11. CATHETERIZATION OF THE ADULT PATIENT WITH CONGENITAL HEART DISEASE

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Adult patients with congenital heart (ACH) disease are a product of medical and surgical advances of the second half of the twentieth century. A review at our own institution revealed that the 17-year survival of those born with all types of congenital heart disease was 84%. Despite the achievements of pediatric caregivers, many of these patients, particularly those with complex lesions, face a world not yet fully prepared for their adult care. Throughout the stages of their adolescent and adult years, congenital heart disease “survivors” present with problems often different from those encountered in their pediatric lives. Novel medical, surgical and catheter-based diagnostics and therapeutics have led to an increased use of the catheterization laboratory in the management of these patients. While simple defects predominated in the catheterization laboratory three decades ago, a marked shift toward more complex lesions was evident 20 years later. In the past decade however, with the advent of interventional procedures, simple lesions such as pulmonary stenosis, patent ductus arteriosus and atrial septal defect are once again being catheterized.

In this chapter we will:
1. Review the primary differences in presentation of the ACH patient for the catheterizing physician trained in the management of congenital heart disease patients.
2. Review the general principles of catheterization of ACH patients.
3. Outline the risks and needs for specific groups of ACH patients.
4. Briefly describe techniques, indications and results of catheterization-based management of particular lesions in the ACH patient.

1. THE ACH PATIENT

The ACH patients will, in general, manifest increasing and cumulative effects from complex palliative procedures such as modified Fontan operations, surgical scarring, insufficient myocardial preservation during prior surgery, uncorrected abnormalities in ventricular pre-load and after-load, alteration in red blood cell mass and tissue perfusion, and progressive tendency to decreased myocardial compliance with aging. These effects contribute to increasing systolic and diastolic ventricular dysfunction, and to the progressive incidence of arrhythmia seen in these patients, atrial enlargement, sluggish atrial flow and thrombosis, and development of atrial arrhythmia (see Chapter 14). In some patients, inadequately controlled pulmonary blood flow may lead to pulmonary vascular disease, in situ thrombosis, and after-load effects on pulmonary ventricular function.
Prior to, and during catheterization of ACH patients, one must consider coexisting morbidity that may be rarely seen in pediatric patients with congenital heart disease. Diabetes mellitus, obesity, systemic hypertension, hypercholesterolemia, peripheral- and cerebro-vascular disease, coronary atherosclerosis, chronic lung disease, chronic erythrocytosis, spinal disease, alcohol and drug use, and potential for pregnancy, all can influence patient well-being and safety during catheterization. A review of the potentials of these illnesses at the time of catheterization is beyond the scope of this text, though a few general principles are offered:

1. Catheterization of the ACH patient may reveal previously unrecognized anatomic or physiologic abnormalities, which will require intervention. Recognition of the catheterization laboratory as an “operating theater”, with commensurate institutional support and generalized, yet appropriate, patient consent is requisite.

2. Variation in systemic blood pressure is generally less well tolerated in the adult patient, and volume resuscitation or pressor support should be contemplated for either SBP $\leq 90$ mm Hg or a significant deviation from baseline. In particular, adequate hydration prior to study is very important in cyanotic polycythemic ACH patients. Support for acute hemodynamic compromise must be available, including appropriately sized mechanical ventricular and ventilatory support, and inhaled and intravenous pulmonary vasodilators.

3. Recent pre-catheterization assessment of lower extremity venous thrombosis in patients at increased risk (presumed paradoxical embolization, obesity, cigarette smoking, oral contraceptive use, peripheral vascular disease, raised systemic venous pressures or non-pulsatile systemic venous flow) may reduce potential for dislodgment and embolization, especially with use of large caliber sheaths and catheters. At the beginning of the catheterization an initial femoral – iliac – vena caval angiogram is recommended in all such patients.

4. When a transseptal study is performed, particular caution should be taken to avoid manipulation in the atrial appendage due to increasing risk of left atrial thrombus.

5. Long-standing erythrocytosis reduces glomerular filtration rate and increases viscosity, raising risk for contrast-induced acute tubular necrosis and vascular thrombosis.²

6. Prior to catheterization, a measurement of beta-HCG and a discussion of teratogenic risks of radiation is mandatory for ALL women.

2. GENERAL PRINCIPLES FOR CATHETERIZATION OF THE ACH PATIENT

Patient size, chamber dilation and vessel distortion present additional technical challenges that can be overcome with increased experience. Options for vascular access may be more varied (radial, brachial, axillary, internal jugular,