatı olan sekiz yaşındaki erkek çocuk, tam düzeltme ameliyatı oldu. Erken ameliyat sonrası dönemde superior vena cava tıkanıklığı bulguları gelişti. Ameliyat sonrası 72. saatte yapılan kalp kateterizasyonu vena cava ve sağ atriyum arasında, sistemik venöz baffle darlığı olduğunu gösterdi. Stent perkütan olarak vena kavaya başarıyla yerleştirilerek darlık giderildi. Hastanın belirtileri kısa bir sürede kayboldu ve hasta hızlıca ekstübe edildi. Takip sırasında sistemik baffle açıklığının mükemmel düzeyde sağlandığı gözlemlendi.

Systemic venous pathway (SVP) stenosis is one of the post-operative complications of the mustard repair procedure for complete transposition or double switch operations. An obstructed SVP following atrial switch operations is the most common indication for stent delivery in the venous system.<sup>[1,2]</sup> Covered and bare metal stents are the most-suitable treatment options for vessel stenosis in congenital heart diseases.<sup>[2-5]</sup>

transposition of the great arteries, pulmonary stenosis, and aortopulmonary collaterals along with large atrial septal defects.

#### Abbreviations:

CP	Cheatham platinum
PAP	Pulmonary artery pressure
RA	Right atrium
SVC	Superior vena cava
SVP	Systemic venous pathway

#### CASE REPORT

An 8-year-old boy was admitted with a small restrictive ventricular septal defect, congenitally corrected

After an arterial double switch operation which included pulmonary xenograft conduit repair, the post-operative intensive care unit stay was uneventful. After extubation, the patient's superior vena cava (SVC) dilatation presented with obstruction symptoms as well as facial swelling, head fullness, and dyspnea. The patient had also hepatomegaly, tibial edema, and abdominal distension. A

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chest X-ray revealed cardiomegaly with pleural effusion. Echocardiography demonstrated a patent inferior part of systemic venous return with SVC dilatation and dynamic obstruction during the diastolic phase of the ventricular contraction. During the catheterization, the mean pressure gradients between the SVC and the right atrium (RA) and for the inferior caval vein were 11 mmHg (SVC: 25 mmHg; RA: 14 mmHg) and 14 mmHg, respectively. The mean pulmonary artery pressure (PAP) was 29 mmHg (systolic PAP: 40 mmHg; diastolic PAP: 20 mmHg). Angiography demonstrated a patent pulmonary outflow tract with SVP stenosis. Therefore, a stent to connect the SVC to the RA was planned to implant (Figure 1a and Video 1\*).

Under general anesthesia and after right femoral vein cannulation, SVC was crossed by a 6F right-guided catheter. The catheter was then exchanged for a standard extra-stiff wire guide, and the 6F sheath was replaced with a long 14 French sheath. Based on SVC angiographic findings (SVC diameter: 12 mm), a cheatham platinum (CP) (Numed NY, USA) balloon expandable, 34 mm (CP8Z34) stent and balloon in a balloon (14 mm) were selected. The balloon was then inflated at 3 atm at a diameter of 14 mm with a procedural time of 15 min and a fluoroscopy time of eight min.

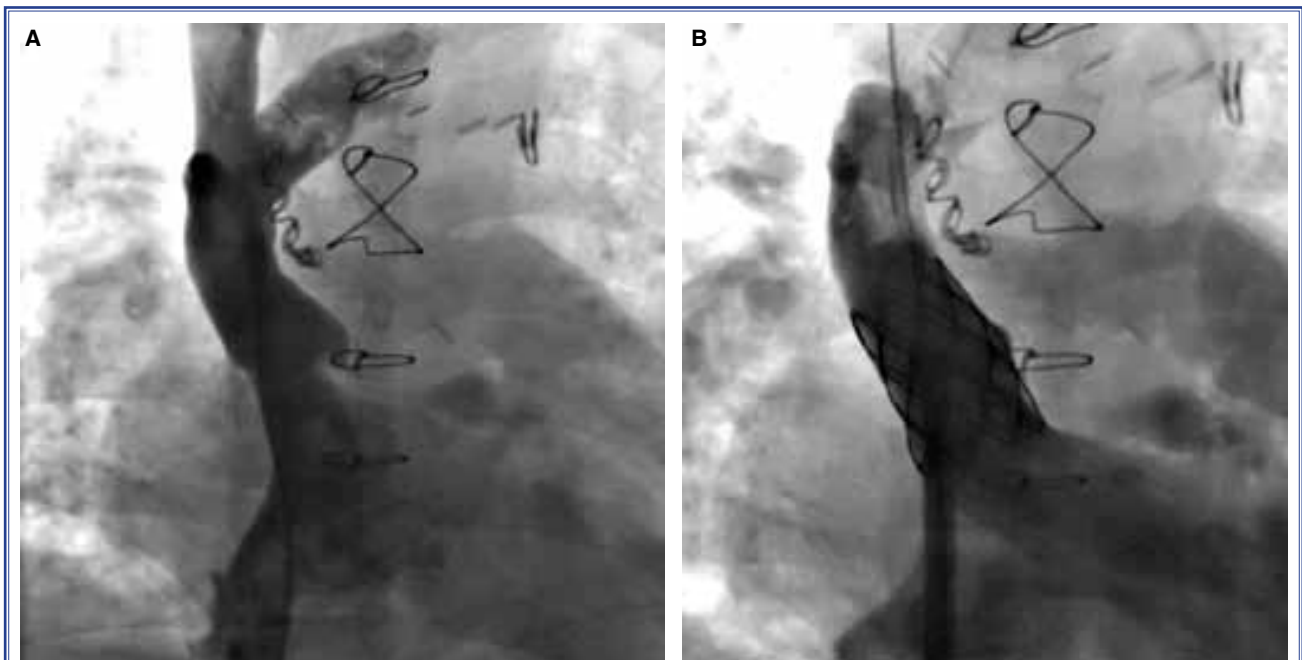
Repeated hemodynamic measurement demonstrated a complete lack of obstruction without a pressure gradient in the upper part of the SVC-to-RA stent. Angiography also showed that the stent was properly placed (Figure 1b and Video 2\*). Ascites was reduced, and abdominal distension resolved soon after the stent implantation.

The patient without any clinical signs of jugular vein restenosis was discharged with aspirin therapy. Repeated echocardiography showed no residual stenosis and the room air saturation remained at 93% during follow-up. At 6-10 months of surgery, the patient is still asymptomatic.

## DISCUSSION

Several conditions may lead to stenosis of the systemic veins, including SVC and IVC obstruction after an atrial switch operation.<sup>[1-3]</sup> The reported incidence of systemic venous baffle obstruction after an atrial switch (Mustard or Senning) operation ranges from 0% to 20%.<sup>[2]</sup> Obstruction of the SVP is a rare, yet known complication in the early post-operative course of the Senning procedure.<sup>[2-5]</sup>

It is established that acute obstruction of the SVC pathway after atrial repair is not well tolerated. It may



**Figure 1.** Angiocardiogram of the patient with superior vena cava and systemic venous baffle obstruction after the senning procedure (A) and immediately after the stent procedure (B).

be responsible for the venous insufficiency and superior vena cava syndrome.<sup>[1-4]</sup> In the early post-operative course, clinical systemic venous stenosis was dramatically symptomatic in our patient. As a result, we may conclude that it is unlikely certain to what degree stenosis intervention should be performed in symptomatic patients. In addition, one editorial paper reported that the importance of mild gradients across the polyvinylpyrrolidones (arbitrarily <3 mmHg) is unknown, whereas gradients of 10-15 mmHg was found in patients with protein-losing enteropathy.<sup>[3,4]</sup>

Stents are used as the primary approach in such lesions, as the procedure is simple and effective.<sup>[1-3]</sup> There are no evidence-based protocols which serve as indications for the intervention; however, stenting is known to be critical in many asymptomatic patients with minimal gradients and modest stenosis (33-40%). Therefore, clinical symptomatic obstructions or obstructions larger than 50% of the large systemic veins as confirmed by angiography are considered appropriate for stent implantation.<sup>[2]</sup> Patients with atrial baffles are also good candidates for stenting.<sup>[2]</sup> A self or balloon-expandable and a covered or bare metal CP stent is the best choice for the vessels as well as for SVP stenosis.<sup>[3-5]</sup>

Utilizing a balloon expandable stent in combination with a CP stent is a simple and effective procedure which can be used successfully for the treatment of SVP obstruction after the Senning procedure

thanks to its ability for rapid and dramatic relief in selected patients.

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

**\*Supplementary video file associated with this article can be found in the online version of the journal.**

## REFERENCES

1. Michel-Behnke I, Hagel KJ, Bauer J, Schranz D. Superior caval venous syndrome after atrial switch procedure: relief of complete venous obstruction by gradual angioplasty and placement of stents. *Cardiol Young* 1998;8:443-8. [CrossRef](#)
2. Tzifa A, Marshall AC, McElhinney DB, Lock JE, Geggel RL. Endovascular treatment for superior vena cava occlusion or obstruction in a pediatric and young adult population: a 22-year experience. *J Am Coll Cardiol* 2007;49:1003-9. [CrossRef](#)
3. Rosenthal E, Qureshi SA. Stenting of systemic venous pathways after atrial repair for complete transposition. *Heart* 1998;79:211-2.
4. Ward CJ, Mullins CE, Nihill MR, Grifka RG, Vick GW 3rd. Use of intravascular stents in systemic venous and systemic venous baffle obstructions. Short-term follow-up results. *Circulation* 1995;91:2948-54. [CrossRef](#)
5. Daehnert I, Hennig B, Wiener M, Rotzsch C. Interventions in leaks and obstructions of the interatrial baffle late after Mustard and Senning correction for transposition of the great arteries. *Catheter Cardiovasc Interv* 2005;66:400-7. [CrossRef](#)

**Key words:** Child; double switch; stenosis; transposition of great arteries; stents.

**Anahtar sözcükler:** Çocuk; double switch; darlık; düzeltilmiş büyük damar transpozisyonu; stent.