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Diagnostic Cardiac Catheterization in Infants and Children

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The mission of the pediatric catheterization laboratory continues to evolve. Although the total number of patients catheterized per year has not changed a great deal in the past several years, the proportion undergoing an interventional procedure continues to increase, from 6% in 1984 to 68% in 1997 (Fig. 6.1).

Echocardiography has greatly influenced the catheterization laboratory patient profile. The remarkable diagnostic precision of this tool has essentially eliminated the need for preoperative diagnostic catheterization in many of the common congenital lesions, such as tetralogy of Fallot, total anomalous pulmonary venous return, and endocardial cushion defects (1)(2). On the other hand, echocardiography identifies lesions, formerly undetected by other noninvasive means, that may be now therapeutically managed in the laboratory, such as a “silent” patent ductus arteriosus (PDA) and a small atrial septal defect (ASD). In addition, it is being used more frequently, particularly in the transesophageal mode, during interventional procedures such as septal device placement (3).

Although obtaining hemodynamic and/or diagnostic data was the sole reason for study in some 400 patients (approximately one third of our total catheterizations), these data remained “part and parcel” of most of the other studies as well, being used for example to accurately quantitate results of interventions. This chapter presents our current catheterization methodology, including information on sedation, anesthesia, and vessel access. In addition a few specialized procedures such as ASD creation, transseptal puncture, endomyocardial biopsy, coronary angiography, and pulmonary vein wedge angiography are described. Details of interventional techniques for management of lesions such as ASD, ventricular septal defect (VSD), PDA, stenotic valves, and collateral vessels are presented in Chapters 28 and 34.

CATHETERIZATION PROTOCOL

General

Patient Population

At least 80% of our population come as outpatients, fasting, for their studies, all necessary precatheterization tests having been completed in the preceding days. Older patients are given nothing by mouth after midnight, and infants are given clear liquids up to 3 hours before the procedure. Patients with a hematocrit greater than 50% are admitted the preceding day and adequate hydration is maintained via intravenous fluids. All patients have an intravenous line placed on arrival. Because of the very high incidence of interventional procedures, each patient has an assigned nurse throughout the study. Anesthesiologists provide general anesthesia for some 25% of the studies and medication advice and supervision for many of the others. At the conclusion of the study, many of the patients are discharged the same day, including all noninterventional cases except for PDA occlusion (4) and a few older pulmonary stenosis balloon valvotomy patients. All available patient information is discussed in some detail by the catheterizing physicians, including the anesthesiologist, immediately before the study, and procedure plans are then formulated.

FIG. 6.1.

Number of cardiac catheterizations per year, 1984 through 1997, at Children's Hospital, Boston, with the increasing proportion of interventional procedures indicated by the shaded areas.

Sedation and Anesthesia
Significant changes have occurred over the past decade, and continue to be made, in sedation and anesthesia administration, largely because of the increasing proportion of interventional studies. Approximately 80% of patients are studied using “conscious” sedation; the remainder are intubated and receive general anesthesia from the outset. There are some in whom sedation alone is insufficient during the procedure, who then require general anesthesia.

Sedation

This modality is used for all noninterventional studies; for balloon dilation of pulmonary and aortic valvar stenosis, coarctation, most pulmonary artery stenoses, and most stent placements; and for coil occlusion of aortopulmonary collaterals and PDAs. For those patients weighing 10 kg or less, intravenous morphine (0.025 to 0.05 mg/kg) and midazolam (0.025 to 0.05 mg/kg) are given, and additional doses may be given during the procedure to a maximum total for each of 0.4 mg/kg. In patients weighing 10 to 25 kg, demerol compound (thorazine 6.25 mg/mL, phenergan 6.25 mg/mL, and demerol 25 mg/mL) is given subcutaneously for premedication to a maximum of 2 mL, the dose based on weight and arterial oxygen saturation (5), (6). During the procedure if additional sedation is necessary, midazolam and morphine (each 0.025 to 0.05 mg/kg IV) may be given to a maximum total for each of 0.3 mg/kg. If Demerol compound premedication is not used initially, then midazolam alternating with morphine using the above dosages may be given to a total for each of 0.4 mg/kg. For those patients weighing more than 25 kg, midazolam may be used for premedication and, alternating with morphine, during the study at the above dosage levels. If these are unsatisfactory, droperidol, 0.025 mg/kg IV over 15 to 30 minutes, may be given twice.

Anesthesia

Although this sedation technique is referred to as “conscious sedation” in adults, in our experience most of the patients are in fact "unconscious"—that is, they often exhibit no response on needle insertion for local anesthesia. Respiratory depression is not uncommon, and frequent blood gas monitoring is mandatory. General anesthesia from the outset of the procedure is undertaken in all those in whom placement of a septal double-umbrella device is planned (7), (8). It is also used for those with bilateral peripheral pulmonary stenosis with right ventricular systolic pressure at or above systemic arterial level when balloon dilation is planned, particularly if concomitant left-sided heart obstruction is present, and in patients with severe congenital mitral stenosis. In addition, all those to undergo mapping and ablation electrophysiologic procedures receive general anesthesia.

This is a very complex and complicated technique. It requires management by an experienced anesthesiologist and is beyond the scope of this text. In brief, drugs such as morphine and fentanyl (analgesics) or thiopental and propofol (anesthetics) are used for induction, and inhaled nitrous oxide, oxygen, and isoflurane are used throughout the study together with a muscle relaxant such as pancuronium. Many of these agents have hemodynamic ill effects, although some, such as remifentanil, are considered free of these (9).

Catheterization Study

Although most lesions today have been correctly identified by physical examination and noninvasive techniques before the study begins, setting out with an open mind rather than simply trying to prove the existence of what one already thinks is present is extremely important. Physicians who catheterize “with blinders on” miss important diagnoses.

Catheters are introduced percutaneously into the femoral vessels in the great majority of cases at the level of the superior ramus of the pubis, using the smallest catheters that are adequate to obtain the necessary information. Before the infusion of any heparin or heparinized flush solution, an activated clotting time (ACT) is measured and heparin (100 IU/kg) is then administered intravenously to a maximum of 5,000 IU, with additional doses to maintain the ACT at greater than 200 seconds throughout the study. The initial phase of the procedure is aimed at gathering physiologic data. A right-sided heart catheterization is performed, measuring oxygen saturation in the superior vena cava and recording pressures and saturations in the midlateral right atrium (RA); inflow and outflow portions of the right ventricle (RV); main, right and left pulmonary arteries (PA); and pulmonary capillary wedge positions. The atrial septum is explored and, if traversed, pressures and saturations are recorded in the left atrium (LA) at least one pulmonary vein, and left ventricle (LV). The catheter is then repositioned in a distal PA branch. An arterial catheter, placed in most cases, is used to record pressure, oxygen saturation, and blood gas values in the descending and then
ascending aorta. The arterial catheter is advanced to the LV, oxygen saturation is measured, and pressure is recorded simultaneously with pulmonary capillary wedge pressure. Oxygen consumption is measured using a flow-through metabolic rate meter (Waters Inc., Rochester, MN), as right and left heart chamber pressures and saturations are recorded during catheter withdrawal for use in computation of pulmonary and systemic flow and resistance values. In the absence of intracardiac shunting, cardiac output may also be assessed by thermodilution (10).

The next phase of the study consists of angiography with particular emphasis on appropriate patient positioning for optimum visualization of specific cardiac anatomy. If no other data collection or procedures are contemplated, the renal shadows are viewed under fluoroscopy (11), the catheters are removed, and bleeding at the entry sites is controlled with a minimum of 15 minutes of local pressure. In our experience, protamine sulphate administration has rarely been necessary, at most once per year. We have, however, encountered rare episodes of pelvic bleeding due to arterial catheter entry proximal to the pubic ramus, some of which have required surgical intervention, manual vessel compression after catheter removal being ineffective in such a setting.

Other Vascular Entry Sites

Umbilical Vessels

In the neonate, venous access is feasible for some 3 days after birth. Anatomically, the umbilical vein enters at the bifurcation of the portal vein, opposite to which the ductus venosus arises, the latter then coursing posteriorly to open into the inferior vena cava. This is a tortuous course, approximately 180° in the lateral view, and catheter passage is more safely accomplished in the laboratory where angiographic delineation aids catheter entry (Fig. 6.2). Use of a tip deflector system (Cook, Bloomington, IN) facilitates catheter negotiation to the heart and also from RA to RV. At our own institution, a survey several years ago involving multiple operators revealed successful entry for 70% of neonates in the first day of life, 56% in the second, and 43% in the third. Although catheterization of right heart structures is more difficult with this technique than from a femoral venous approach, access to left heart structures, including the aorta, is relatively easy. The umbilical vein is commonly used for balloon atrial septostomy (see later discussion). The umbilical artery is traversible longer than the umbilical vein, occasionally to age 10 days. Anatomically, this vessel turns acutely posteriorly and distally to join the iliac artery, and a further 90° turn by the catheter is then necessary to reach the descending aorta. Nevertheless, passage is surprisingly easy, and this route has provided a means for arterial pressure and saturation monitoring, aortography, and even balloon dilation of critical valvar aortic stenosis (12).

FIG. 6.2.

Posteroanterior (A) and lateral (B) views outlining contrast course via umbilical vein to portal vein (open arrow), then via ductus venosus (closed arrow) to inferior vena cava.

Transhepatic Venous Route

In patients in whom femoral venous channels are occluded and those with congenital inferior vena cava interruption, a transhepatic venous approach has been increasingly used both for diagnostic and interventional procedures (13-15). We use the expertise of our radiology colleagues to help place these lines. After local anesthesia, a long 22-gauge needle is inserted in the midaxillary line, approximately halfway between the diaphragm and the inferior liver edge, and advanced with the use of ultrasound guidance and contrast injections into a large central hepatic vein. A sheath and dilator are then advanced over a guidewire to the RA. In addition to acquiring diagnostic data, this route has been used for interventional procedures such as transseptal studies and placement of double-umbrella devices in atrial defects using sheaths as large as 11F even in children. At the conclusion of the procedure, the hepatic tissue channel is occluded with coils during final catheter withdrawal. This procedure has been surprisingly uncomplicated, the most frequent problem being transient abdominal discomfort, presumably due to peritoneal irritation.

Internal Jugular Vein
This vessel, usually the right, large even in infants, is frequently used for RV biopsies, in double-umbrella device closure of apical muscular VSDs, in patients with bidirectional Glenn shunts, and in electrophysiologic studies. The ipsilateral arm is placed at the patient's side, and the head is turned to the opposite shoulder. A catheter in the vessel from a femoral venous approach, if available, provides an excellent guide for needle entry. The percutaneous needle, after local anesthesia, is advanced in the depression between the two heads of the sternomastoid, above the medial end of the clavicle, in a direction posterior to the latter, while gently aspirating. A wire, sheath, and dilator are then introduced. Complications have been surprisingly few, consisting mainly of local hematomas or inadvertent entry of the more medially located carotid artery.

**Subclavian Vein**

This vessel is commonly used in patients of all ages, even in babies weighing 3 kg (16). It is necessary where thrombosis of iliac veins or infrarenal inferior vena cava has occurred after prior catheterization. In addition, palliative surgical procedures, such as the bidirectional Glenn procedure, which consists of anastomosis of the cephalad end of the superior vena cava with the PA, make this approach necessary to reach the PA. We prefer the left subclavian vein, largely because later catheter manipulation seems easier. The patient is first placed in a slight Trendelenburg position, a small rolled-up towel is placed lengthwise under the thoracic spine, and the patient's left arm is positioned at the side. Lidocaine is injected down to the periosteum at the junction of the clavicle and first rib. A 19-gauge, short, bevelled, thin-walled needle is used to enter the skin at the junction of the medial and middle thirds of the clavicle in adults, or 1 to 2 cm lateral to that point in infants and children (17). The needle is advanced toward the suprasternal notch, while maintaining an orientation that is both perpendicular to the spine and parallel to the floor as the needle tip passes between the clavicle and first rib, with continual aspiration of the syringe. Once venous blood returns freely, a preformed guidewire is inserted and advanced to the superior vena cava, and an appropriate-sized sheath with a backbleed valve is then inserted. At the conclusion of the study, the sheath is removed and bleeding is controlled with at least 15 minutes of gentle pressure on the first rib. Outlining the vessel by prior contrast injection in the left hand is particularly helpful. In addition to acquisition of hemodynamic and angiographic data, this approach has been of considerable value in reaching distal PAs when the RV is large and in placement of PA stents.

**EQUIPMENT USED IN CATHETERIZATION**

**Catheters**

Based on issues of patient safety, teaching responsibilities, and cost, balloon-tipped flow-directed transvenous catheters are used initially in all age groups. These may be either end-hole or side-hole (angiographic) in type and range from 4F to 7F in diameter. Acute bends may be shaped by hand just before insertion, especially with the smaller catheters. Deflector wires may be used within these catheters to help attain access to desired sites (e.g., LV from LA, aorta from LV). For arterial studies, ultrathin-walled white Teflon pigtail catheters ranging from 3.2F to 7F are used (18),(19); they provide satisfactory contrast flow rates (e.g., up to 35 mL at 35 mL/sec through a 6F, 80-cm catheter). Using a Y adapter (USCI, Billerica, MA), satisfactory pressures and angiograms (up to 20 mL at 20 mL/sec) are obtainable even with a 0.035-inch guidewire in place through a 7F pigtail catheter (20).

**Contrast Material**

Currently only the osmolar (nonionic or dimeric) contrast materials are used, their cost having decreased substantially in recent years. We make every effort not to exceed a total dose of 5 mL/kg for the entire study.

**Radiographic Equipment**

Because of contrast agent constraints and the necessity of acquiring as much anatomic information as possible from each injection, biplane equipment is essential for pediatric catheterization. Given the broad range in patient age and size, a wide range of image intensifier modes is necessary, from 5 inches for neonates to 12 to 14 inches for older patients in whom it is necessary to image the PAs and larger cardiac chambers. Precise patient positioning is essential...
to provide optimum visualization of anatomy (21),(22). The most common views used are the long axial oblique (for VSD, Fig. 6.3), the right anterior oblique (for subaortic stenosis, Fig. 6.4), the four-chamber or hepatoclavicular (for endocardial cushion defect, Fig. 6.5), and the sitting up or cranial angled view (for central PA anatomy, Fig. 6.6). All cineangiograms in our institution are now recorded with the use of digital enhancement and stored in a RAID (Redundant Array of Independent Discs) system and on individual compact disks.

FIG. 6.3.

Long axial oblique (analogous to 30° left anterior oblique, 30° cranial) view of left ventricular cineangiogram in an infant with a membranous ventricular septal defect (arrow).

FIG. 6.4.

Subaortic stenosis, right anterior oblique view, discrete (A, arrow) and diffuse muscular (B, arrow).

FIG. 6.5.

Four-chamber or hepatoclavicular view of endocardial cushion defect with common atrioventricular valve (dashed line) in atrial systole and ventricular septal defect (arrow) in an infant.

FIG. 6.6.

MPA cine, with cranial angulation (“sitting up” view) with (L) anterior oblique, in postoperative patient with tetralogy of Fallot showing bilateral pulmonary artery stenosis, severe (arrow), at the original of the left pulmonary artery.

SPECIAL PROCEDURES

Atrial Septostomy

Since the introduction of balloon atrial septostomy (BAS) by Rashkind and Miller in 1966 (23), this procedure has been the standard initial therapy for infants with d-transposition of the great arteries. This congenital anomaly, in which the pulmonary and systemic circuits are in parallel rather than in series, often results in severe hypoxia and acidosis immediately after birth. In this setting, BAS improves bidirectional mixing at the atrial level, resulting in an immediate rise in arterial oxygen saturation with alleviation of acidosis. This procedure is also of value in neonates with left atrial outflow obstruction (e.g., mitral atresia), because an initial atrial septal opening frequently narrows within weeks of birth.

Vascular entry is obtained either by way of the umbilical vein or percutaneously from a femoral vein. The septostomy catheter we most commonly use is the 5F Miller-Edwards catheter (American Edwards Laboratories, Santa Ana, CA), which requires a 7F sheath for insertion to allow passage of the unrecessed balloon. For the umbilical venous approach, we use a sheath with a valve (back-flow adapter) to avoid air embolism. The catheter tip is advanced to the middle LA, using biplane fluoroscopy. The balloon is held against the atrial septum and inflated rapidly (to a maximum of 4 cc). The catheter is then advanced 1 or 2 mm before being pulled briskly to the junction of the inferior vena cava and RA, advanced to the RA, and then rapidly deflated. This sequence is usually repeated at least twice to ensure that an adequate atrial septal opening has been created. It results in a rapid rise in arterial blood oxygen saturation, evidence of bidirectional shunting at the atrial level, and abolition of the interatrial pressure gradient. Alternatively, this procedure can be carried out at the bedside under two-dimensional echocardiographic guidance. Complications are extremely rare, but tears of pulmonary veins and atrial walls have occurred.
The BAS technique just described is usually ineffective after 1 month of age and in those neonates with very thick atrial septa. In such patients, the atrial septum is crossed in a location other than the patent foramen with a transseptal needle and sheath. Then, via the sheath a guidewire is passed, preferably to a left pulmonary vein, after which a succession of angioplasty balloon catheters of increasing size are inflated in the newly created atrial hole. An alternative method, now uncommonly used, consists of placing a catheter with a retractable blade (24) via a sheath into the LA. Blade lengths of 9.4, 13.4, and 20 mm are available (Cook). The blade is opened carefully in the LA under biplane fluoroscopic monitoring and then withdrawn slowly across the atrial septum. This procedure is followed by a balloon septostomy to further enlarge the resulting opening. If oxygen saturation rise and pressure gradient reduction are inadequate, the procedure is repeated using a larger blade and/or balloon catheters until the mean residual left-to-right atrial gradient is less than or equal to 3 mm Hg (25).

**Transseptal Left-Sided Heart Catheterization**

The availability of biplane fluoroscopy has significantly reduced the complication rate associated with this technique. In addition, introduction of the long Mullins sheath (USCI) has made transseptal catheterization easier and has broadened the indications for this procedure (26). Current indications include LA access for balloon valvotomy of mitral stenosis, LV catheterization in the presence of an aortic valve prosthesis, and device closure of VSD. We have not encountered any fatal complications related directly to this procedure.

Available equipment includes 6F and 7F long sheaths and needles of appropriate size. The sheath is introduced percutaneously by way of the right femoral vein, although we have used the left femoral vein after additional prebending of the distal needle shaft. We prefer to attach the needle lumen to a syringe filled with contrast material rather than a pressure transducer, to allow precise location of the needle tip after its initial extrusion. Pressure measurement alone via the needle tip can give misleading information about tip location. Availability of two-dimensional echocardiographic visualization during the procedure is also helpful when the atrial septal location is unusual.

**Selective Coronary Arteriography**

In general, ventriculography and aortography are sufficient to identify the major proximal coronary arteries in such conditions as tetralogy of Fallot, d-transposition of the great vessels, or double-outlet right ventricle. However, selective coronary injections are necessary in a variety of anomalies, including Kawasaki disease, coronary artery fistulae, and pulmonary atresia with intact ventricular septum, and frequently when origin of the left anterior descending artery from the right coronary artery is suspected. Borrowing from the enormous experience of our colleagues with adult patients, miniaturized catheters for use in pediatric patients, including infants, have been developed. We use a variety of such catheters, ranging from 4.5F to 7.3F, in Judkins or Amplatz configurations (Cook) with varying secondary loop sizes. We use standard techniques and angiographic views (see Chapter 11) and appropriately smaller doses of contrast material.

**Pulmonary Vein Wedge Angiography**

Some patients, as a result of a congenital defect or prior cardiac surgery, have complete occlusion of a proximal pulmonary artery, usually the left. Although an injection in the aorta or even in a collateral vessel may outline the size and location of the isolated PA, it is essential before any surgical reanastomosis is undertaken to identify unequivocally the presence and size of that PA. A balloon-tipped end-hole catheter is placed across the atrial septum and into a pulmonary vein in the appropriate lung. With the balloon inflated, up to 0.3 mL/kg of nonionic contrast agent (which causes less coughing than high-osmolar ionic agents) is injected, followed immediately by an equal volume of saline. The parenchymal vessels are usually well outlined by this method, with back-filling, if present, of the mediastinal segment. On occasion, the main and contralateral PA may also fill if they are in continuity. It is important to use biplane cineangiography for these injections to identify accurately the degree of proximal extension of the vessel relative to the bronchus on that side.

**Endomyocardial Biopsy**
The technique of endomyocardial biopsy has improved considerably in recent years (see Chapter 20). Many bioptomes, some as small as 3F, are now available, and together with preformed long sheaths they have made biopsy a rather safe procedure, even in newborns. The number of biopsies at our institution has increased markedly, mostly because of cardiac transplantation. The great majority of procedures involve right ventricular biopsy, although left-sided specimens can be obtained if necessary. Because the procedure is repeated frequently in patients with a cardiac transplant, a different venous percutaneous access site is usually used with each procedure. The vessels used are the internal jugular, subclavian, and femoral veins for right-sided biopsies. If left ventricular samples are required, the approach is transseptal in infants and retrograde from a femoral artery in older patients.

**SPECIAL CATHETER TECHNIQUES**

With continued widespread use of homografts to bypass stenotic or atretic lesions, particularly in the right side of the heart, obstruction over time is increasingly encountered. With the advent of balloon dilation and stents (27), localization of the exact sites of obstruction to make use of these therapeutic modalities is important. To this end, a very useful double-lumen balloon catheter (Figs. 6.7 and 6.8) is available with the ports 3 cm apart in the 7F size and 2 cm in the 6F size (28).

**FIG. 6.7.**

Simultaneous pressure tracings proximal (A) and distal (B) to obstructed valve in right ventricle to pulmonary artery homograft recorded using double-lumen 7F catheter.

**FIG. 6.8.**

Simultaneous pressure tracings in a 21-year-old patient with tricuspid atresia with right atrium=right ventricle valved conduit. **A:** No obstruction present at valve. **B:** Significant obstruction (*shaded*) evident between distal conduit and right ventricle.

A number of lesions amenable to balloon dilation, such as peripheral pulmonary stenosis and aortic coarctation, require measurement of pressures across the dilated areas together with angiography to assess the results of such therapy. To facilitate such studies safely, cutoff pigtail catheters with Y adapters are used over guidewires. A review of 737 such procedures found that satisfactory data were obtained with only one instance of wire dislodgment. Contrast volume and flow rates were determined using a variety of catheter and wire combinations (20).

Older patients with tetralogy of Fallot who had a transannular right ventricular outflow patch placed as part of their surgical repair frequently have right and/or left PA origin stenosis, very dilated RA and RV chambers, and both tricuspid and pulmonary regurgitation. From a femoral venous approach, it is often difficult to reach the distal PA branches for balloon/stent therapy. In many, an approach from the left subclavian vein provides a more stable catheter course. In addition, use of a large guiding sheath with its tip just proximal to the PA origins allows easier passage of dilating balloons and stents. Use of Amplatz coronary artery catheters also allows easier access to these PAs, especially the left.

**SUMMARY**

The past 15 years have seen substantial changes in the cardiac catheterization of infants and children, both as a diagnostic tool and particularly as an important therapeutic modality. The diagnoses (e.g., complex congenital heart disease), techniques (e.g., umbilical artery and vein access), patient management (e.g., deep sedation or even general anesthesia), and specific procedures (e.g., atrial septostomy) in this population are not part of the day-to-day world of the adult cardiac catheterization specialist. We hope that this overview of pediatric catheterization and the companion chapter on pediatric intervention (Chapter 28) will contribute to a more rounded overview of invasive cardiology.