# Fontan Conversion Still An Effective Alternative

#### BY BRUCE JANCIN Denver Bureau

SAN DIEGO — Fontan conversion accompanied by arrhythmia surgery and pacemaker implantation remains a safe and effective alternative to cardiac transplantation for patients with failing Fontan circulation, Dr. Constantine Mavroudis reported at the annual meeting of the Society of Thoracic Surgeons.

During the past several years patients with failing Fontans have been presenting at an older age, with more complex lesions and more-difficult-tomanage atrial arrhythmias as the increasing popularity of transcatheter ablation procedures has delayed surgical referral.

Yet results remain excellent due to evolution in surgical techniques and advances in pacemaker therapy, said Dr. Mavroudis, the Willis J. Potts Professor of Surgery at Northwestern University and surgeon-in-chief at Children's Memorial Hospital, Chicago.

His retrospective study examined 111 consecutive patients who underwent atriopulmonary to total cavopulmonary artery extracardiac Fontan conversion at the hospital since late 1994.

The patients' mean age was 23 years, with a mean 14-year interval between Fontan and Fontan conversion. Fourteen patients had undergone prior Fontan revisions.

Dr. Mavroudis divided the experience into three periods based upon changes in arrhythmia surgery techniques.

The first epoch consisted of simple isthmus cryoablation, a strategy abandoned after nine patients.

The next 51 had right atrial maze procedures for right atrial reentry tachycardia.

The most recent 51 patients—those treated since 2003—have routinely received the more elaborate biatrial Cox maze-III procedure, which incorporates cryoablation of the left atrium. This change occurred in response to a shift in the predominant presenting arrhythmia from right atrial reentry tachycardia to more challenging cases of atrial fibrillation.

The classic Cox maze-III was supplemented with one additional cryoablation lesion running between the bases of the right and left atrial appendages and across the dome of the atria to reduce the incidence of postoperative atrial tachycardia.

There were one early and six late deaths among the 111 patients. Six patients required cardiac transplantation, with two of the six late deaths in the series coming 4 and 24 days post transplantation. The other four donor heart recipients are alive 5-7 years later. With follow-up extending to 12 years, 88% of patients have experienced an improvement in New York Heart Association functional class. The arrhythmia recurrence rate was 13.5% overall, declining to just 8% in the most recent group comprised of 51 Cox maze-III-treated patients.

Postoperative arrhythmia recurrence in patients with preoperative refractory atrial fibrillation took the form of atrial tachycardia, Dr. Mavroudis continued, which is far more easily treated.

In a multivariate analysis, the strongest risk factor for death or transplantation was protein-losing enteropathy, present preoperatively in three patients and associated with an 87-fold increased risk.

A right or ambiguous ventricle and preoperative moderate to severe atrioventricular valve dysfunction also predicted poor outcome.

Discussant Dr. Joseph A. Dearani praised Dr. Mavroudis for heading



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DR. MAVROUDIS

what the congenital heart disease surgery community recognizes to be the world's premier Fontan conversion program.

"The most important contribution from their experience is the thorough understanding of the different atrial arrhythmias that occur in the failing Fontan circulation and their methods of ablation, which have evolved over time," observed Dr. Dearani, a cardiothoracic surgeon at the Mayo Clinic, Rochester, Minn.

"The importance of the need to address both atria at the time of operation cannot be overemphasized," he added.

Noting that the late deaths and cardiac transplantations in the Chicago series all occurred relatively early—within a year after Fontan conversion—Dr. Dearani asked whether it might make more sense to consider protein-losing enteropathy, significant atrioventricular valve regurgitation, and severe ventricular dysfunction to be contraindications to Fontan conversion and instead send affected patients directly to heart transplantation.

Dr. Mavroudis, however, answered that he would not yet include atrioventricular valve regurgitation as a contraindication. This is because his recent experience using Alfieri valvuloplasty has, so far, been very encouraging.

## Fetal Intervention Aids Outcomes In an Uncommon Heart Defect

### BY BRUCE JANCIN Denver Bureau

SAN DIEGO — Fetal intervention appears to boost midterm survival of children with hypoplastic left heart syndrome and an intact or highly restricted atrial septum, Dr. Vladimiro L. Vida said at the annual meeting of the Society of Thoracic Surgeons.

Nine of 14 such patients at Children's Hospital of Boston who underwent fetal intervention were alive at 6 months, compared with 6 of 16 in whom surgeons waited until after

birth to perform urgent atrial decompression. That translates to a 6-month survival rate of 64% in the fetal intervention group compared with 37% in patients who received what was until now considered state-of-theart care starting imme-

diately postnatally, noted Dr. Vida, a pediatric cardiac surgery fellow at the hospital.

Fetal intervention consisted of balloon atrial septectomy or aortic balloon dilation. The purposes were to eliminate the immediate postnatal cyanosis that accompanies this form of congenital heart disease, achieve circulatory stability, and promote normal vascular and parenchymal development. The hypothesis was that this would improve both shortand long-term outcome in patients with this otherwise very-poor-prognosis abnormality, the physician explained.

Based upon the favorable experience to date, Dr. Vida recommended encouraging referral for fetal intervention in all patients identified prenatally as having hypoplastic left heart syndrome (HLHS) with an intact atrial septum by fetal echocardiography, or with poor flow or reversal of flow in the pulmonary vein.

Outcomes in HLHS have improved in the last decade, but the subgroup with an intact or highly restrictive atrial septum has lagged behind. Fortunately, it is not a common defect. The 30 affected patients Dr. Vida reported on were among 230 consecutive HLHS patients managed at Children's Hospital of Boston in a period beginning in January 2001. Twenty-five of the 30 were diagnosed prenatally as having an intact atrial septum, so surgeons were standing by to perform transcatheter atrial septectomy to create an interatrial communication immediately after birth in most of those who didn't undergo fetal intervention.



median age of 4 days. A modified Blalock-Taussig shunt was utilized for this purpose in 26 of 30 patients. Delayed sternal closure was required in 24. Operative mortality was 30%, and five additional patients died in the interstage period. Both operative and interstage mortality were lower in patients who had undergone fetal intervention.

Five patients required early revision of their Blalock-Taussig shunts—four because of poor circulation and one because of shunt thrombosis.

Early shunt revision, need for postoperative

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DR. VIDA

support using extracorporeal membrane oxygenation, a longer cardiopulmonary bypass time, and the presence of pulmonary venous pathology were associated with increased risk of in-hospital mortality, Dr. Vida continued.

Discussant Dr. Jeffrey P. Jacobs hailed Dr. Vida's report as "truly groundbreaking." Soon-to-be-published data provided to him by colleagues at Children's Hospital of Philadelphia underscore the extremely high risk of HLHS with an intact or highly restrictive atrial septum when managed conventionally that is, starting postnatally, added Dr. Jacobs of the University of South Florida, Tampa.

Thirty-day survival of patients in the Philadelphia series was just 38%. Moreover, there was no difference in outcome between those diagnosed pre- and postnatally. Survival wasn't improved by a strategy involving early aggressive postnatal intervention based upon prenatal diagnosis.



Surgical palliation was performed at a

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