13. HOW TO CATHETERIZE SOME COMMON COMPLEX LESIONS

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1. SINGLE VENTRICLE (SV).
2. TETRALOGY OF FALLOT WITH PULMONARY ATRESIA (TOF - PA).
3. PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM (PA-IVS).

Following the dramatic advances in surgical and interventional catheterization techniques, prolonged survival is now common in patients with complicated lesions, some heretofore considered inoperable such as hypoplastic left heart syndrome (HLHS). In the course of management of 3 such complex lesions, namely SV, TOF - PA and PA - IVS, surgery and catheterization at planned intervals are necessary. In the past 10 years at our institution, among 6,441 patients (11,593 catheterizations) were 952 patients with SV, including patients with HLHS, tricuspid atresia (TA), corrected transposition with a hypoplastic right ventricle (SLL-hypoRV), malaligned common atrioventricular canal and a straddling atrioventricular valve with hypoplasia of a ventricle. The final surgical procedure, a modified Fontan approach is preceded by staged surgeries, catheterizations and interventional procedures. These patients underwent 2,318 catheterizations and 1,198 interventional procedures. There were also 517 patients with TOF-PA in this population (1,319 catheterizations and 873 interventional procedures), the goal in these being to achieve a biventricular repair with an adequate pulmonary arterial tree and 178 patients with PA-IVS (351 catheterizations and 207 interventional procedures) the desired end point in these being achievement of a biventricular repair and when not feasible a modified Fontan approach.

It is the purpose of this chapter, based on our experience to outline the catheterization methodology and interventional procedures involved in these 3 groups of patients at each of several stages. We recognize that medically indicated deviations from these schedules inevitably occur, yet the methodology, data acquisition and interventions required at these unscheduled studies are often similar to those necessary at the elective catheterizations.

1. SINGLE VENTRICLE (SV).

In this group surgery in the neonatal period is usually based on echocardiography alone; for example, a stage 1 Norwood procedure in HLHS or a pulmonary artery band placement in those with TA and unrestricted pulmonary blood flow. The subsequent surgical course in these patients usually involves a bidirectional Glenn and then a fenestrated Fontan, each preceded by a catheterization.

Cath 1: Pre Bidirectional Glenn (BDG).

This first catheterization is usually at 6 months of age just prior to a bidirectional Glenn anastomosis (BDG). In many, a single femoral venous line (5F) alone may be adequate to acquire all the necessary data - if however, aortic arch obstruction,
distal pulmonary artery stenosis or pulmonary artery hypertension is suspected, a small 3F pigtail is placed in a femoral artery. The **Physiological Data** required include saturations and pressures in superior vena cava (SVC), right atrium (RA), left atrium (LA), ventricle (V), pulmonary vein (PV), ascending and descending aorta (AO), a PV wedge pressure and an oxygen consumption to enable precise calculation of pulmonary artery resistance. If the PV mean wedge pressure exceeds 18-20 mm with a ventricular end diastolic pressure (VEDP) less than 10, then direct pulmonary artery (PA) measurement is mandatory. This latter is acquired via the arterial route using the 3F catheter with the pigtail cut to 180°. This catheter is first placed over a wire beyond the shunt origin in the subclavian artery, withdrawn slowly until the modified tip engages the proximal orifice. Next, an 0.018” torque wire (Mallinckrodt Medical, St. Louis, MO 63134) is advanced to a distal PA and the catheter advanced over the wire. It should be remembered that the mandril of this wire proximal to the floppy platinum coated tip is quite stiff and should thus be hand-shaped to conform to the aortic arch and shunt curves prior to insertion.

**Anatomical Details** necessary include visualization of the (L) innominate vein and any branches (such as an LSVC-CS/LA), RSVC, both (R) and (L) PA and their branches, (Fig. 13-1) all pulmonary veins (since occasionally one may drain anomalously or be stenotic), aortic arch, shunt origin course and insertion, ventricular size and function and atrioventricular valve competence.

**Figure 13-1**: Visualization of shunt (arrow) and pulmonary arteries after stage 1 Norwood procedure for hypoplastic left heart syndrome, injecting in innominate artery proximal to distal occluding balloon: catheter course antegrade.

**Interventional Possibilities**: If aortic arch obstruction is identified, this should be dilated, if possible using the antegrade route (Fig. 13-2). Distal PA stenoses should also be balloon dilated (Fig. 13-3) with either a retrograde or antegrade approach using small balloons available to 8mm diameter (Boston Scientific, Watertown, MA 02172: B. Braun, Bethlehem, PA 18018), which can be advanced through a 4F sheath. Any large A-P collaterals should be coil occluded as should an LSVC-CS/LA as the latter will likely enlarge following the BDG: it is important to occlude an LSVC-CS/LA on the cardiac side of the entrance of its azygos vein (see below).

**Cath 2: Post BDG Prior to Modified Fontan Procedure.**
This study is usually undertaken between ages 1 and 3 years. Vascular access involves placement of two venous lines (both 5F), one in a femoral vein and the other in the (L) subclavian vein. A femoral arterial line (4F) is also required.

**Physiological Data** required include saturation values from SVC, (R) and (L) PA, atria, PVs, V and AO. It should be mentioned that a step up in saturation may be encountered especially in the (L) upper lobe PA due to A-P collateral flow.