

**Down sendromlu pulmoner hipertansiyonu olan bir çocuk hastada  
geniş patent duktus arteriozusun fenestrasyon uygulanmış  
muscular VSD occluder cihazı ile kapatılması**

**Closure of wide patent ductus arteriosus using a fenestrated muscular  
VSD occluder device in a pediatric patient with Down syndrome and  
pulmonary hypertension**

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**Özet-** Zamanında tedavi edilmemiş geniş patent duktus arteriozusun ve önemli pulmoner hipertansiyonu bulunan hastalar kardiyologlar için önemli bir sorundur. Bu hastalarda kateter laboratuvarında uygulanan reverzibilite ve balon oklüzyon testleri hastanın sonraki planlaması için karar vermede yardımcı olabilir. Kateter yoluyla duktusun kapatılması planlanan hastalarda bu amaçla geliştirilmiş cihazların yanısıra off-label kullanımı olan farklı cihazlar da kullanılabilir. Reverzibilite testi sınırdaki pozitif olarak bulunduğunda, seçilmiş hastalarda fenestre edilmiş cihaz kullanılabilir. Bu yazıda, geniş duktusu ve sistemik pulmoner hipertansiyonu olan, muscular VSD occluder cihazının fenestre edilerek off-label kullanılmasıyla kateter yoluyla kapatma işlemi yapılan 10 yaşında Down sendromlu bir hasta sunuldu.

Patent ductus arteriosus (PDA), even excluding those detected in premature babies, is a frequently seen (9-12%) anomaly among all congenital heart diseases.<sup>[1]</sup> Closure of PDAs with audible persistent murmurs which also cause significant left- to-right shunts or overloading of the left ventricle is recommended. Transcatheter closure of PDA has been used prevalently in line with development of new devices, loading systems, and techniques, and accumulating experience of the physicians, and it has become the first line of treatment within the last 20 years.<sup>[2]</sup> Transcatheter or surgical closure of the PDA is contraindicated in the presence of advanced stage pulmonary vascular disease, and active infective endocarditis.

**Summary-** Patients with wide patent ductus arteriosus and significant pulmonary hypertension not treated in time constitute a significant problem for cardiologists. For these patients, tests that could aid in decision-making for further planning include reversibility and balloon occlusion tests performed in the catheterization laboratory. Devices developed for the closure of ductus as well as different devices with off-label use may be employed in patients scheduled for transcatheter occlusion. When result of reversibility test is borderline positive, the use of fenestrated device may be applicable for selected patients. Presently described is case of a 10-year-old patient with Down syndrome who had a wide ductus and systemic pulmonary hypertension. Transcatheter closure procedure was performed with off-label use of a fenestrated muscular ventricular septal defect occluder device.

In advanced age patients with wide PDA, and pulmonary hypertension (PHT) application of vasoreactivity, and balloon occlusion tests before transcatheter closure may aid the clinician. In this article transcatheter closure procedure performed in a 10-year-old Down syndrome patient with wide ductus, and secondary systemic PHT with off-label use of fenestrated Amplatzer® muscular VSD occluder device.

**Kısaltmalar:**

PDA Patent ductus arteriosus  
PHT Pulmonary hypertension  
PVR Pulmonary vascular resistance  
SVR Systemic vascular resistance

Submitted on: 06.30. 2016 Accepted for publication on: 12.09. 2016

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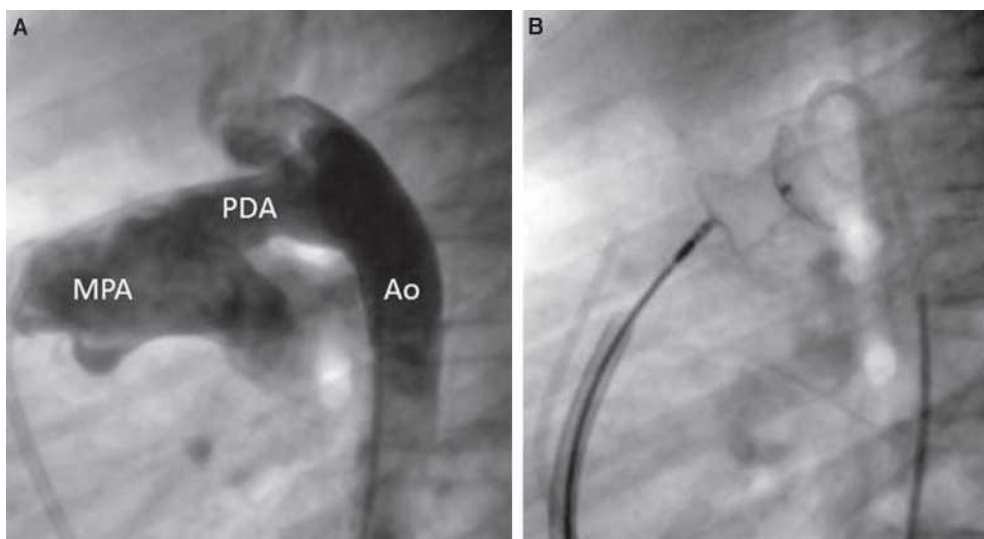
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### CASE PRESENTATION

On physical examination of a 10-year-old patient followed with the diagnosis of Down syndrome and received diagnosis of PDA, and significant PHT, sharp, and harsh second heart sound was heard over all foci, and also a 3/6 systolic murmur was detected. On echocardiographic examination wide, bidirectional shunt, more prominently left-to-right shunt, and systemic PHT were detected. Cardiac chambers, and systolic functions were normal. During the first cardiac catheterization applied anatomy of PDA was examined. PDA was 14 mm long with a diameter of 11 mm, and an ampulla measuring 14 mm (Figure 1a). Some parameters were as follows: pulmonary artery pressure: 101/58 (72) mmHg, aortic pressure: 110/64 (80) mmHg, pulmonary vascular resistance (PVR) 9.5 Wood units  $m^2$ , PVR/SVR (systemic vascular resistance) 0.8. Nitric oxide and vasoreactivity tests yielded negative results. Ductal closure occlusion test was applied, but pulmonary artery pressure did not change anyway, so transcatheter closure procedure failed. Subsequently bosentan treatment was initiated. Control angiography was planned six months later. Before, and after reversibility tests performed with nitric oxide: PVR, and PVR/SVR ratios were measured 8.8 Wood units  $m^2$  vs PVR 6.3 Wood units  $m^2$ , and PVR/SVR 0.67, vs, PVR/SVR 0.45, and application of transcatheter

closure of PDA was decided. The most suitable device for the closure procedure was thought to be Amplatzer® muscular VSD occluder device (AGA Medical Corporation, Golden Valley, MN, USA) with two retention discs. Since the patient had systemic PHT, the device was fenestrated with a 10F sheath catheter. The device was advanced transvenously through within 12 F sheath and placed in PDA via antegrade approach. During control injection left-to-right shunt was observed through fenestration. Aortic, and pulmonary artery pressures were 118/71 (86) mmHg, and 72/40 (50) mmHg, respectively, and the device was freed. The procedure was terminated without any complication (Figure 1b). Control echocardiograms obtained the following day revealed that the device remained in situ, and mild degree of left-to-right shunt persisted without narrowing left pulmonary artery or coarctation of descending aorta. The patient hadn't any complaints as detected at polyclinic controls of the patient at post procedural sixth month, and first year. On echocardiographic examination of the patient who didn't receive medical treatment, estimated pulmonary artery pressure via induction of mild degree of median pulmonary artery pressure was 35 mm Hg, and a slight displacement of the ductal occluder device used from left to right was detected.



**Figure 1. (A)** Angiogram obtained after Injection of contrast material with the patient laid in 90° lateral decubitus position demonstrates a wide patent ductus arteriosus. **(B)** Angiogram following ductal closure with Amplatzer® muscular VSD occluder device Ao: Aorta; PDA: Patent ductus arteriosus, MPA: Main pulmonary artery.

## DISCUSSION

Transcatheter closure of patent ductus arteriosus was firstly realized using Ivalon device in the year 1967.

This procedure had been the first interventional treatment procedure in pediatric cardiology.<sup>[2]</sup> For this procedure many coils as Gianturco coil, Duct-Occlud, and Nit-Occlud, and multiple number of devices as Amplatzer PDA occlusion device may be used. Closure of patent ductus arteriosus through percutaneous route using occluders is an effective, and safe treatment modality with higher success rates, and lower possibility of developing complication, and hence it is preferred over surgical treatment in recent years.<sup>[13]</sup>

In advanced- aged patients with wide PDA, and pulmonary hypertension because of the risk of device embolization use of classical ductal occluders may be risky. Publications on application of Amplatzer® muscular VSD occluder or ASD occluder with two retention discs with lower risk of embolization, and higher procedural success rates are available, and successful results have been reported with off-label use of these devices.<sup>[4-6]</sup> In older children with Down syndrome, irreversible PHT may develop.<sup>[7]</sup> In older people with significant PHT, and positive reversibility tests, application of transcatheter closure has been reported as a safe, and effective procedure with lower risk relative to surgical treatment. In patients with irreversible PHT ductal closure may lead to the development of right ventricular insufficiency. In the catheterization laboratory to evaluate whether status of pulmonary hypertension, and the patient's health is suitable for the procedure, application of vasoreactivity, and balloon occlusion tests is an appropriate approach. In patients with negative test results, bosertan, and iliprost may be used to prepare the patient for transcatheter closure procedure or surgical treatment.<sup>[9]</sup> In our elder patient in addition to wide PDA, systemic secondary PHT was detected, so priorly drug treatment was applied for PHT. Then during control cardiac catheterization, reversibility test positivity was detected, and proceeded with transcatheter closure procedure. For the procedure Amplatzer® muscular VSD occluder device was preferred. On echocardiographic examination pulmonary artery pressure of the patient with bidirectional shunt was measured in the catheterization laboratory, then occlusion test was performed with a device closing the defect. Since an expected drop in pulmonary artery pressure could not be

achieved, the device was fenestrated before its application and a small residual left-to-right shunt was left.

In patients with pulmonary hypertension following transcatheter closure, postprocedural complications as device embolization or malposition, bleeding, endocarditis, left pulmonary artery stenosis, aortic coarctation, progressive pulmonary vascular disease, hemolysis, and residual shunt may occur.<sup>[2-4]</sup> In a study by Zabal et al. 168 patients with PHT who had undergone transcatheter ductal closure procedure were evaluated, and in conclusion, effectiveness, and safety of this treatment method were emphasized. In 18 patients Amplatzer® muscular VSD occluder device was used, and the authors indicated that this device can be preferred in wide PDA patients with higher pulmonary artery pressure.<sup>[4]</sup> We didn't observe development of any complication in our case, and manifestations of PHT regressed.

In conclusion, in advanced aged patients with wide PDA, and significant PHT, before transcatheter closure reversibility, and balloon occlusion tests should be performed. In these patients instead of using devices manufactured for ductal closure, use of devices with fenestrated double retention skirts may decrease complication, and increase procedural success rates.

***Conflict of interest: None declared***

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*Anahtarsözcükler:* Muscular ventricular septal defect occluder cihazı; patent duktus arteriozus; pulmoner hipertansiyon; transkate- ter kapatma.

*Keywords:* Muscular ventricular septal defect occluder device; patent ductus arteriosus; pulmonary hypertension; transcatheter closure.