LETTERS TO THE EDITOR

Obstruction of the right pulmonary veins after the modified Fontan operation

To the Editor:

We read with great interest the report by Kreutzer and associates[1] in the June issue of the Journal. They described their experience with conversion of Fontan circuits into cavopulmonary anastomoses (CPA) with a lateral atrial tunnel. In their group of eight cases substantial relief of symptoms was encountered only in patients with obstruction to the right pulmonary veins by a grossly dilated right atrium. Improvement in arrhythmias or protein-losing enteropathy did not occur.

Our experience with conversion of a modified Fontan circuit into a CPA with a lateral atrial tunnel consists of four cases. Obstruction of the right pulmonary veins by a grossly dilated right atrium was diagnosed before the operation in two. In both cases the obstruction was diagnosed with the aid of transesophageal echocardiography (TEE). TEE demonstrated compression of the right pulmonary veins on the two-dimensional images. Obstruction was confirmed by color flow mapping and pulsed Doppler studies. In the third case the right pulmonary veins were only intermittently narrowed. The narrowing occurred in early diastole and coincided with a peak of accelerated flow on the Doppler tracing of the pulmonary vein. Our fourth patient was operated on before the introduction of TEE in our department but, in retrospect, probably had pulmonary venous obstruction as well. We were not aware of the possibility of right pulmonary venous obstruction at that time, and catheterization data failed to reveal it. However, despite a stenosis in the left pulmonary artery and a widely patent right pulmonary artery, pulmonary perfusion scans showed 60% of the pulmonary blood flow to pass through the left lung.

All patients were treated with the creation of a lateral atrial tunnel, consisting of polytetrafluoroethylene* in three and right atrial appendage tissue in one.[2] A fenestration in the tunnel was made in only one case. Concomitant procedures included relief of pulmonary arterial stenosis in two patients and creation of a ventricular septal defect to a blind right ventricle in one. The latter patient was also the one with the fenestration in the tunnel. She required prolonged ventilatory and inotropic support and was discharged in rather poor condition with marked cyanosis. Improvement in her condition was not achieved until another reoperation was performed to close the fenestration and the ventricular septal defect to the now aneurysmal right ventricle. Recovery after the second reoperation has been rapid, and her exercise tolerance is far better now than before the conversion to a lateral atrial tunnel. The three patients without fenestration in the tunnel had a remarkably uneventful hemodynamic recovery without effusions, and all show improved circulation and exercise tolerance on follow-up (2 to 6 years). However, atrial arrhythmias that were present before the conversion to a lateral atrial tunnel persisted afterward. None of our patients in this small series had chronic effusions or protein-losing enteropathy.

We assume that obstruction to the right pulmonary veins is not the exception in patients who have had a modified Fontan operation. TEE with Doppler flow mapping is a powerful tool to evaluate residual problems after a Fontan procedure[3] and can reliably diagnose pulmonary venous obstruction.[4] TEE may be superior to magnetic resonance imaging in identifying intermittent narrowing of the right pulmonary veins. The latter finding is possibly an early stage of pulmonary venous obstruction, in which the compression occurs only at the point of maximal right atrial dilation. We recommend the regular use of TEE in the follow-up of all patients after modified Fontan procedures.

We strongly endorse the conclusion of Kreutzer and associates[1] that patients with proven obstruction to the right pulmonary veins by a grossly dilated right atrium are likely to benefit from conversion to CPA with a lateral atrial tunnel. The mortality of this operation is low in selected cases.

Nynke J. Elzenga, MD[a]
Gertie C. M. Beaufort-Krol, MD[a]
Tjark Ebels, MD[b]
Division of Pediatric Cardiology[a]
Divison of Cardiothoracic Surgery[b]
University Hospital Groningen
Groningen, The Netherlands

*Gore-Tex patch and tube, registered trademark of W. L. Gore & Associates, Inc., Newark, Del.

REFERENCES

1. Kreutzer J, Keane JF, Lock JE, Walsh EP, Jonas RA, Castaneda AR, et al. Conversion of modified Fontan procedure to a lateral atrial tunnel. J Thorac Cardiovasc Surg 1996;111: 1169-76.
2. Ebels T, Elzenga NJ, Brenken U, Barns JL, van Os J, Eijgelaar A. Right atrial auricle as inter-caval tunnel in the total cavo-pulmonary connexion: method to avoid the sinus node and its blood supply. In: Minami K, editor. Cardio-thoracic surgery. Amsterdam: Elsevier, 1992:9-16.
3. Stümper O, Sutherland GR, Geuskens R, Roelandt JR, Bos E, Hess J. Transesophageal echocardiography in evaluation and management after a Fontan procedure. J Am Coll Cardiol 1991;17:1152-60.
4. Ren WD, Nicoloii GL, Lestuzzi C, Canterin FA, Golia P, Gervesato E. Role oftransesophageal echocardiography in evaluation of pulmonary venous obstruction by paracardiac neoplastic masses. Am J Cardiol 1992;70:1362-6.

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