THE SURGICAL TREATMENT of most forms of congenital heart disease is a remarkable success story. Mortality figures are low even for complex conditions such as transposition of the great arteries and tetralogy of Fallot. Despite the immediate operative success, however, important residua, sequelae and late complications have been reported in the more common conditions (table 1).

In this issue of Circulation, the University of Minnesota pediatric and adult cardiology groups add another chapter to reports of abnormalities after surgery for congenital heart disease. They investigated 11 survivors of the Fontan procedure for treatment of tricuspid atresia. Their report illustrates three recurring themes in follow-up studies for postoperative congenital heart disease patients.

First, the surgical mortality is very low, with no perioperative deaths in their 16 consecutive patients. Second, clinical assessment of cardiac status (New York Heart Association classification) reveals that most patients are asymptomatic — nine of 13 (69%) in their series — or have only minimal symptoms. Third, with continued follow-up and postoperative investigations, significant residua, sequelae and complications become apparent (table 2). Of particular interest are the late deaths (two of 16 died suddenly, suggesting dysrhythmia), the low cardiac index at rest, and the abnormal response to exercise. Although the numbers of patients are small, the message seems clear: In this group of postsurgical patients, cardiac performance is suboptimal and the incidence of sudden death (possibly due to dysrhythmia) is high. These latter problems (dysrhythmia and abnormal cardiac performance) apply to postoperative tetralogy and transposition patients and suggest that similar mechanisms may play a role in such abnormalities in these cyanotic patient groups (table 3).

How do the data regarding the Fontan operation guide the physician attempting to provide optimal care to his or her patient with tricuspid atresia? As with most therapy for complex congenital heart disease, the answers are not clear. In certain subsets of patients, particularly those who fulfill Fontan’s criteria for an optimal candidate for his procedure, the postoperative results appear to be good. These ideal patients, however, would probably also do exceptionally well with a shunt procedure. There are no data that compare similar subsets of tricuspid atresia patients after a Fontan vs a shunt or multiple shunts in terms of longevity, morbidity and cardiovascular performance. The many unanswered questions in the treatment of tricuspid atresia patients (table 4) indicate the need for detailed investigations and careful analysis of long-term follow-up data for patients after operations for congenital heart disease.

For future follow-up studies for postoperative patients, important areas of concern are listed in table 5. First, detailed quantitative preoperative data must be available for comparisons to allow proper assessment of perioperative events that could influence outcome. Second, perioperative data must be as detailed as possible. With recent improvements in methods for intraoperative myocardial protection, many studies showing postoperative alterations in ventricular function or dysrhythmia may not be applicable and probably should not be exclusively used in current decision-making regarding therapy until corroborative evidence is available on patients operated upon with current techniques for ischemic protection of the heart during surgery. Third, detailed postoperative investigations should be performed and, in most patients, should be delayed for at least 6-12 months after repair unless signs and symptoms indicate the need for earlier study. Cardiovascular performance should be assessed both at rest and with exercise or afterload stress.

In obtaining these postoperative data, who should have a postoperative catheterization? The answer continues to change. Echocardiography and radionuclide angiography provide much of the information needed for a complete postoperative evaluation. However, further correlative studies are needed to compare catheterization and noninvasive assessments for different subsets of postoperative patients in order to clarify which patients will require catheterization for a comprehensive evaluation.

What are some of the unresolved problems still facing the physician who treats patients with congenital heart disease? A partial list is shown in table 6. There is still much to be learned.

Finally, in dealing with complex congenital heart disease, postoperative abnormalities may be inevitable in conditions in which reparative or palliative rather than corrective more properly describes the operative options. The task at hand for surgeons, pediatric cardiologists and adult cardiologists is to continue to evaluate treatment practices critically with extended longitudinal follow-up and detailed postop-
TABLE 1. Residua, Sequelae and Late Complications After Surgery for Congenital Heart Disease

<table>
<thead>
<tr>
<th>Preoperative diagnosis</th>
<th>Residua, sequelae, complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>Open VSD with CHF; elevated Rp; AV block; ventricular dysfunction</td>
</tr>
<tr>
<td>Endocardial cushion defect</td>
<td>Mitral, tricuspid regurgitation; elevated Rp; residual shunt; AV block; atrial dysrhythmia</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>Residual or recurrent coarctation; aneurysm; aortic regurgitation</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>Residual or recurrent stenosis; aortic regurgitation; ventricular dysfunction; altered ventricular distensibility</td>
</tr>
<tr>
<td>Transposition of the great arteries (atrial repair)</td>
<td>Atrial dysrhythmia and conduction disturbances; ventricular dysfunction; caval obstruction; pulmonary venous obstruction; increased Rp; residual shunt; pulmonary stenosis; tricuspid regurgitation</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Ventricular dysrhythmia; AV block; aneurysm of RV outflow tract; VSD; residual or recurrent stenosis; pulmonary regurgitation; ventricular dysfunction; tricuspid regurgitation; increased Rp (usually in patients with prior shunt procedure)</td>
</tr>
<tr>
<td>Pulmonary atresia plus ventricular septal defect, truncus arteriosus, transposition post–Rastelli repair</td>
<td>All of those listed for tetralogy plus conduit stenosis</td>
</tr>
</tbody>
</table>

Abbreviations: VSD = ventricular septal defect; CHF = congestive heart failure; Rp = pulmonary vascular resistance; AV = atrioventricular; RV = right ventricular.

TABLE 2. Postoperative Residua and Sequelae in Tricuspid Atresia Patients After the Fontan Operation

<table>
<thead>
<tr>
<th>Residua or sequelae</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>&quot;Late&quot; death</td>
<td>3/16 (19%) died 2, 6, and 25 mos. after operation: one from sepsis, two sudden</td>
</tr>
<tr>
<td>Elevated right atrial pressure at rest</td>
<td>8/8 (100%)</td>
</tr>
<tr>
<td>Low cardiac index (&lt; 2.5) at rest</td>
<td>5/8 (63%)</td>
</tr>
<tr>
<td>Elevated pulmonary vascular resistance</td>
<td>2/8 (25%)</td>
</tr>
<tr>
<td>Marked elevation of right atrial pressure with exercise</td>
<td>5/5 (100%), mean pressure 23–30 mm Hg</td>
</tr>
<tr>
<td>Abnormal cardiac index response to exercise</td>
<td>3/5 (60%)</td>
</tr>
</tbody>
</table>

TABLE 3. Possible Causes for Abnormal Diastolic or Systolic Ventricular Function and Dysrhythmia in Patients After Surgery for Congenital Heart Disease

I. Secondary to the defect or defects

A. Hemodynamic overload — chronic hypertrophy — fibrosis
B. Intermittent myocardial oxygen supply-demand imbalance — fibrosis
   1. $O_2$ demands — wall tension, heart rate and $O_2$
      catecholamine stimulus during stress
   2. $O_2$ supply — wall tension; cyanotic patient — $O_2$ content, polycythemia — viscosity — coronary vascular resistance
C. Residual shunts, obstructive lesions, valvular insufficiency

II. Complications of treatment

A. Inadequate myocardial protection during operative interventions — ischemia — fibrosis
B. Inadequate myocardial protection during low output states: e.g., severe congestive heart failure pre- or postoperatively with low coronary perfusion pressure
C. Intraoperative coronary air embolus and myocardial ischemia
D. Altered coronary supply: tetralogy with injured or transected coronary artery, post-Jatene arterial switch for transposition
E. Ventricular scar from repair
F. Atrioventricular block and ventricular pacing

III. Acquired disease

A. Coronary artery disease
B. Myocardial disease: myocarditis, endocarditis with valvular or myocardial damage, cardiomyopathy
C. Hypertensive cardiovascular disease

operative investigations to try to minimize postoperative abnormalities and maximize both survival and quality of life for the increasing numbers of survivors of palliative or reparative operations.

References

TABLE 4. Unresolved Problems Regarding Surgical Treatment of Tricuspid Atresia

- Long-term fate of patients with Fontan procedure vs those palliated with shunts
- Subsets of patients who are better candidates for shunts? for Fontan? (with follow-up objective data documenting a better outcome)
- Optimal age for the Fontan procedure
- Optimal options for different subsets of patients having the Fontan procedure: conduit vs no conduit, valved or non-valved conduit, direct pulmonary artery anastomosis vs use of right ventricle, caval valves, Glenn procedure
- Incidence and timing of conduit obstruction (or outgrowth of conduit) requiring reoperation
- Optimal early palliation for symptomatic infants
- Use of ventricular septal defect enlargement as palliation
- Left ventricular functional abnormalities in different subsets of postoperative patients
- Rhythm abnormalities in different subsets of postoperative patients

TABLE 5. Assessing the Results of Surgery: Areas of Concern In Designing Investigations

I. Preoperative status
   A. Hemodynamic assessment: degree and duration of abnormalities
   B. Ventricular assessment: size, degree of hypertrophy, compliance, systolic function
   C. Cyanotic patients: degree and duration of hypoxemia

II. Perioperative data
   A. Intraoperative arrest time or aortic cross-clamp time
   B. Method of myocardial protection
   C. Postoperative assessment of cardiac function — intensive care unit
   D. Complications: hypotension, acidosis, arrest, ventilator time, inotropic requirements
   E. Rhythm or conduction abnormalities

III. Postoperative data
   A. Hemodynamic residua
   B. Rhythm problems — ambulatory monitor, exercise, electrophysiology studies in selected patients
   C. Resting and stressed ventricular function: exercise, afterload stress

TABLE 6. Unresolved Problems Regarding Postoperative Congenital Heart Disease Patients

| Problem Description                           | Degree and Duration of Abnormalities
|----------------------------------------------|-----------------------------------------------
| Transposition of the great arteries          | Progressive systemic ventricular dysfunction as adults? Increasing incidence of dysrhythmia with longitudinal follow-up? Morbidity and mortality of the arterial switch procedure?
| Tetralogy of Fallot                          | Cause and treatment for ventricular dysrhythmia? Ventricular function in patients repaired as infants? Contribution of outflow patch, pulmonary insufficiency to abnormalities of cardiac performance? Use of valved conduits to improve abnormal right ventricular function?
| Pulmonary atresia, intact ventricular septum | Fate of various-sized right ventricles with different operative therapy?
| Truncus arteriosus, pulmonary atresia with VSD, TGA after Rastelli | Ventricular dysfunction or dysrhythmia in long term survivors? Indications to replace conduit?
| Single ventricle                             | Fontan vs septation vs palliative shunts as optimal treatment for various subsets of patients? Ventricular dysfunction?

Abbreviations: VSD = ventricular septal defect; TGA = transposition of the great arteries.

Assessing the results of surgery for congenital heart disease: a continuing process.
T P Graham, Jr

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