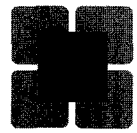

Congenital heart diseases and coronary artery anomalies: a comparison of experiences

THE CARE OF CHILDREN with congenital cardiac malformations has improved, and corrective cardiac surgery has been extended to a wider array of anomalies in neonates and infants. For the surgery to be successful, the coronary arterial patterns and variants in cardiac malformations must be understood and recognized.

In three conditions in particular—transposition of the great arteries, tetralogy of Fallot, and pulmonary atresia with intact ventricular septum—the coronary arterial pattern has significance. In transposition of the great arteries, the conventional surgical approach using an atrial switch procedure (e.g., Mustard procedure) is being replaced by one using an arterial switch procedure. In the latter operation, performed in neonates, the aorta and pulmonary trunk are each transected and reconnected to the opposite ventricle. A major component of the operation is to remove the origins of the coronary arteries from the posterior wall of the aorta that arises from the right ventricle and to place the origins into the anterior wall of the newly constructed aorta that arises from the left ventricle. An abnormality of the origin or course of the coronary arteries[1–4] complicates the latter part of the procedure and may contradindicate performance of an arterial switch.

■ *See Tuzcu and associates (pp 147–152)*

In tetralogy of Fallot, corrective operations are preferred to palliative procedures. In 2% to 10% of patients with this condition,[5–7] the anterior descending coronary artery arises from the circumflex artery. In its course to the anterior interventricular groove, the anterior descending artery passes across the right ventricular outflow area. Correction, involving incision in the outflow area, cannot be performed in such patients because of the coronary arterial pattern.

Finally, patients with pulmonary atresia and intact ventricular septum, especially those with a hypoplastic right ventricle, have coronary arterial anomalies including stenosis or atresia of the proximal left coronary artery.[8,9] As long as pulmonary atresia maintains right ventricular systolic pressure, coronary perfusion is maintained by blood exiting the right ventricular myocardial sinusoids into the coronary arterial system. After the obstruction is relieved, right ventricular pressure falls and myocardial ischemia ensues in those patients with coronary obstructive conditions.

A knowledge of the origin and course of the coronary arteries is necessary to properly manage a cardiac malformation. Cardiologists and radiologists must apply the knowledge of the coronary arterial system and its variants to correctly image and identify the coronary arteries of infants and children being considered for surgery. The cardiac surgeon must use the information about the coronary arteries to determine what surgical procedure is appropriate or how to modify an existing procedure in accordance with the anatomic variations from normal.

The study by Tuzcu and associates published in this issue of the *Cleveland Clinic Journal of Medicine* adds to our understanding of coronary arterial anomalies associated with cardiac malformations. This study is an extension of one published in 1982 of 38,703 patients studied by coronary arteriography at the Cleveland Clinic.[10] Among this large group, 601 (1.55%) were found to have a congenital coronary arterial anomaly, the most common being separate origins of the circumflex and anterior descending branches (182 patients) and a circumflex artery arising above the right sinus of Valsalva (176 patients). In the current study, 1,000 congenital anomalies were found among 66,884 coronary arteriograms (1.5%). Of the 1,000, 101 (10%) had a coexistent cardiac malformation.

Tuzcu and associates' 101 cases can be divided into three categories: cases in which the coronary anomaly was not expected (51 cases), cases in which a coronary arterial anomaly would be expected because of the nature of the associated cardiac malformation (11 cases with corrected transposition and 10 cases with univentricular heart), and mitral valve prolapse (29 cases). There is question whether mitral valve prolapse should be considered a congenital cardiac anomaly.

In Tuzcu and associates' current study, 5% of patients had a congenital coronary anomaly that was not specific for a particular cardiac malformation. The study does not provide data on the incidence of coronary anomalies in patients with a congenital cardiac malformation.

Some information on incidence can be obtained from reviewing the data of the Northern Great Plains Regional Cardiac Program, a registry system of 20 pediatric cardiac centers. Data are available from 12,867 children with a congenital cardiac malformation who have undergone cardiac catheterization, cardiac surgery, or died of a cardiac condition. In this data base of 12,867 children there were 604 (4.7%) with a coronary anomaly identified through coronary arteriography. Since coronary arteriography is an uncommon procedure in children, the frequency may well be higher. In 91 of the 604 there was a primary coronary artery anomaly, including 49 with anomalous origin of the left coronary from the pulmonary trunk, 34 with a coronary arteriovenous fistula, and 8 with other primary conditions. There were another 258, 91 with corrected transposition and 167 with a univentricular heart, who had the anticipated coronary anomalies associated with these respective conditions. Finally there were 255 (2%) with an unpredicted, coexistent coronary anomaly.

Most of the unpredicted coronary anomalies occurred in patients with cyanotic forms of congenital heart disease. Eighty-six of the 255 patients with unpredicted coronary anomalies were among the 1,187 (7%) of patients with tetralogy of Fallot, usually the left anterior descending artery arising from the right coronary artery. As others have reported, such anomalies occur in 2% to 10% of patients with this malformation.[5-7] In addition there were 29 other patients with pulmonary atresia and ventricular septal defect, an analogous condition, with coronary arterial anomalies.

As indicated previously,[1-4] coronary anomalies occur in transposition of the great arteries and must be recognized preoperatively. Twenty-six of 777 patients (3%) in our registry with transposition of the great arteries showed anomalies of origin of coronary arteries from the aorta.

While an uncommon condition, accounting for 1.5% of cases of congenital cardiac anomalies, pulmonary atresia with intact ventricular septum is often associated with coronary anomalies,[8,9] especially fistulous connections from the right ventricle to the coronary branches. Major coronary anomalies were described in 22 of 57 patients in the registry with a hypoplastic right ventricle.

Unlike Tuzcu and colleagues, we found few cases (6, or 1%) of stenosis of the left coronary artery among the 617 registry cases of aortic stenosis, and we found no cases of a short or absent left main trunk. In marked contrast, Tuzcu and associates identified this anomaly in 7 of 18 cases with bicuspid aortic valve. The reason for this difference is unknown but could represent a problem of ascertainment (e.g., requiring coronary arteriography) or of definition (What is a "short" main trunk?), or it could be related to the age differences in the populations.

In the study of Tuzcu and associates, 13 of 101 congenital coronary anomalies were classed as an arteriovenous fistula, and, among registry patients, 52 of the 255 cases had such a fistula. In 46 of the 52 a cyanotic form of congenital cardiac malformation was present. The reason for this association is unknown.

Neither Tuzcu and associates nor the Northern Great Plains Regional Cardiac Program has determined the incidence of coronary anomalies in patients with a cardiac malformation, because of study design. But both sets of data do indicate that coronary arterial anomalies may coexist with a wide variety of cardiac malformations, mostly with cyanotic forms; that the anomalies may have important diagnostic, therapeutic, or functional complications; and that they should be sought in all patients being imaged.

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