Introduction

The use of catheters to investigate cardiac function dates back to the 19th century. Three early 20th-century pioneers in cardiac catheterization won the Nobel Prize for Medicine in 1956 (Werner Forssman, Andre Cournand, and Dickinson Richards). Forssman’s efforts cost him his job at the time, but Cournand and Dickinson went on to use right heart catheterization in a systematic effort to understand cardiac function. For many years diagnostic cardiac catheterization was the primary method for delineating the anatomy in congenital heart disease (CHD) as well as providing physiological data.

Interventional catheterization was first described in 1953 by Rubio-Alvarez to treat pulmonary stenosis. In 1968 Rashkind and Miller described balloon atrial septostomy for palliation of transposition of the great arteries, and in the last few years the number of transcatheter interventions has increased dramatically.

The history of anesthetic involvement in cardiac catheterization dates to the early 1950s. In 1953 anesthesia for angiocardiography at the Brompton Hospital was described using thiopentone and succinylcholine. The use of rectal thiopentone and intravenous meperidine, and rectal and intramuscular barbiturates were described in 1956. In 1958 Smith described the “CM3” mixture for sedation (chlorpromazine, demerol, and promethazine), which was widely used until recently.

Indications for diagnostic catheterization have become more limited in the last decade as other less invasive imaging techniques have become available. However, diagnostic catheterization is still required in selected cases to resolve the anatomy in complex patients and to make hemodynamic measurements. As the number of diagnostic catheterizations has diminished, the use of cardiac catheters for therapeutic purposes has increased. At the Hospital for Sick Children (HSC) in Toronto, interventional catheterizations currently outnumber diagnostic studies by two to one. Interventional procedures are used to avoid surgery, for lesions not amenable to surgical treatment (e.g. peripheral pulmonary artery [PA] stenosis) or to palliate patients and postpone surgical repair. As opposed to diagnostic catheterization the focus is on treatment, not precise diagnosis, and maintenance of baseline hemodynamics is less critical. Some procedures are performed in high-risk patients with the potential for serious complications, and are associated with major hemodynamic disturbances. Another group, electrophysiology (EP) procedures, include delineation and transcatheter ablation of abnormal conducting pathways and implantation of pacemakers. Anesthetic and sedative drugs have effects on cardiac conduction, and particular agents should be chosen to minimize this effect.

Environment

The cardiac catheterization laboratory is an inhospitable environment for the anesthesiologist (Fig. 25.1). It is often remote from the operating room, frequently undersized, of necessity not brightly illuminated, and filled with equipment that makes access to the patient difficult. Patients are frequently transferred some distance to recovery or intensive care facilities, and during transport the anesthesiologist must be satisfied that the patient is in a stable condition. The patient should be transferred with suitable monitoring, appropriate personnel and oxygen, resuscitation equipment, and drugs.

Radiation exposure is a hazard to both the patient and staff during cardiac catheterization. Non-stochastic effects such as erythema and cataracts are a direct result of cellular injury and are dose related. Stochastic effects are the result of injury to DNA. The risk of injury is increased with increasing dose (amount of energy absorbed); however, the magnitude of effect is not dose related. Exposure is measured in rem (radiation equivalent in man) or Sieverts (100 rem = 1 Sievert). Background exposure is 0.1 rem/year. The risk of a fatal cancer is increased by 0.04% per rem of life-time exposure, and thus no level of radiation exposure can be considered
Procedures are longer, more complex and more likely to involve interventions. Most children will not tolerate this without some pharmacological suppression of consciousness. The benefits of general anesthesia or sedation should be considered in relation to the individual patient and procedure.

Sedation may be administered without the presence of an anesthesiologist. Recognition that adverse events occur in association with this practice has lead to a series of guidelines being produced. The most recently revised (and most applicable) guidelines are those of the American Society of Anesthesiologists (ASA). Four levels of sedation and anesthesia are recognized: minimal, moderate, or deep sedation; and general anesthesia. A purposeful response to verbal stimulation can be produced during moderate sedation. During deep sedation only painful or repeated stimuli elicit a purposeful response. Sedation to a depth that no response or only reflex withdrawal can be produced is defined as general anesthesia. The distinction between deep sedation and anesthesia is often arbitrary, and in the UK, no distinction is made between the two.

The ASA guidelines specify that the individual supervising the sedation must be able to rescue the patient if the level of sedation enters the next level. To ensure that a potentially uncooperative child remains still during cardiac catheterization, deep sedation or general anesthesia is required. Therefore the individual supervising the sedation must have the skills to manage general anesthesia, to support the airway, ventilation, and cardiovascular system. The risk of major complications during cardiac catheterization and the physiological limitations of the catheter laboratory can be an inhospitable place for both the anesthetist and patient. X-ray and hemodynamic monitoring equipment limits access to the patient.

**Anesthetic considerations**

As early as 1958, a satisfactory state for pediatric cardiac anesthesia was described: (i) freedom from pain; (ii) absence of restlessness; (iii) no respiratory depression; and (iv) sedation light enough to allow a normal response to a selective ether test. While selective ether tests are no longer conducted, the requirements for cardiac catheterization at present are broadly similar. General anesthesia is not essential to achieve these aims; in adult patients it is routine to conduct cardiac catheterization with minimal or no sedation. However, cardiac catheterization is different in children. Procedures are longer, more complex and more likely to involve interventions. Most children will not tolerate this without some pharmacological suppression of consciousness. The benefits of general anesthesia or sedation should be considered in relation to the individual patient and procedure.

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status of the patients require the presence of an individual who is skilled in resuscitation other than the operator. Deep sedation supervised by a non-anesthesiologist sometimes occurs, but this situation does not provide optimum care for children.

Inadequate preoperative assessment and inadequate monitoring contribute to poor outcome during sedation. Assessment and monitoring of patients undergoing cardiac catheterization, whether under sedation or general anesthesia, should be comparable to that of a patient undergoing any other operative procedure. A means of monitoring adequacy of ventilation and the airway is mandatory; oxygen masks or nasal cannulae can be adapted to allow monitoring of end-tidal carbon dioxide.\(^\text{10}\)

The risk of airway obstruction increases as the level of consciousness decreases. Access to the airway is limited during the procedure, and intubation of the trachea and control of ventilation provide the safest option in most patients, unless it is desirable to avoid positive pressure ventilation (e.g. during diagnostic procedures in some patients with Fontan physiology). Total intravenous anesthesia can be achieved in many patients without specific support of the airway, although care and very close observation is required. In patients who develop airway obstruction, the laryngeal mask airway (LMA) is well tolerated and allows maintenance of the airway. In spontaneously breathing patients, intubation of the trachea requires deeper anesthesia and produces higher arterial carbon dioxide concentration and greater depression of the cardiovascular system. Patients at increased risk of airway obstruction, such as those with Down’s syndrome, and patients at risk of ventilatory failure, including neonates of low birth weight, should have their airway secured by endotracheal intubation.\(^\text{11}\) Endotracheal intubation is usually advisable if transesophageal echo is required. Continuous monitoring of the airway and ventilation is mandatory with any of these techniques.

Vascular access is most commonly achieved via groin vessels, although vessels in the neck, umbilical vessels and the arms may also be used. Observation of the patient is further limited by sterile drapes and by reduction in lighting levels. Reliance on electronic monitoring is normal and monitoring devices should be positioned out of the way of the radiological view.

Analgesia for cardiac catheterization can be provided by infiltration of local anesthetics at vascular access sites. It is simple and if performed correctly should not complicate vascular access. When performed prior to cannulation the need for deepening of sedation or anesthesia is minimized.

**Complications**

Care should be taken in positioning the patient since neuropathy due to traction on the brachial plexus has been reported.\(^\text{12}\) When biplane fluoroscopy is used it is common to position the patient’s arms above the head. Such patients may require intubation and positive-pressure ventilation. Children with congestive heart failure, pleural effusions, or airway anomalies may not tolerate the supine position while awake. Temperature should be monitored and devices such as forced air warmers may be used for smaller patients.

Major complications are rare (2%) except during interventional procedures.\(^\text{13,14}\) Serious complications include arrhythmias, vascular damage at access sites, bleeding, perforation of vessels or the heart, cardiac tamponade, vascular thrombosis, air embolus, catheter fragment embolus, valvular incompetence, allergy to contrast medium or drugs, and stroke. Risk factors for complications include lower patient age or size (less than 5 kg) and the particular intervention performed. The overall mortality rate is 0.14% (0.28% for interventional procedures) and is most often due to either perforation of the heart and great vessels or due to the patient’s underlying disease.

Arrhythmias are the most common complication, occurring in 2.6% of all procedures. Arrhythmias, including heart block caused by mechanical stimulation, are usually transient and respond to withdrawal of the catheter. Contributory factors such as electrolyte disturbance, hypercarbia and excessive catheter manipulation within the heart should be minimized. Equipment for defibrillation should be immediately available. Pacing can be instituted to treat heart block or supraventricular tachycardia (SVT). Other causes of arrhythmias must be considered including cardiac ischemia, coronary air embolus and direct damage to the myocardium or conducting system.

Catheters used for interventional procedures have a larger diameter than those used during diagnostic studies, and the risk of vascular damage at the site of insertion is increased, as is the risk of damage to heart structures. Complications such as perforation of the heart or vessels or valvular incompetence may require urgent surgical intervention. Blood should be immediately available and interventional procedures in children should only be conducted in hospitals where facilities for cardiac surgery exist.\(^\text{6,15,16}\) In the event of sudden blood loss, rapid transfusion and arterial monitoring is possible via the vascular access sheaths placed for the procedure. During balloon angioplasty it may be possible for the cardiologist to tamponade a rupture by reinflation of the balloon. However, emergency surgical repair will often be required.

Thrombosis and thromboembolism may occur at any site where the vascular endothelium is disrupted. Heparin is given in a dose of 50–150 U/kg prior to arterial cannulation, when the systemic circulation is entered, and during procedures that inevitably cause damage to the vascular endothelium. The use of anticoagulants will increase the likelihood of hematoma formation at vascular access sites. Protamine may be used to reverse heparinization but caution must be exercised due to the risk of adverse reactions.
Serious complications occur in 0.7–3.3% of EP studies in children. Complications are related to vascular access, catheter manipulation, or the use of radiofrequency energy. The Radiofrequency Ablation Registry provides data on complication rates for radiofrequency ablation. This includes a mortality rate of 0.2%, complete heart block in 1%, and valvular regurgitation in 0.5% (G. Van Hare, pers. comm., PAPCA registry data). Complications are higher in patients with CHD and heart block is more common when the abnormal pathway is close to the normal conducting system. Coronary artery injury has been demonstrated in animal models of radiofrequency ablation (G. Van Hare, pers. comm., PAPCA registry data).

**Diagnostic catheterization**

Advances in other imaging techniques, most notably echocardiography and magnetic resonance imaging, have provided less invasive methods for demonstrating cardiac and vascular anatomy and reduced the need for diagnostic cardiac catheterization. These modalities are less successful to date in assessing physiological parameters, although this may change. Careful consideration is given to the indications for diagnostic catheterization due to the invasive nature of the procedure and the inevitable radiation exposure. Current indications can be summarized as follows:

1. **To measure central and peripheral intravascular pressures and derive hemodynamic information such as pulmonary and systemic vascular resistance (SVR), shunt fractions, and cardiac output (CO).** The most common situation for this type of investigation is in preparation for the Fontan procedure.
2. **To define cardiac and vascular anatomy:** poor windows can defeat the most expert echocardiographer and certain anatomical features are difficult to visualize. This occurs in a minority of cases and usually implies either complex anatomy or complicating factors in the patient such as lung disease.
3. **To evaluate myocardial function and to assess the effects of drugs and respiratory interventions on the cardiovascular system; for example, during investigation of patients with pulmonary hypertension.** Endocardial biopsies, coronary artery angiography and assessment of myocardial function are part of the routine surveillance of patients following heart transplantation. Endocardial biopsy is also diagnostic in cases of cardiomyopathy and viral myocarditis. Diagnostic studies are also an integral part of transcatheter interventional procedures.

**Anesthetic considerations**

The anesthesiologist’s role during diagnostic catheterization is to provide care so that the patient emerges from the procedure with minimal psychological or physiological trauma, and the cardiologist derives meaningful data on which to base decisions about the child’s future treatment. All sedative and general anesthetic agents have hemodynamic effects and all depress respiration; this in turn influences the results of the investigation. It is important that the anesthesiologist has a clear understanding of the information being sought in the investigation, and that the cardiologist has some understanding of the effects of sedatives on the cardiovascular system.

More than 30% of children with CHD have reparative surgery in the neonatal period and more than 50% are operated on in the first year of life. The majority of children come to surgery without cardiac catheterization; the necessary information is acquired by echocardiography. A majority of patients presenting for diagnostic cardiac catheterization are either infants presenting with some complexity of their condition or as postoperative patients. Preoperative assessment should include a careful review of the child’s medical history, current symptoms, and any available diagnostic tests. Appropriate laboratory investigations will depend on the child’s physical condition and any medications prescribed. As a minimum, a hemoglobin level should be measured and blood group ascertained.

Particular concerns during cardiac catheterization of neonates and small infants include airway management, limited cardiovascular reserve, hypothermia, and changes in intravascular volume. Hypovolemia can arise from extensive blood sampling during the procedure or as a result of blood loss during catheter placement or exchange. It is difficult to monitor as bleeding is hidden on the drapes, therefore serial hematocrit measurements are helpful. The use of check-flow valves markedly reduces bleeding from the sheath. Hypervolemia is also a concern as fluid is routinely administered through the sheath and catheters. Extensive angiographic studies result in the administration of significant amounts of contrast and this should be limited. The use of low osmolarity non-ionic contrast has reduced the number of side effects, but the total volume of fluid can still be considerable.

Postoperative patients are often understandably anxious when returning to hospital. There are benefits from premedication even with parental presence at induction of anesthesia. Midazolam 0.25–0.75 mg/kg is a safe and effective oral premedication for children with CHD and has the advantage of a rapid onset. These children are unlikely to have normal hemodynamics and may have severely limited cardiovascular and respiratory reserve. The most common cardiac sequelae of surgery for CHD are arrhythmias and myocardial dysfunction. Recurrent laryngeal and phrenic nerve palsy are recognized complications of cardiac surgery and result in limited respiratory reserve, as does congestive heart failure. Vascular access can be extremely difficult in children who have had prolonged hospitalization. Venous
thrombosis (and the attendant complications) is being increasingly recognized as a source of morbidity in children, thus increasing the difficulty of cardiac catheterization.21

In addition, 25% of children with CHD have other congenital anomalies. Craniofacial, airway, and intrathoracic anomalies are a major cause for concern. The combination of a patient with a challenging airway, limited cardiac reserve, and difficult vascular access in an area of the hospital often some distance from colleagues, epitomizes the challenge for the pediatric anesthesiologist in the cardiac catheterization laboratory.

**Procedure**

The routine approach to diagnostic catheterization is via the femoral vein and/or artery using the Seldinger technique. It is common to elevate the pelvis to facilitate venepuncture. Following cavopulmonary connection, subclavian or internal jugular vein cannulation is required to study the pulmonary vascular bed. After dilation and placement of an appropriate sheath, the catheter is advanced through the circulation; pressure and oxygen saturation measurements are then made in sequence. Oxygen saturation measurements allow the calculation of shunts, and in combination with measurements or estimates of oxygen consumption, allow calculation of CO and pulmonary blood flow (see Tables 25.1 and 25.2 for normal data and calculations). The use of a high inspired concentration of oxygen at this stage will introduce errors due to dissolved oxygen and pulmonary vasodilation. Whenever possible 21% oxygen should be administered and normocapnia achieved. Cardiac output can be measured using thermodilution techniques in patients without shunts.

**Anesthetic techniques**

It has been common to provide sedation for cardiac catheterization with large single doses of oral or intramuscular sedation. Regimens include a mixture of meperidine, promethazine, and chlorpromazine (DTP) and large doses of oral chloral hydrate. The limitations of such techniques are considerable, as the dose cannot be titrated to response and the duration of action may be greatly prolonged in infants.22 Periods of either excess sedation or inadequate sedation are common. These techniques have been rightly superseded by use of shorter acting agents with rapid onset.23 Table 25.3 summarizes agents in common use.

Many of these agents have steep dose–response curves and need to be carefully titrated to achieve a predetermined endpoint. The results of excess sedation are predictable: loss of airway reflexes, respiratory depression, and cardiovascular compromise. Specific antagonists exist for opioids and benzodiazepines; however, initial management should be support of the airway and ventilation. In an analysis of critical incidents due to sedation, no correlation could be found between particular agents or mode of administration and outcome.22,24 In specific situations combinations of two agents may be useful: opioids and sedatives may be of value for painful procedures and benzodiazepines reduce the incidence of hallucinations when ketamine is used.

The effects of sedative and analgesic drugs on the heart are variable. Inhalational anesthetics cause peripheral vasodilation, varying degrees of myocardial depression, and affect sinus node function and cardiac conduction tissue. Sevoflurane has a less direct myocardial depressant effect than halothane: in normal children sevoflurane produces a fall in

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**Table 25.1 Normal cardiac catheterization data.**

<table>
<thead>
<tr>
<th></th>
<th>Pressure in mmHg</th>
<th>Oxygen saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Newborns</td>
<td>Older children</td>
</tr>
<tr>
<td>Right atrium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a wave</td>
<td>3–8</td>
<td>5–10</td>
</tr>
<tr>
<td>v wave</td>
<td>2–6</td>
<td>4–8</td>
</tr>
<tr>
<td>Mean</td>
<td>0–4</td>
<td>2–6</td>
</tr>
<tr>
<td>Right ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>65–80</td>
<td>15–25</td>
</tr>
<tr>
<td>End-diastolic</td>
<td>2–7</td>
<td>3–8</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>65–80</td>
<td>15–25</td>
</tr>
<tr>
<td>Diastolic</td>
<td>35–50</td>
<td>8–12</td>
</tr>
<tr>
<td>Mean</td>
<td>40–70</td>
<td>10–16</td>
</tr>
<tr>
<td>PA wedge</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a wave</td>
<td>6–10</td>
<td>8–14</td>
</tr>
<tr>
<td>v wave</td>
<td>7–11</td>
<td>10–17</td>
</tr>
<tr>
<td>Mean</td>
<td>5–8</td>
<td>7–13</td>
</tr>
<tr>
<td>Left atrium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a wave</td>
<td>4–7</td>
<td>6–12</td>
</tr>
<tr>
<td>v wave</td>
<td>6–12</td>
<td>8–15</td>
</tr>
<tr>
<td>Mean</td>
<td>3–6</td>
<td>5–10</td>
</tr>
<tr>
<td>Left ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>65–80</td>
<td>90–120</td>
</tr>
<tr>
<td>End-diastolic</td>
<td>3–7</td>
<td>2–5</td>
</tr>
<tr>
<td>Aorta</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>65–80</td>
<td>90–120</td>
</tr>
<tr>
<td>Diastolic</td>
<td>45–60</td>
<td>60–75</td>
</tr>
<tr>
<td>Mean</td>
<td>55–65</td>
<td>70–90</td>
</tr>
<tr>
<td>Flows</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary (Qp)</td>
<td>3.5–5.0</td>
<td>3.5–5.0</td>
</tr>
<tr>
<td>Systemic (Qs)</td>
<td>3.5–5.0</td>
<td>3.5–5.0</td>
</tr>
<tr>
<td>Resistances</td>
<td>Woods units x m² BSA</td>
<td></td>
</tr>
<tr>
<td>Pulmonary (Rp)</td>
<td>8–10</td>
<td>1–3</td>
</tr>
<tr>
<td>Systemic (Rs)</td>
<td>10–15</td>
<td>15–30</td>
</tr>
</tbody>
</table>

BSA, body surface area; PA, pulmonary artery.
### Table 25.3 Suitable agents for sedation during cardiac catheterization.

<table>
<thead>
<tr>
<th>Agent</th>
<th>Suggested dose</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midazolam</td>
<td>Oral 0.25–1.0 mg/kg</td>
<td>After i.v. bolus 4 min before peak effect</td>
</tr>
<tr>
<td></td>
<td>i.v. bolus 50–150 µg/kg</td>
<td>Steeper dose–response curve than diazepam</td>
</tr>
<tr>
<td></td>
<td>Infusion: 1–2 µg/kg/min</td>
<td></td>
</tr>
<tr>
<td>Ketamine</td>
<td>1–2 mg/kg</td>
<td>Psychic disturbances in 5–30%</td>
</tr>
<tr>
<td></td>
<td>Infusion 50–75 µg/kg/min</td>
<td>Prolonged recovery time</td>
</tr>
<tr>
<td>Propofol</td>
<td>1–2 mg/kg</td>
<td>High risk of loss of airway reflexes</td>
</tr>
<tr>
<td></td>
<td>Infusion: 100–200 µg/kg/min</td>
<td></td>
</tr>
<tr>
<td>Nitrous oxide</td>
<td>Up to 50%</td>
<td>Seldom adequate as sole agent, useful adjunct during skin infiltration</td>
</tr>
<tr>
<td>Morphine</td>
<td>0.05 mg/kg</td>
<td>Risk of respiratory depression and nausea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prolonged action</td>
</tr>
<tr>
<td>Fentanyl</td>
<td>1 µg/kg</td>
<td></td>
</tr>
<tr>
<td>Alfentanil</td>
<td>20 µg/kg then 0.5 µg/kg/min</td>
<td>Much smaller doses required after Fontan procedure</td>
</tr>
<tr>
<td>Remifentanil</td>
<td>0.05–0.15 µg/kg/min</td>
<td>High potential for apnea</td>
</tr>
<tr>
<td>Etomidate</td>
<td>0.1–0.3 mg/kg i.v. bolus then 25–50 µg/kg/min infusion</td>
<td>Transient adrenal suppression, pain and thrombophlebitis; excellent maintenance of baseline hemodynamics</td>
</tr>
</tbody>
</table>

### Flows

#### Pulmonary:

\[ Q_p = \frac{V_{O_2}}{(S_{V_2}O_2 - S_{A_2}O_2) \times Hgb \times 1.34 \times 10} \]

#### Systemic:

\[ Q_s = \frac{V_{O_2}}{(S_{A_2}O_2 - S_{A_2}O_2) \times Hgb \times 1.34 \times 10} \]

#### Effective pulmonary:

\[ Q_{EP} = \frac{V_{O_2}}{(S_{V_2}O_2 - S_{A_2}O_2) \times Hgb \times 1.34 \times 10} \]

### Resistances

#### Pulmonary:

\[ R_p = \frac{P_{AP} - R_{AP}}{Q_p} \]

#### Systemic:

\[ R_s = \frac{A_{OP} - R_{AP}}{Q_s} \]

### Shunts

#### Pulmonary to systemic:

\[ \frac{Q_p}{Q_s} = \frac{S_{V_2}O_2 - S_{A_2}O_2}{S_{A_2}O_2 - S_{A_2}O_2} \] (a flow ratio)

#### Left to right:

\[ Q_p - Q_{EP} \] (absolute flow)

#### Right to left:

\[ Q_p - Q_{EP} \] (absolute flow)

Ao, aorta; AoP, aortic pressure; Hgb, hemoglobin; LAP, left atrial pressure; MV, mixed venous; PA, pulmonary artery; PAP, pulmonary artery pressure; PV, pulmonary vein; Q_{EP}, effective pulmonary flow; Q_p, pulmonary flow; Q_s, systemic flow; R_{AP}, pulmonary vascular resistance; R_s, systemic vascular resistance; S, saturation.
SVR without an increase in heart rate (HR) resulting in no change in cardiac index. Isoflurane preserves contractility in children with CHD, yet produces greater myocardial depression in infants.

Intravenous agents such as propofol, midazolam and ketamine are all used for sedation and general anesthesia. Propofol has been studied in children with intracardiac shunting undergoing cardiac catheterization; it decreases SVR resulting in significant decreases in the ratio of pulmonary to systemic flow (Qp:Qs). In patients with elevated pulmonary vascular resistance (PVR), propofol caused pulmonary vasodilation and varying degrees of bradycardia. When a combination of oral midazolam and ketamine was used for cardiac catheterization, patients often required intravenous supplementation and developed airway obstruction.

Cardiac catheterization, patients often required intravenous supplementation and developed airway obstruction. Ketamine causes sympathetic stimulation, salivation and bad dreams. It has negative inotropic effects on isolated myocardium, but this is masked in the intact patient due to the sympathomimetic effects. Ketamine increases oxygen consumption (VO2), leading to a potential source of error in hemodynamic calculations unless VO2 is actually measured. There are conflicting reports on the effects of ketamine on PVR. Etonidate maintains CV stability and has been used for induction of anesthesia in children with CHD and end-stage cardiomyopathy. There are few reports of its use for cardiac catheterization, but baseline hemodynamics are preserved during procedures lasting up to several hours. The use of short acting intravenous narcotics such as alfentanil and remifentanil for sedation of spontaneously breathing children has also been reported. These drugs generally produce good hemodynamic stability. The problems of respiratory depression (apnea and airway obstruction) and vomiting limit the usefulness of these agents unless the airway and ventilation are controlled. In addition the use of remifentanil with sevoflurane and positive pressure ventilation is associated with a decrease in HR and arterial blood pressure.

Respiratory manipulations also affect hemodynamics. A switch from positive to negative pressure (cuirass) ventilation increased CO by 11% in healthy children, 28% in postoperative cardiac patients, and 54% in patients with a Fontan circulation. A similar increase in CO has been demonstrated on extubation of postoperative Fontan patients. If the child has a large left-to-right shunt then the effect of breathing high concentrations of oxygen are twofold: first, it is necessary to consider the contribution of dissolved oxygen when calculating pulmonary blood flow using the Fick equation; secondly, high oxygen levels decrease PVR thereby increasing Qp:Qs. Similarly, changes in arterial carbon dioxide tension or pH will affect pulmonary blood flow. From the perspective of deriving the best hemodynamic data, the ideal situation is a patient spontaneously breathing room air; however, airway obstruction, carbon dioxide retention, or atelectasis also produced potential sources of error in the study.

**The patient with pulmonary hypertension**

Primary pulmonary hypertension is rare in children, as in adults. However, children with CHD and large left-to-right shunts are at risk of acquired pulmonary hypertension if their cardiac anomaly is not dealt with in a timely fashion. Infants with outflow obstruction to the pulmonary vascular bed (e.g. mitral stenosis or pulmonary vein obstruction) are also at risk.

When surgery is planned for children with pulmonary hypertension it is important to carefully assess pulmonary vascular reactivity, in order to decide if the lesion is operable. These investigations require teamwork on the part of the anesthesiologist and cardiologist. Children with severe pulmonary hypertension are at risk of sudden death, and require careful anesthetic management, and children whose pulmonary vascular bed is labile can appear inoperable if improperly managed.

Pulmonary hypertension causes right ventricular failure. The thin-walled right ventricle (RV) responds to pressure loading by dilating and becoming hypertrophied. This reduces right coronary blood flow rendering the subendocardial region vulnerable to ischemia. The dilated RV interferes with left ventricular geometry and function causing increased left ventricle (LV) end-diastolic pressure and decreased stroke volume. Tricuspid regurgitation can also occur. Acute increases in PA pressure and PVR are poorly tolerated, reducing CO, which may lead to arrhythmias and death.

The intimate relationship between PVR, alveolar oxygenation, and carbon dioxide means that the first principle of anesthetic management of the child with pulmonary hypertension is meticulous attention to the airway and to the gas exchange. Hypercarbia or hypoxia will cause an elevation in PVR, which in turn has been shown to cause bronchospasm resulting in greater difficulties with oxygenation and ventilation, inducing a rapid downward spiral. Ineffective bag and mask ventilation, or too large a leak around the endotracheal tube can result in life-threatening acute increases in PA pressure in vulnerable children and is particularly important if there is no intracardiac shunt (e.g. patent foramen ovale or atrial septal defect), because the RV fails and CO is significantly reduced.

The rationale behind the management of children undergoing investigation of pulmonary hypertension is to establish the baseline hemodynamic values and then to intervene to assess the reactivity of the pulmonary vascular bed. This also provides data against which the effects of therapy can be measured, and decisions are based on the lowest value of PVR that can be attained.

For such studies at HSC, all children receive general anesthesia with muscle relaxants and endotracheal intubation. Premedication with oral midazolam is routine. Cautious inhalation induction with sevoflurane has been used if
vascular access is difficult, and avoids the inevitable agitation due to multiple attempts at obtaining intravenous access. A bolus (0.5–1.0 µg/kg) of remifentanil followed by an infusion at doses of 0.25 µg/kg/minute is started and titrated to effect. Atropine can be given if the HR slows significantly. Isoflurane 0.5% or small doses of midazolam are given intravenously to induce amnesia. The patient is ventilated with air and oxygen and the pH and venously to induce amnesia. The patient is ventilated with air and oxygen and the pH and PCO₂ are maintained within normal limits while intracardiac pressures and saturation are measured, and calculations of PVR and the PVR index are made. The patient is then hyperventilated until the PCO₂ is 30 mmHg on 100% oxygen after which all measurements are repeated. Patients who do not respond to hyperoxia and hypocarbia are given 40 p.p.m. nitric oxide and the study repeated. In selected patients the study is used to assess the effects of therapy such as inhaled or intravenous prostacycline, or oral sildenafil.

Endocardial biopsy

Endocardial biopsy is used in three patient groups: the routine surveillance of patients post-cardiac transplant, the assessment of patients with suspected rejection of the transplanted heart, and the diagnosis of myocarditis. The latter two groups are more likely to have myocardial dysfunction. When seen preoperatively all patients should be questioned as to symptoms of heart failure or arrhythmia, and reports of preoperative echocardiograms and electrocardiograms (ECGs) inspected. The procedure is usually performed by cannulation of the right internal jugular or femoral vein. The biopsy catheter is passed into the heart and samples of endocardium taken for histology. Stress testing, coronary angiogram, and ultrasound examination of the coronary arteries may be performed during the same procedure.

Interventional cardiology techniques

Valvotomy

The technique used for balloon dilation of any stenotic valve (aortic, pulmonary, mitral, or bioprosthetic) is essentially similar. A catheter is introduced past the stenosis and pressures are measured proximal and distal to the lesion. A wire is positioned across the valve and the catheter withdrawn. The wire is used to guide a balloon catheter into position across the stenotic valve and to stabilize the balloon. The size of balloon used is dictated by the size of the valve annulus, because the purpose of the procedure is not to dilate the valve annulus but to separate the fused valve leaflets. Larger balloons reduce the gradient, but increase the risk of regurgitation. During inflation the valve is completely obstructed; for this reason, sustained single inflations are avoided. When a balloon is inflated across a stenotic valve, a “waist” is seen at the site of stenosis, and short inflations are repeated until this waist disappears.42

Angioplasty

Vascular angioplasty involves the inflation of a balloon across an area of vascular stenosis using a similar technique to balloon valvotomy. It results in tearing of the endothelium and subsequent healing by scar formation and re-endothelialization. The technique may be applied to native blood vessels or to surgical conduits. Established indications are recurrent aortic coarctation, systemic venous stenosis and PA stenosis. Complications include rupture of the vessel, dissection, late aneurysm formation, thrombosis, and restenosis.

Endovascular stents

Modification of balloon angioplasty by the placement of endovascular stents has been applied to PA stenosis, systemic
venous stenosis, and to coarctation of the aorta (Fig. 25.2). The placement of endovascular stents improves the initial success of angioplasty, reduces the incidence of recurrence and may reduce late aneurysm formation. The stents used are deployed over a balloon that is positioned and inflated to dilate the stent and the vessel, then withdrawn, leaving the stent in situ.

The technique is limited by the large size of catheters required to deploy stents and a failure of the stents to grow with the patient; however, newer designs of stents that allow redilation reduce this limitation. Large catheters increase the risk of damage to vascular and cardiac structures, especially thrombosis of the femoral vessels. Further complications are malposition or embolization of the stent.

### Closure of shunts

A number of devices are available to close intravascular...
In older infants, in whom the DA is closed, there is no alternative route for pulmonary blood flow; therefore, inflation of the balloon leads to greater hemodynamic upset. Cardiac output decreases dramatically, resulting in significant hypotension. This usually improves on deflation of the balloon, but if ventricular dilation develops the blood pressure may not recover promptly. Bolus doses of epinephrine and occasionally more prolonged inotropic support may be required.

A modification of the technique is used for treatment of pulmonary atresia with intact ventricular septum in which radiofrequency energy is used to perforate the membrane at the pulmonary annulus. Because the pulmonary valve leaflets are absent and the membrane destroyed, pulmonary insufficiency is inevitable, and the risk of inadvertent perforation of the right ventricular outflow tract is higher during this procedure.

Pulmonary artery stenosis

Pulmonary artery stenosis may be congenital or acquired, often following surgery. Congenital stenosis is associated with three conditions: reduced pulmonary blood flow (such as tetralogy of Fallot), branch stenosis in association with Williams’ syndrome or congenital rubella, or as an isolated stenosis at the site of insertion of the DA. Stenosis may occur after any surgery involving manipulation of PAs, particularly creation of shunts or PA banding. Stenosis can arise at any point in the pulmonary vascular bed and multiple sites are not uncommon. Two thirds are confined to proximal vessels. The right ventricular pressure is raised when the main PA is affected or when multiple peripheral stenoses are present. If only one branch PA or the peripheral PAs are affected, then the PA pressure may be normal at rest.

Surgery is the treatment of choice for proximal PA stenosis but surgical treatment of peripheral PA stenosis is difficult. Transcatheter angioplasty is initially successful in 50% but restenosis is common and long-term benefits are seen in less than 35% of patients. In a large series published in 1992, the mortality was 1–2% and the risk of serious complications
5%. Complications include vessel rupture, hemoptysis, paradoxical embolism, balloon rupture, aneurysm formation, air embolism, and unilateral pulmonary edema.

The outcome of PA stenosis is improved with the use of endovascular stents, which reduce the incidence of initial failure, and restenosis. The large catheter positioned in a stenotic PA increases the degree of obstruction resulting in worsening hypoxia and right ventricular failure. The use of extracorporeal membrane oxygenation has been described in high-risk patients.51

Patients with cavopulmonary shunts have a limited capacity to tolerate obstruction to pulmonary blood flow. Obstruction can occur at the site of the superior vena cava to PA anastomosis, at the site of previous surgery, or at a remote site. Angioplasty, commonly with placement of stents, may be used to relieve the obstruction; however, when required in the early postoperative period, the patient’s condition is often poor.

 Patients with Williams’ syndrome may have multiple peripheral stenoses of the PAs, associated supravalvular aortic stenosis and coronary artery stenosis. In a series of 39 procedures the mortality rate was 7.7%,52 and high right ventricular pressure was a predictor of mortality. Three out of 22 patients initially managed with deep sedation required intubation.

Aortic valvotomy

Balloon dilation of isolated aortic valve stenosis is indicated when the transvalvular gradient is greater than 70 mmHg, or is greater than 50 mmHg with symptoms, or when ECG evidence of ischemia is seen. Neonates have much smaller gradients despite severe stenosis, as flow across the aortic valve may be minimal. Untreated severe stenosis carries a 19% risk of sudden death.52,55

In a series of 630 dilations, the immediate outcome was less than optimal (failure or major morbidity) for 17% of patients and procedural mortality was 1.9%.53 Major complications, including aortic regurgitation, vascular damage, and perforation of the heart or great vessels, occurred in 6.3% of patients. Age less than 3 months, high gradient, the use of small balloon sizes, and the presence of an aortic coarctation predicted poor outcome,54,55 and outcome improved with greater experience of the technique. The procedure is essentially palliative, with 50% of patients requiring further interventions within 8 years.

Damage to the aortic valve is a major concern and severe aortic insufficiency occurs in 1.6% of cases. The valve may be damaged by use of an oversized balloon causing dilation of the valve annulus or by inadvertent puncture of the valve leaflet. The latter complication may be reduced by use of an antegrade approach to the valve via the atrial septum (either puncturing the septum or through the foramen ovale). This approach also eliminates the risk of damage to the femoral artery, the passage of the wire across the stenotic valve is simplified and less hemodynamic compromise occurs. If severe aortic insufficiency occurs then coronary blood supply is compromised and urgent surgical repair may be required, which is associated with high mortality, especially in the neonatal age group.56

The most difficult patients are neonates with critical aortic stenosis. The incidence of major complications in this age group is 16.7% and procedural mortality is 8.3%. There is a risk of sudden death from myocardial ischemia and arrhythmia. The systemic and coronary circulations are dependent on right-to-left flow through the DA, and the LV is greatly hypertrophied with poor compliance. Initial resuscitation includes mechanical ventilation, PGE1, and cautious use of inotropes. Inflation of the balloon across the aortic valve causes complete obstruction of coronary flow. A narcotic based anesthetic technique reduces cardiac work, and minimizes afterload reduction and tachycardia. Care must be taken to maintain preload as any reduction in CO leads to further ischemia. Inotropes, vasopressors, and a defibrillator should be immediately on hand.

Older infants may present with severe degrees of aortic stenosis. They have heart failure and are at risk of ischemia and arrhythmias. Cardiovascular compromise during balloon inflation is inevitable but on deflation of the balloon prompt return of CO is to be expected. The incidence of arrhythmias is higher than during other interventional procedures. As with neonates there is a risk of arterial damage and of aortic regurgitation, which may require surgical intervention. It has been suggested that these patients may be managed safely with sedation.41 However, the patient needs to be still during positioning of the balloon and there is risk of hemodynamic compromise or complications, and at the HSC it is normal to provide general endotracheal anesthesia for this group. Older patients may present with progressive aortic stenosis or with restenosis.

Coartation of the aorta

Balloon angioplasty is used for both native and recurrent coarctation of the aorta.57 The indications are resting hypertension proximal to the coarctation with a gradient of greater than 20 mmHg or the presence of multiple collaterals (Fig. 25.3). The best results are obtained when there is a short discrete coarctation with an otherwise normal aortic arch. Initial success with native coarctation is greater than for recurrent coarctation; however, there is a high incidence of late aneurysm formation (2–6%) and of restenosis (7–12%). In neonates recurrence rates are high and angioplasty is not indicated. In older children with native coarctation the role of angioplasty is controversial.18 Surgery for recurrent coarctation is more difficult and recoarctation is an accepted indication for transcatheter angioplasty. Use of endovascular stents may reduce both restenosis and aneurysm formation, but the risk of damage to the femoral artery and the failure of stents to grow with the patient limit the use of this technique.
The procedure is generally well tolerated but the risk of serious sequelae is significant. Older patients often have significant collaterals and this reduces the incidence and severity of hemodynamic compromise during balloon inflation. Complications occur in 15% of procedures and include aortic rupture or dissection, stroke, femoral artery damage, thrombosis, and late aneurysm formation with a 0.7% mortality. As with surgical repair, postoperative hypertension should be anticipated.

Closure of atrial septal defect and ventricular septal defect

Selected atrial septal defects can be closed with a device. The technique is unsuitable for ostium primum defects or for large defects. A rim of septal tissue is required to allow the device to be anchored. At 1 year, 5–10% of patients have significant residual leaks. Complications are uncommon and include embolization of the device, encroachment on atrioventricular (AV) valves, obstruction of pulmonary or systemic veins, perforation of the heart or great vessels and air embolus. Generally the procedure is well tolerated. The procedure is usually performed with general endotracheal anesthesia due to the use of transesophageal echocardiography and the need for a still patient.

Transcatheter closure of a VSD is a more complex procedure with a greater risk of serious complications. A wire is placed across the defect from the left side. This wire is then snared from the right heart and brought out of the femoral vein so that a continuous connection is made from the venous system and right heart, through the VSD into the LV and out through the aortic valve and arterial system. The device is then deployed via the right heart, avoiding passage of a large catheter through the femoral artery and aortic valve. Complications include blood loss, arrhythmias, atrioventricular or aortic valve regurgitation, and cardiac arrest. In one series, 50% of patients were admitted to intensive care following the procedure. General anesthesia and control of the airway is required due to the high risk of cardiovascular instability, the length of the procedure, and the use of transesophageal echocardiography. The precise role of this procedure has yet to be defined, though with greater experience and improved technology the incidence of complications is likely to be reduced.

Closure of extracardiac connections

Connections between the systemic and pulmonary circulations may occur in isolation (patent ductus arteriosus [PDA]), in association with CHD (aortopulmonary connections with pulmonary atresia), as a complication of cyanotic heart disease (venovenous connection after cavopulmonary connection) or through surgically created shunts. Usually these lesions are closed by embolization with helical wires. The choice of anesthetic technique will depend on the patient’s physiology. Cyanotic patients with a cavopulmonary connection and multiple collaterals may not tolerate positive pressure ventilation. Older patients presenting with large PDAs may have significant pulmonary hypertension and be at risk of right heart failure. Embolization of the device into the pulmonary or systemic circulation may cause vascular occlusion and infarction. Often the device can be retrieved via the catheter but open retrieval may be required.

Atrial septostomy

Balloon atrial septostomy was first described in 1966. The objective is to open a non-restrictive connection between the left and right atria to allow bidirectional mixing of blood. It has most widely been used for the initial palliation of
Other indications are total anomalous pulmonary venous drainage, atrioventricular valve atresia, and pulmonary atresia with intact ventricular septum.

The technique involves the passage of a balloon tipped catheter across the foramen ovale into the left atrium. The balloon is inflated and withdrawn across the septum, tearing the atrial septum. This procedure is repeated until the inflated balloon can be withdrawn without resistance. The procedure can be performed at the patient’s bedside using echocardiographic guidance. A modification of the procedure is applied to older infants, because by 1 month of age the atrial septum is too thick to be torn by the balloon alone. A catheter with a retractable blade is used to initiate the tear.

Complications include arrhythmias, perforation of the heart, balloon rupture, and embolization and damage to heart structures. Patients are often extremely hypoxic, acidic and may have pulmonary edema. Ventilation and PGE1 infusion are the first priorities and resuscitation should continue through the procedure. The effect of successful septostomy is dramatic with a rapid increase in oxygenation and in end-tidal carbon dioxide from improved pulmonary blood flow.

**Electrophysiology studies and radiofrequency ablation**

The purpose of an EP study is to identify the mechanism of the patient’s arrhythmia by recording signals from electrodes placed within the heart. Use of fluoroscopy and reference to anatomic landmarks allows localization of the abnormal pathways and foci responsible for the arrhythmia. Ablation of the abnormal pathway is initially successful in 91% of patients with a recurrence rate of 23%.17,63

The majority of children presenting for EP investigation and treatment are otherwise healthy, have functionally normal hearts, and present with well-tolerated SVT. A minority have either a life-threatening arrhythmia, or an arrhythmia complicating CHD. This is in contrast to the adult population, who more frequently present with ventricular arrhythmias complicating ischemic heart disease. Anesthetic concerns include the length of the procedure, poor access to the patient, and the possibility of anesthetic agents altering the EP of the heart. The effect of anesthetic agents on the ability to study the patient’s arrhythmia depends upon the mechanism of the arrhythmia.

**Pathogenesis of arrhythmia**

The most common mechanism of tachycardia is re-entry. This requires a circuit composed of connected pathways with different conduction velocities and refractory periods (Fig. 25.4). Pre-excitation syndromes occur in a subgroup of patients with re-entry tachycardia. One arm of the circuit is the patient’s atrioventricular node and the other is a congenital muscular pathway between the atrium and ventricle. Tachycardia arises when depolarization occurs in the circuit while one limb is refractory and the other is able to conduct. The depolarization continues around the circuit and reaches the previously refractory pathway, which is now able to conduct, and the circle is therefore perpetuated (Fig. 25.4).64

A second mechanism is altered automaticity. During normal sinus rhythm only the sinoatrial node independently generates rhythmic impulses through spontaneous depolarization of the basement membrane. Other cells demonstrate this activity but at a slower rate, and only control the HR if the sinus node is not functioning or conduction is blocked. When cells are damaged and subjected to extrinsic factors (electrolyte disturbance, hypoxia, hypercarbia, high wall tension, ischemia, and high catecholamine levels) spontaneous depolarization may be accelerated. This leads to rapid repetitive depolarization of a single focus: ectopic tachycardia.

**Electrophysiology techniques**

Figure 25.5 demonstrates the typical electrode catheter position during an EP study for investigation of SVT. The catheters have multiple electrodes along their length. An electrode placed across the tricuspid valve can be positioned to record an ECG from the His bundle. Further electrodes are typically placed high within the right atrium, at the apex of the RV, and “roaming” electrodes can be placed elsewhere in the right heart. Potentials from the left heart can be recorded.
via an electrode within the coronary sinus or from electrodes placed directly within the left heart (via puncture of the atrial septum or retrograde passage through the aortic valve).

Figures 25.6 and 25.7 demonstrate typical ECGs recorded during an EP study. In order to identify the mechanism of the arrhythmia periods of pacing and programmed stimulation are undertaken. Pacing, commonly via the high right atrium and ventricular apex, allows arrhythmias to be provoked or terminated, permits measurement of EP properties of the conduction system and allows for ventricular pacing should complete heart block occur (a rare complication). Drugs may be used to further refine the study. Adenosine blocks normal AV conduction exposing a concealed abnormal pathway. Isoproterenol increases sinoatrial rate, speeds AV node conduction, reduces refractory periods, and increases automaticity of other contractile tissue.

**Radiofrequency ablation**

Destruction of abnormal pathways and automatic foci abolishes the arrhythmia. This is most often accomplished by delivering radiofrequency energy to ablate the area. In the treatment of pre-excitation syndromes a specialized catheter is positioned along the AV ring then adjusted to record the earliest conduction via the abnormal pathway. When delivering radiofrequency energy, the size of lesion created is controlled by measurement of the temperature generated at

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**Fig. 25.5** Typical catheter position during electrophysiology study for investigation of supraventricular tachycardia. Catheters have multiple electrodes along their length. Catheters are positioned across the tricuspid valve to record the signal from the His bundle, in the ventricular apex, in the high right atrium, and within the coronary sinus. In this image contrast has been injected into the coronary sinus.

**Fig. 25.6** Simultaneous surface and intracardiac electrocardiograms (ECGs) are shown. Three paced beats (P) are delivered via the high right atrial catheter followed by a premature stimulus (E) resulting in supraventricular tachycardia (SVT). For each beat the CS 9,10 lead demonstrates three signals: an artifact corresponding to the pacing stimuli; a signal from depolarization of adjacent atrial muscle (A); followed by a signal from the adjacent ventricle (V). The His-bundle depolarization (H) coincides with depolarization of the ventricular septum adjacent to the electrode. During paced rhythm ventricular depolarization (V) occurs earliest in the CS 3,4 electrode (an electrode in the center of the coronary sinus catheter), indicating proximity to the accessory pathway. The pre-excitation (delta wave) seen on the surface ECG (S1) is confirmed by depolarization of CS 3,4 prior to His-bundle depolarization (H). Following initiation of SVT the morphology of the surface QRS becomes normalized indicating activation via the normal conducting system (S2). Earliest retrograde activation of atrial muscle is seen on the CS 3,4 electrode confirming the position of the accessory pathway.
CHAPTER 25  Anesthesia for the cardiac catheterization laboratory

Anesthetic considerations

As with other catheter procedures it is possible to conduct EP studies in conscious patients or with minimal sedation. At the HSC in Toronto it is common to use general anesthesia for procedures in children and adolescents. Though only vascular access is painful, the procedures are often prolonged, multiple venous and arterial access points are required, and periods of arrhythmia are inevitable. Arrhythmias are unpleasant for the patient and urgent interventions are facilitated by anesthesia. Deep sedation offers no advantage to anesthesia and the additive effect of repeated doses of sedative drugs over a period of time must be considered. When vascular access sheaths are placed within the thorax in a spontaneously breathing patient greater care is required to reduce the risk of air embolus.

Preoperative considerations include the presence of CHD, cardiomyopathy, or familial conditions associated with arrhythmia (long QT syndrome and arrhythmogenic right ventricular dysplasia). The patient should be questioned to the frequency of arrhythmia, factors that precipitate the arrhythmia, their symptoms during the arrhythmia, and whether treatment is required to terminate the arrhythmia. Arrhythmias associated with fainting or collapse of the patient are likely to be associated with greater hemodynamic compromise. If anxiety precipitates the arrhythmia, preoperative anxiolysis is indicated. Antiarrhythmics are usually stopped prior to the procedure unless the arrhythmia is frequent and poorly tolerated. The majority of patients are adolescents and discreet questioning as to risk of pregnancy (especially in view of X-ray exposure) and substance abuse is appropriate.

The likely mechanism of arrhythmia is often known from surface ECG (e.g. the presence of delta waves indicates

![Fig. 25.7 Surface and intracardiac electrocardiograms (ECGs) are shown from the same patient as in Fig. 25.6. Ventricular depolarization (V) is earliest in CS 3,4 and pre-excitation is demonstrated on the surface ECG (S2). As radiofrequency energy is delivered pre-excitation is lost with normalization of the surface QRS morphology (S2) and ventricular depolarization on CS 3,4 occurs later in the cardiac cycle.](image-url)
Wolff–Parkinson–White syndrome) or from a holter recording of the arrhythmia. This allows the anesthetic technique to be adapted to avoid suppression of the arrhythmia and to have minimal effects on the patient’s EP. Specific agents may suppress abnormal pathways or foci to a point where their detection and the induction of the arrhythmia is not possible. This leads to false negative studies and failure of the procedure.

Other considerations are the length of the procedures, poor access to the patient, and the potential for hemodynamic compromise. X-ray tables are very firm and patient positioning can be awkward. Care needs to be taken to avoid injury to nerves and pressure areas and large adolescent patients are particularly difficult to position.

Access to the patient is reduced from the use of the subclavian or internal jugular veins for vascular access for some procedures. The need to have X-ray and EP equipment close to the patient further reduces access to the patient, and intravenous lines, ventilator tubing, and anesthetic machines should be positioned accordingly.

Periods of very rapid HR and rhythm are to be expected during an EP study and healthy patients tolerate this well. Studies in patients with atrial arrhythmias often fall into this category, and beat-to-beat monitoring of arterial pressure may not be necessary. Greater hemodynamic compromise occurs when the ventricular rate is very high, when the focus is ventricular, or when the patient has reduced cardiac reserve. Continuous arterial pressure monitoring via a femoral artery catheter should be considered in these patients. Vasopressors may be required to improve perfusion pressure though α-adrenergic agonists have reflex effects on the EP of the heart. Discussion with the cardiologist prior to and during the procedure is vital in the management of more difficult patients. Most arrhythmias can be terminated rapidly by overdrive pacing. Drugs (other than adenosine) or external cardioversion are rarely required and may force the cancellation of the procedure.

Anesthetic drugs and the cardiac conduction system

Anesthetic agents influence the EP of the heart. Effects are mediated via the sympathetic and parasympathetic systems or via the cardiac conduction system and myocardium. The significance of these effects depends upon the mechanism of the tachycardia.

Anesthetic drugs and pre-excitation syndromes

Supraventricular tachycardia due to pre-excitation syndromes requires conduction of impulses in a circuit involving the functional atroventricular node and accessory pathway. Effects upon the accessory pathway are most critical and can be characterized by two EP variables: the accessory pathway effective refractory period (APERP) and the coupling interval. The APERP is the minimal time between two impulses that are still conducted by the accessory pathway. The coupling interval is the maximal time between two impulses able to precipitate a SVT. A false negative EPS due to suppression of conduction via the accessory pathway is most critical following radiofrequency ablation, because it may lead to recurrence of SVT.

Enflurane, isoflurane and halothane at 1 minimum alveolar concentration (MAC) have been shown to cause prolongation of the APERP in both adults and children. It has been suggested that these agents be avoided during ablation of accessory pathways. Enflurane has a greater effect than other agents. However a further study of isoflurane and a study of sevoflurane failed to demonstrate significant electrophysiological effects at 1 MAC. Conversely, isoflurane and halothane prolong the coupling interval, potentially increasing susceptibility to SVT. From animal studies it is clear that inhalational agents do have a number of electrophysiological effects when administered in sufficient dose; however, the clinical significance of this may be limited.

A number of clinical studies have demonstrated no direct effect of propofol on conduction at doses of 100–150 µg/kg/minute other than slight prolongation of atrial refractory period. In studies on isolated hearts, significant electrophysiological effects are apparent only at concentrations unlikely to be achieved clinically. Midazolam and alfentanil in combination do not have direct effects on cardiac conduction. Droperidol produces a marked prolongation of APERP and should be avoided. Sufentanil in very high doses produces a slowing of cardiac conduction, though it is not clear if this is true of other opiates. Vecuronium has no EP effects but other neuromuscular blocking agents have not been studied.

Despite the electrophysiological effects of volatile agents, it is possible to induce SVT due to re-entry in most patients. A technique utilizing opioids, nitrous oxide, and a low concentration of volatile is acceptable, and sevoflurane may be preferable to isoflurane in this circumstance. The successful use of propofol for maintenance of anesthesia during EP studies for re-entry tachycardia has been described, but it remains uncertain whether the risk of false negative studies is reduced in comparison to volatile anaesthetics.

Anesthetic drugs and automatic tachycardia

Ectopic arrhythmias resulting from increased automaticity behave differently under anesthesia. Many of the extrinsic factors, which promote automaticity, are minimized during steady state anesthesia. Typically, catecholamine levels are low and cardiac work is decreased.

The direct effects of inhalational anesthetics on automaticity are complex. As described above, the abnormal behavior of these foci is related to an acceleration of spontaneous depolarization of the cell membrane. Halothane reduces the
rate of depolarization in sinoatrial cells producing a predictable reduction in HR.76 However, when uninjured Purkinje cells exposed to epinephrine are exposed to halothane, the rate of spontaneous depolarization is increased.77 The most likely substrate for ectopic tachycardia is injured Purkinje cells already demonstrating increased automaticity. Volatile agents do not affect these cells or their response to epinephrine.77 In intact hearts, halothane decreased the ability to induce ventricular tachycardia in dogs though enflurane had no effect on the ability to induce ventricular tachycardia during human EP studies.78,79

In a series of 150 patients with SVT, anesthetized with propofol infusions, seven patients had arrhythmias due to increased automaticity.75 The arrhythmia could not be induced in four of these patients despite the infusion of isoproterenol, though EP studies were successful in all of the 143 patients with re-entry tachycardia. In a further case report an incessant SVT due to increased automaticity was terminated by propofol.80

Given this data it is difficult to suggest a best anesthetic technique for EP studies in patients with ectopic tachycardia. Propofol should be avoided, as should higher doses of narcotics. Limiting the dose of volatile anesthetics during attempts to induce the tachycardia and the replacement of endogenous sympathetic activity with sympathomimetic drugs such as isoproterenol is one approach. The avoidance of general anesthesia can be considered in older patients; however, EP studies for automatic tachycardia are feasible in anesthetized patients. In a series of 12 children with automatic tachycardia, anesthesia did not appear to add to the difficulties in studying the arrhythmia.81

**Implantation of pacemakers and defibrillators**

Pacemaker implantation is less common in children than in adults.82 It is indicated for complete heart block (congenital or acquired) or for sinus node dysfunction leading to symptomatic bradyarrhythmia. Bradycardia may complicate CHD or be surgically produced. Innovative indications include anti-tachycardia pacing of SVT and implantation of defibrillators.

In cooperative older patients the procedure can be performed awake, but for small children general anesthesia is required. Anesthesia may be associated with worsening of bradycardia. Treatment with atropine (not effective for complete heart block) and isoproterenol may be instituted followed by the rapid placement of a temporary pacemaker wire, or the use of transthoracic or esophageal pacing. Transthoracic pacing requires a general anesthetic and possibly muscle relaxation; however, placement of pacing pads prior to induction of anesthesia facilitates treatment.

There are practical problems associated with placement of pacemakers in small children. Wires must be sufficiently long to accommodate patient growth and subcutaneous placement of the pacemaker may be impossible (the abdomen is a commonly used site). Epicardial wires are required in small infants and when access to the heart via the venous system is not possible (post-Fontan).

Implantation of defibrillators is a rare procedure in children and is indicated for life-threatening ventricular arrhythmias.83 Implantation is similar to implantation of pacemakers; however, it is usual to test the defibrillator by induction of ventricular fibrillation which is unpleasant for a conscious patient. Indications include isolated arrhythmias associated with long QT syndromes and patients with hypertrophic cardiomyopathy or arrhythmogenic right ventricular dysplasia who may have more general myocardial disease.84–86 Often, patients present after “near miss” sudden death or death of a close family member, and they and their families are often extremely anxious. Preoperative anxiety can be sufficient to induce arrhythmia and premedication with an anxiolytic is advisable.

**Conclusion**

The trend toward more invasive therapeutic cardiac catheterization procedures in younger, smaller, and sicker patients increases the potential for hemodynamic and respiratory instability. As such, preparation and vigilance for these procedures by the anesthesiologist is essential. Approaching these procedures as if the patient were undergoing surgery will help ensure the best outcome. Indeed, in the future an increasing number of combined surgical and catheter interventions may be performed at the same setting, in a modified catheterization laboratory that is fully equipped for surgery.87

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