

## **You Never Know**

By Jo Ann Simons

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For almost 25 years, I lived with a false sense of security. When my son, Jonathan Derr, was born in 1979 with Down syndrome and Tetralogy of Fallot, I was reassured that his heart defect could be corrected. I turned over Jon's cardiac care to the team of talented professionals dedicated to his care and devoted myself to understanding DS and doing everything to maximize Jonathan's potential. I even altered my career choice and forged a professional path in the disability field.

Jonathan had successful cardiac surgery as a toddler and grew healthy and strong. He became a successful athlete and excelled academically and socially. His cardiac care turned into bi-annual visits. Occasionally I forgot to schedule the visit and he went three or four years between cardiac visits.

So it was with much surprise that we learned in October 2003 that the results of a cardiac MRI demonstrated the need for a pulmonary valve replacement. The cardiologist told us that while surgery was inevitable, it was not immediately necessary. It was Jon who decided to have the surgery "while I was healthy." We supported Jon's decision, since waiting would not improve either the outcome or the surgical technique. The MRI result, together with earlier test results showing that Jonathan had high cholesterol, led me to decide that care from an adult congenital heart disease team was appropriate. So we switched cardiologists.

Surgery was scheduled for December 1, 2003, and Jon began to be seen by the Boston Adult Congenital Heart group at Children's Hospital. Prior to surgery Jon underwent a bolter test and an exercise tolerance test. Both tests indicated ventricular tachycardia. During uneventful surgery, Jon had his pulmonary valve replaced and a cryoablation to address the ventricular tachycardia.

Prior to discharge, Jonathan had a positive ventricular stimulation study. The members of his cardiac and electrophysiology teams thought that the test results merited an implantable cardiac defibrillator. Jon's physicians discussed at length with all of us the lack of absolute data and the data that did exist. While none of us wanted him to have to go under general anesthesia again, or to live with the uncertainty of having a defibrillator, we decided to support the team's recommendation and go ahead with implantation. Jonathan was discharged 10 days after his valve surgery for a two-day break to enjoy the comforts of our

home. The defibrillator implantation was successful.

We are grateful to the teams of physicians and nurses at Boston's Children's Hospital for Jonathan's treatment and recovery. I am haunted, however, by the unknown numbers of Tetralogy of Fallot patients who are as unaware as I was of the need for aggressive and competent follow-up care as they become adults.