

Revision of Fontan to TCPC in a patient undergoing repair of subaortic stenosis

Subaortik stenozun tamiri sırasında Fontan'ın TCPC'ye dönüştürülmesi

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Subaortic stenosis is a complication which develops in the follow-up period of patients with univentricular atrioventricular connection to a dominant left ventricle when the ventriculoarterial connection is discordant. Nine-year-old male patient presented with cyanosis. Physical examination revealed 5/6 degree systolic murmur on the left parasternal border. The patient underwent Fontan operation with diagnosis of tricuspid atresia, right ventricular hypoplasia, transposition of the great arteries and ventricular septal defect at the age of four and subaortic stenosis developed afterwards. Ventricular septal defect was enlarged to relieve subaortic obstruction, and revision of Fontan to extracardiac conduit total cavopulmonary connection was performed at the same time. Contegra bioprosthesis was used for conduit. The patient was discharged without any complication.

Key words: Aortic stenosis, subvalvular/diagnosis/surgery; Fontan procedure; transposition of great vessels/surgery.

Subaortic stenosis is a time-manifested complication in patients with left-side dominant univentricular heart and ventriculo-arterial discordance. The degree of stenosis will rise with narrowing of ventricular septal defect (VSD), which worsen the functional status of the patient. The obstruction usually manifests itself with a decrease in effort capacity with time. Although there is no apparent difference in early outcome between the extracardiac conduit total cavopulmonary connection (TCPC) and the conventional modified Fontan, TCPC has superior hemodynamic efficiency and less complication related to the right atrium like dysrhythmias in long-term follow-up.^[1] Therefore revision of Fontan to extracardiac conduit TCPC provide potential benefits in late outcome during enlargement of VSD operation.

Tek ventrikül fizyolojisinde, dominant sol ventrikül ve ventrikülo-arteryel diskordansı olan hastalarda ameliyat sonrası gelişen komplikasyonlardan biri subaortik stenozdur. Dokuz yaşındaki erkek hasta siyanotik bir durumda getirildi. Fizik muayenede sternum sol kenarında 5/6 derece sistolik üfürüm saptandı. Hasta dört yaşındayken triküs pit atrezisi, sağ ventrikül hipoplazisi, büyük arter transpozisyonu ve ventriküler septal defekt tanılarıyla Fontan ameliyatı geçirmiş, ameliyat sonrası dönemde subaortik stenoz gelişmişti. Subaortik stenozun giderilmesi amacıyla ventriküler septal defekt genişletildi ve eş zamanlı olarak önceden yapılmış olan Fontan, ekstrakardiyak konduit ile total kavo-pulmoner konneksiyona dönüştürüldü. Konduit olarak Contegra biyoprotezi kullanıldı. Hastada ameliyat sonrası dönemde herhangi bir sorun olmadı.

Anahtar sözcükler: Subvalvüler aort stenozu/tanı/cerrahi; Fontan prosedürü; büyük damar transpozisyonu/cerrahi.

CASE REPORT

A 9-year-old boy with New York Heart Association Class III was admitted to our clinic. He had cyanosis. There was a 5/6 degree systolic murmur on the left parasternal border on physical examination. He had tricuspid atresia and right ventricular hypoplasia with ventriculo-arterial discordance and a large VSD on echocardiography in neonatal period. He underwent pulmonary artery banding and atrial septostomy operation on second month to prepare a single ventricle repair. After 4 years a Fontan (atrio-pulmonary connection) operation was performed, which includes connection of right atrium to pulmonary artery. Restrictive VSD with 40 mmHg gradient was determined between right and left ventricles on echocardiography.

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Ventricular septal defect narrowed spontaneously as time passes, which was large at birth, and subaortic stenosis then becomes apparent. Systolic pressure of hypoplastic right ventricle was measured as 90 mmHg, systemic left ventricular pressure was 130 mmHg and the systolic pressure in the ascending aorta was determined as 72 mmHg on cardiac catheterization. It became clear that the subaortic stenosis was caused by restrictive ventricular septal defect (Fig. 1). Revision of Fontan to extracardiac TCPC was decided to relieve the cyanosis and to improve the effort capacity. Ventricular septal defect was enlarged with resection of the antero-superior rim, the previous Fontan operation was revised to extracardiac conduit TCPC using bovine jugular vein



Fig. 1. Catheterization image of subaortic stenosis caused by restrictive ventricular septal defect in preoperative period.

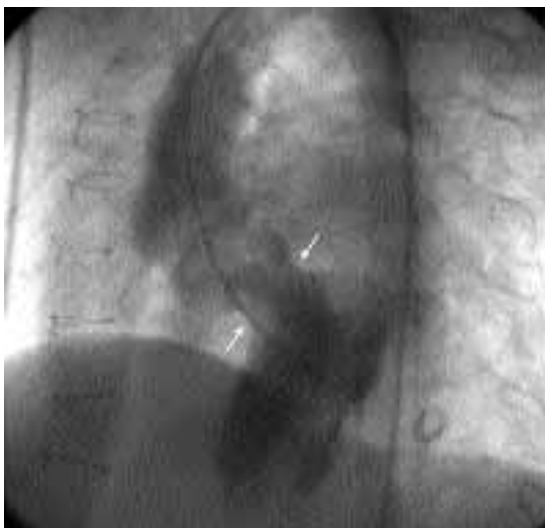


Fig. 2. Relief of subaortic stenosis with enlargement of ventricular septal defect in postoperative period.

xenograft (Medtronic's Contegra, Medtronic, Inc, Minneapolis, Minn) no: 20 without fenestration. Left ventriculo-aortic continuity through enlarged VSD was seen on left ventriculography in postoperative period (Fig. 2). The patient was extubated in 6 hours; central venous pressure was measured 20 mmHg. He was discharged without any complication. There was no gradient between the right and left ventricle and subaortic stenosis on control echocardiography. Extracardiac conduit TCPC was demonstrated on control cardiac catheterization (Fig. 3).

DISCUSSION

The mechanisms of subaortic stenosis after Fontan operation are related to hypertrophy of the subaortic component that incorporates the infundibulum and trabeculae. Careful morphological investigation is essential in patients with risk factors for subaortic stenosis. Mild deterioration of the ventricular compliance due to progressive subaortic stenosis may result in reduction of the cardiac function and patient's quality of life. Adequate surgical treatment to relieve the stenosis is considered essential after the Fontan operation.^[2] Enlargement of the VSD including antero-superior rim resection, can provide satisfactory long-term relief with univentricular atrioventricular connection to a dominant left ventricle, atrioventricular discordance and subaortic stenosis.^[3] Heart block is a rare complication of rim resection. There was not any conduction problem in our case.

Damus-Kaye-Stansel (end-to-side pulmonary trunk-aortic anastomosis) is another approach to relieve the subaortic obstruction. The aorta is brought into direct and wide communication with the large or main or dominant chamber.^[4]

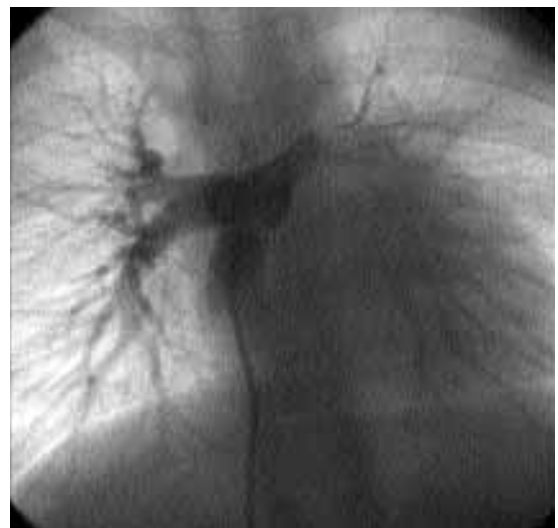


Fig. 3. Contegra bioprosthesis for extracardiac conduit total cavopulmonary connection.

Fontan revision to extracardiac or intra-arterial conduit total cavopulmonary anastomosis can be performed with success, and sometimes with dramatic improvement, in properly selected patients with complications referable to right atrium-pulmonary artery or modified right atrium-pulmonary artery connection, such as thrombosis, pulmonary venous obstruction, or arrhythmias. Revision may also be beneficial in patients with no complications directly related to the right atrium-pulmonary artery connection but with other indications for operation. Indications for Fontan revision to extracardiac TCPC in our patient was decline of the effort capacity and increase of the cyanosis with time. In the failing patient who both lacks complications specifically related to the right atrium-pulmonary artery connection and has other specific indications for operation (e.g., bulboventricular foramen obstruction, atrioventricular valve regurgitation), revision to total cavopulmonary anastomosis may be ill advised. At present, such patients with severe exercise intolerance, effusions or ascites, and protein losing enteropathy are not considered candidates for revision.^[5]

Recent studies focusing on the pattern of infradiaphragmatic venous return in patients undergoing the Fontan procedure have shown that forward (antegrade) flow into the pulmonary circulation is significantly reduced in the upright position because of increased reversal of flow into the IVC and the hepatic veins caused by gravity.^[6] These findings might at least partly explain subnormal exercise tolerance in patients undergoing the Fontan procedure. The use of a valved conduit in the Fontan procedure was found to have no major complications attributable to the valve. Baslaim and coworkers presented 4 TCPC cases using Contegra conduit.^[7] The Contegra bioprosthesis consists of a heterologous bovine jugular vein with a trileaflet venous valve and natural sinuses. The advantages of Contegra xenograft are easy tailoring and suturing, easy availability, different size options, and the presence of a valve in the conduit. Our report is the second presentation in English literature of the use of Contegra xenograft in extracardiac TCPC.

In conclusion, revision of the failing Fontan shunt to extracardiac conduit TCPC can be performed with sig-

nificant improvement in most patients and low mortality (10%) and morbidity. Optimal selection criteria for conversion have yet to be determined, but most patients can be expected to benefit to some extent. Revision of functioning Fontan circuit to extracardiac conduit TCPC provides more potential advantages in long-term period with superior hemodynamic status which improves effort capacity of the patient. Furthermore extracardiac conduit TCPC has less right atrial complication. Therefore, patients with functioning Fontan shunt, undergoing operation with another indication expect better long-term survival rates with less complication with revision of Fontan to extracardiac conduit TCPC. The use of a valved conduit may be beneficial in a long-term period with decrease of reversal flow into the IVC caused by gravity. Contegra bioprosthesis has superiority in surgical handling, easy availability and includes a venous valve which decreases reversal flow caused by gravity.

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