Successful Modified Fontan Procedure in an Adolescent After Left Pneumonectomy

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SUMMARY. We performed a Waterston's anastomosis on a patient with complicated tricuspid atresia when she was two months of age. At age 14 years she required left pneumonectomy for massive, idiopathic hemoptysis. Four months after the pneumonectomy we substituted a modified Fontan anastomosis for the Waterston anastomosis. Restudy when she was 17 years of age showed continued satisfactory function.

KEY WORDS: Tricuspid atresia — Fontan procedure — Waterston anastomosis — Hemoptysis — Pneumonectomy

The Fontan procedure and modifications thereof have been used since the early 1970s as a definitive procedure for patients with tricuspid atresia, other forms of hypoplastic right heart syndrome, and various types of single ventricle [1, 3-5]. Original guidelines for the operation included age, anatomy, physiology, prior surgery, etc. [2]. As the procedure was increasingly done, many of these initial guidelines were found to be less than absolute. At the present time, the major factors which appear to influence the results of the Fontan procedure include the following: size and integrity of the pulmonary vascular bed, pulmonary vascular resistance, ventricular function, and atrioventricular valve function. We present herein an adolescent who had a modified Fontan performed using the right pulmonary artery following a left pneumonectomy for severe hemoptysis.

Case Report

A 17-year-old black woman was restudied 3 years after placement of a modified Fontan anastomosis between the superior vena caval/right atrial junction and the right pulmonary artery. She was noted to be cyanotic at birth and catheterization revealed tricuspid atresia, severe pulmonary stenosis, and both atrial and ventricular septal defects. A Waterston anastomosis was performed at the age of 2 months with a good rise in oxygen saturation and moderate congestive heart failure. Subacute bacterial endocarditis occurred on two occasions at 9 and 10 years of age, both successfully treated without clinical deterioration. A positive tuberculin conversion was treated with isoniazid for 1 year. By the age of 14 years, she was moderately limited in physical activity and remained on digoxin and diuretics. She then had her first episode of moderate hemoptysis and was transferred to Tisch Hospital for further evaluation and treatment. A cardiac catheterization done at that time (Table 1) revealed that the ventricular septal defect had closed and there was functional pulmonary atresia, as well as anatomic tricuspid atresia. The Waterston shunt was open to the right lung with moderate pulmonary hypertension; the pulmonary vascular resistance was at the upper limits of normal and the atrial defect was nonrestrictive. No left pulmonary artery could be found. This was thought to be due to probable kinking at the site of the Waterston anastomosis followed subsequently by loss of perfusion to the left pulmonary artery from the heart when the ventricular defect closed. Ventricular function was good and there was no atrioventricular valve regurgitation. While in the hospital for further observation, she had a massive hemoptysis and was taken to the operating room. A clot was found in the left main bronchus and when removed, there was fresh bleeding which could not be localized or controlled. A left pneumonectomy was therefore performed and her postoperative course was unremarkable. Bronchiectasis probably related to the tuberculosis was thought to be the cause for the hemoptysis, since no markedly dilated bronchial collaterals were found. The left upper lobe was markedly fibrotic as was a segment of the left lower lobe. There was no active tubercular infection. The right lung, fed by the Waterston anastomosis, continued at risk for the development of pulmonary hypertensive changes and the patient was readmitted 4 months after the pneumonectomy for a modified Fontan procedure. At operation, a communication was fashioned between the right pulmonary artery and the superior vena caval/right atrial junction. No pericardial or prosthetic material was used. The atrial defect was...
closed and the Waterston anastomosis was taken down. At the end of the operation, the right atrial pressure was 10–12 mmHg. Cardiac function was good and normal sinus rhythm resumed. Atrial flutter occurred in the early postoperative period and was controlled with digoxin and quinidine. There were no other problems. One month after discharge, she developed an upper respiratory infection with a cough and had another episode of hemoptysis, bringing up about 50 ml of bright red blood. She was immediately readmitted to Tisch Hospital but no further bleeding occurred. Bronchoscopy showed a normal right bronchial tree. There was mild tracheitis with inflammation but no bleeding site was found. She was discharged on maintenance digoxin and quinidine and she presently considers herself to be asymptomatic. She was restudied prior to entering college. On examination, vital signs were normal with no clubbing, cyanosis, or edema. There was no organomegaly or ascites. A grade 2/6 systolic flow murmur was present along the upper left sternal border with no radiation. Catheterization (Table 2) revealed a normal cardiac output with a right atrial mean of 12 mmHg and a right pulmonary arterial mean of 10 mmHg. There was no obstruction at the anastomosis and there was no shunt. The systemic circuit was normal with a left ventricular end-diastolic pressure of 8 mmHg, good contractility, and very minor mitral regurgitation.

### Discussion

Although the Fontan procedure does not result in an anatomically normal heart, it does restore near-normal physiology with obliteration of the right-to-left shunt. The original limitations suggested for the procedure [2] have been much modified with children under 4 years, as well as adults now having the operation. Dysrhythmias per se do not appear to be an absolute contraindication [3]. Although patients with a single ventricle of the right ventricular anatomic type seem to do less well than those with a left ventricle, this is not a contraindication for attempting the Fontan [7]. Significant atrioventricular valve regurgitation can be lessened by repair or replacement of the valve as long as ventricular function is adequate [3]. Hypoplasia of one pulmonary artery has also not prevented successful use of the Fontan procedure as long as pulmonary vascular resistance is normal [6]. In our patient, the presence of some degree of pulmonary hypertension, thought to be due to the shunt flow since the pulmonary vascular resistance was near normal, did not negatively influence the results of the operation, in spite of all the cardiac output going into only one lung. Although our patient may still be at risk to the late development of complications, she has remained problem-free since the early atrial flutter was controlled. Other patients, thought to be better candidates for a Fontan, have been plagued with recurrent effusions, edema, and protein-losing enteropathy, despite seemingly unobstructed, adequate operations [4, 5, 7]. The reasons for this ambiguity of clinical results still remain to be elucidated. Although arteriovenous fistula formation in the lungs may occur following the Fontan procedure as it has after a Glenn anastomosis, this has not yet been identified in our patient.

### References

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