

Tetralogy of Fallot – Introduction

SURGICAL THERAPY FOR TETRALOGY OF FALLOT IS one of the success stories of the early era of cardiac surgery. Despite the many advances of the ensuing fifty years, such as new techniques for imaging, extension of surgical therapy to the neonatal period, and improved treatment in the intensive care unit, there has been an increasing recognition that the outcomes for many patients over the long term remain less than optimal. Mortality for surgical correction of tetralogy of Fallot, even in very small neonates, approaches zero at most institutions. Patients remain at risk, however, for right ventricular dysfunction, congestive heart failure, and an ongoing need for future therapeutic interventions, including cardiac catheterization and re-operation. Surgical reconstruction of the abnormal subpulmonary right ventricular outflow tract often results in pulmonary insufficiency and scarring, leading to ventricular volume overload and dysfunction. Postoperative right ventricular dysfunction may be associated with congestive heart failure, poor exercise tolerance, arrhythmias, and sudden death. The chapters which follow in this section of our supplement focus on the current state of knowledge concerning treatment and management of patients with pulmonary insufficiency following surgical repair of tetralogy of Fallot. There is considerable controversy concerning the clinical significance of pulmonary insufficiency; there is no standard technique for the assessment of right ventricular function; and there are considerable disagreements concerning the optimal timing of surgical re-intervention to replace the pulmonary valve. I believe that the reviews will help clarify these important issues.

In the first article, Bob Anderson and Paul Weinberg discuss the clinical anatomy of tetralogy of

Fallot. Their report focuses on the specific morphologic criteria for diagnosis, as well as identification of the significant variations within the phenotypic framework. Bob Boucek and Rick Martinez present the current “state of the art” for echocardiographic assessment of right ventricular function. This is of critical importance in the determination of the optimal time for replacement of the pulmonary valve, as well as for assessment of outcomes following surgery. Anthony Chang discusses the time course of right ventricular adaptation to progressive insufficiency of the pulmonary valve, as well as medical and surgical strategies to improve right ventricular function before and after surgery for replacing the valve. Jim Quintessenza and his colleagues discuss current surgical options for replacing the pulmonary valve. In addition, they introduce their new surgical technique, which uses a prosthetic bicuspid valve made of polytetrafluoroethylene. This section of our supplement concludes with a debate between me and Carl Backer, in which we address the issue of aggressive versus conservative surgical intervention for patients with pulmonary insufficiency following surgical repair. I am sure that there will be something within the pages that follow that will prove of interest to all our readers.

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