Chapter

22

D-Transposition of the Great Arteries (Atrial Switch)

Denise C. Joffe and Michael J. Eisses

Case Scenario

A 35-year-old woman with dextro-transposition of the great arteries presented to the Adult Congenital Heart Disease clinic with a history of worsening exercise tolerance and shortness of breath. As a newborn she had a surgical balloon atrial septostomy performed, followed by a Mustard atrial switch procedure in infancy. A cardiac computed tomography scan now shows a possible small baffle leak, and thus she was scheduled for diagnostic cardiac catheterization with possible baffle leak closure.

Recent transthoracic echocardiogram shows:

- *Atrial switch repair of dextro-transposition of the great arteries*
- *Mildly depressed systemic right ventricular function*
- *No evidence of baffle stenosis or leaks*

Key Objectives

- Understand the anatomy of dextro-transposition of the great arteries and an atrial switch procedure.
- Describe the long-term sequelae of atrial switch anatomy in a patient with dextro-transposition.
- Describe the complications of an intraatrial baffle procedure.
- Describe the anesthetic considerations in patients with intraatrial baffle procedures who are undergoing noncardiac surgery and catheterization procedures.
- Describe the transesophageal echocardiogram in a patient with a Mustard procedure.

Pathophysiology

What is the anatomy of d-transposition of the great arteries?

In "simple" dextro-transposition of the great arteries (d-TGA) the great vessels originate from the wrong ventricle. The aortic valve is anterior and rightward of the pulmonic valve resulting in a discordant connection between the ventricles and the great arteries. Systemic blood return flows from the vena cavae to the right atrium (RA), right ventricle (RV), and aorta. Pulmonary venous blood flows from the left atrium (LA) to the left ventricle (LV) and exits the pulmonary artery (PA), returning again to the pulmonary veins. ***Blood flows in a parallel fashion, with deoxygenated blood recirculating to the body and oxygenated blood recirculating to the lungs.*** (See Chapter 21.) The resultant physiology is not compatible with life unless there is mixing between circulations. The most common source of mixing in simple d-TGA is an atrial septal defect (ASD), patent foramen ovale (PFO), or patent ductus arteriosus (PDA). In simple d-TGA there are no other associated cardiac defects.

What is the difference between an atrial switch and arterial switch procedure?

Although both procedures correct the abnormal physiology created by parallel circulations, they do so in significantly different ways. An ***atrial switch*** procedure reroutes atrial blood via a baffle to the correct great vessel, albeit via the wrong ventricle. (See Figure 22.1.) In contrast, in an ***arterial switch*** operation (ASO) the great vessels are transected and moved to the appropriate ventricle, and the coronary arteries are reimplanted into the neoaortic root, thereby correcting the anatomy. (Figure 21.4.) Importantly, in contrast to the atrial switch procedure, the LV becomes the systemic ventricle after an ASO.

Clinical Pearl

*An **atrial switch** procedure reroutes atrial blood via a baffle to the correct great vessel, albeit via the wrong ventricle. In contrast, in an **arterial switch** procedure the great vessels are transected and moved to the appropriate ventricle, and the coronary arteries are reimplanted into the neoaortic root, thereby correcting the anatomy. (See Figure 21.4.)*

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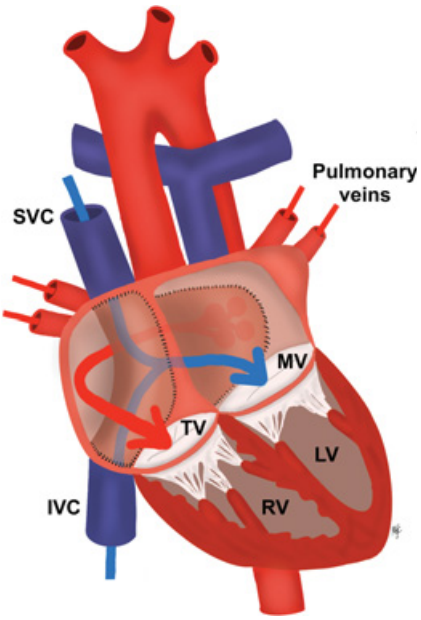

Figure 22.1 A Mustard repair for d-TGA. The SVC and IVC are baffled to form the SVP that directs blood to the MV and LV (blue arrow). Blood from the pulmonary veins enters posterior and to the right of the baffle, draining to the TV and RV (red arrow). IVC, inferior vena cava; LV, left ventricle; MV, mitral valve; RV, right ventricle; SVC, superior vena cava; TV, tricuspid valve. Courtesy of Michael Eisses, MD.

What is the history of management for patients with d-TGA?

Prior to the development of either the atrial switch or ASO, survival of neonates with d-TGA was improved by increasing intracardiac mixing of blood with a surgical atrial septostomy (Blalock–Hanlon–Thomas septostomy). Soon after, the introduction of the percutaneous balloon septostomy by Dr. Rashkind eliminated the need for surgery. However, a simple atrial septostomy did not eliminate shunts or correct the physiology, and patients succumbed early because of severe hypoxemia, congestive heart failure or pulmonary hypertension (PH). Survival beyond infancy came with the introduction of surgical repairs that corrected the discordant anatomy and eliminated the shunts. Although it was recognized from the outset that the optimal procedure to correct d-TGA was an ASO, the ability to transfer the coronary arteries required more sophisticated surgical techniques and equipment than was available at the time. In the late 1960s, Drs. Senning and Mustard developed ingenious but different ways of rerouting atrial blood to the opposite ventricle (see Figure 22.1), and atrial switch procedures became the procedure of choice for the next 15–20 years until the ASO became standard treatment for simple d-TGA. The atrial switch is no longer performed for children with d-TGA, but as of 2008, there were an estimated 9000 adult congenital heart disease (ACHD) patients in the United States who had undergone atrial switch procedures performed for the correction of d-TGA.

Do any cardiac lesions still require an atrial switch procedure as part of the repair?

As previously described, an atrial switch is no longer performed in patients with d-TGA. An atrial switch or a hemi-atrial switch (also called a hemi-Mustard or hemi-Senning; the inferior vena cava (IVC) alone is baffled to the mitral valve while the superior vena cava (SVC) is utilized in a bidirectional Glenn shunt) may be necessary in a small subset of patients with complex congenital heart disease (CHD). For example, patients with levo-TGA (l-TGA) or heterotaxy syndrome may require an atrial switch or hemi-switch as part of their repair. (See Chapter 23.) Therefore, although the number of patients requiring an atrial switch procedure has decreased significantly in the past 30 years, these procedures are still occasionally performed.

Clinical Pearl

Atrial switch procedures were historically performed in patients with d-TGA, and these patients are now between 35 and 60 years old. Atrial switch procedures are still occasionally performed in patients with complex anatomy such as levo-TGA and heterotaxy syndrome.

What is the difference between a Mustard and a Senning procedure?

The Senning procedure uses complicated incisions and suturing of pericardium in situ to create the baffle, whereas the Mustard procedure uses the patient's excised pericardium to create the baffles and is an easier procedure to master. Although hard to conceptualize, the shape of the single piece of pericardium used in creating a Mustard baffle is rectangular. In both procedures, the baffles are fashioned so that the SVC and IVC limbs join together in what appears to be a "Y" connection, creating a systemic venous pathway that then directs blood to the left-sided mitral valve (MV), into the LV, and then to the PA. Oxygenated pulmonary venous blood remains on the posterior side of the baffle until it reaches the RA and the tricuspid valve (TV) and is then ejected from the RV into the aorta. Close inspection of Figures 22.1 and 22.2 demonstrates why these are referred to as atrial switch procedures. Blood returns to the correct circulation albeit via the wrong ventricle. The LV is also referred to as the subpulmonary ventricle and the RV as the subaortic ventricle. (See Figure 22.3.)

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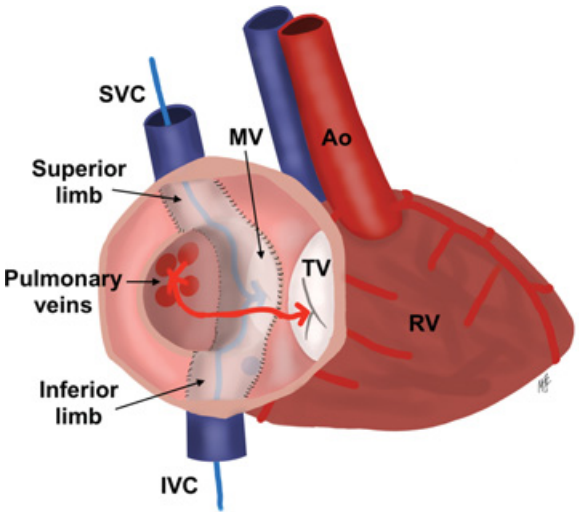

Figure 22.2 A Mustard procedure for d-TGA. A rectangular shaped baffle is used to create the SVP and PVP. The original interatrial septum is excised, and the baffle sutured as depicted creating the pathways. The PVP (red arrow) opens posteriorly and leads to the TV and RV. The SVP (blue arrow) is composed of the superior and inferior limbs that merge and are directed to the MV and LV. Ao, aorta; IVC, inferior vena cava; PA, pulmonary artery; RV, right ventricle; TV, tricuspid valve. Courtesy of Michael Eisses, MD.

It is impossible to distinguish the type of atrial switch using imaging techniques, but it is important to know whether the patient had a Mustard or Senning procedure because certain complications are more common depending on the procedure.

What are the long-term sequelae for patients with atrial switch anatomy after correction of d-TGA?

Common complications center around the constructed intraatrial baffles, including atrial arrhythmias, sinus node dysfunction, and baffle leaks and stenoses. Other long-term sequelae include systemic ventricular (RV) dysfunction and systemic atrioventricular valve regurgitation (for these patients, TR). (See Figure 22.4.)

Atrial arrhythmias (e.g., atrial flutter, supraventricular reentrant tachycardias) and sinus node dysfunction (e.g., junctional rhythm and atrioventricular block) are a common problem, occurring in up to 30% of patients by the age of 20 years. The etiology is likely multifactorial and includes extensive baffle suture lines across the atria with subsequent fibrosis, although injury to the sinus node artery itself during surgery is possible. Abnormal looping of the ventricles also contributes to heart block. Between 20% and 50% of patients are pacemaker-dependent by age 50 years, with many requiring implantable cardioverter defibrillators (ICDs). Lead placement is backwards in these patients and can result in confusion, as leads are seen on the left side of the heart. The atrial lead is placed in the left atrial appendage and ventricular leads in the LV (subpulmonary atrium and ventricle), unlike the typical lead placement in the RA appendage and RV of a normal heart.

Symptoms of ***baffle obstruction*** depend on which venous pathway is affected. Systemic venous baffle obstruction is more common after the Mustard procedure whereas pulmonary venous baffle obstruction is more common after the Senning procedure. Obstruction of the systemic venous baffle occurs in up to 30% of patients. The superior limb is more susceptible because it is smaller than the inferior limb and also often has pacing leads. Patients usually have few signs and symptoms because obstruction generally develops over time, and the SVC can decompress via the azygous system to the inferior limb. Patients with obstruction of the inferior limb may present with ascites and liver dysfunction. Obstruction of the pulmonary venous baffle usually occurs at the distal end of the baffle as the pulmonary venous blood is directed into the TV. (See Figure 22.4.) Patients usually present with symptoms similar to those of mitral stenosis, including progressive shortness of breath (SOB), pulmonary edema, and PH. Although it might be difficult to identify the baffle obstruction with transthoracic echocardiography (TTE), indirect evidence of PH such as LV (subpulmonary) hypertrophy and elevated velocity of the MR jet should heighten the suspicion for obstruction and prompt further imaging investigation.

Baffle leaks are less common than baffle obstruction, but leaks occur in about 15% of patients. They result in a predominantly left-to-right shunt akin to an ASD from the higher pressure pulmonary venous pathway to the lower pressure systemic venous pathway, and cause symptoms of pulmonary overcirculation such as SOB and exercise intolerance. Most small defects are well tolerated and do not require closure as the hemodynamic burden is not significant. Some, however, may require closure in order to avoid systemic emboli when pacemaker or ICD leads are necessary. In the presence of systemic venous baffle stenosis and a proximal leak, patients can develop symptomatic right-to-left-shunting.

Clinical Pearl

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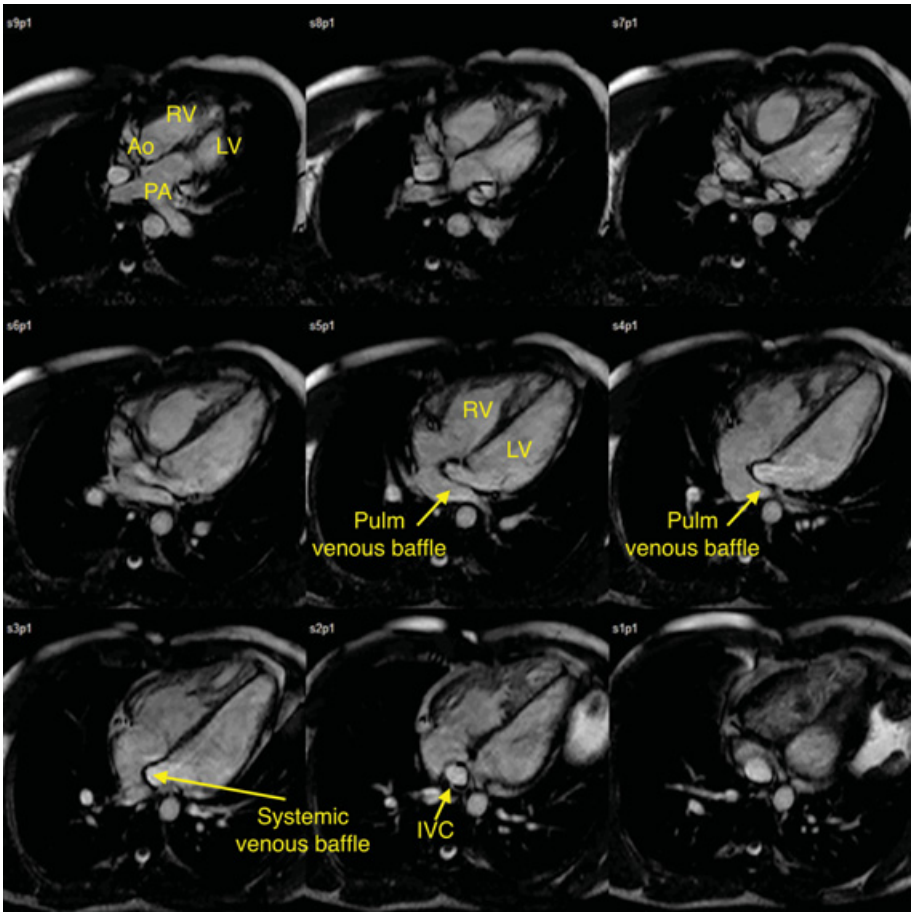

Figure 22.3 d-Transposition of the great arteries status post atrial baffle (Senning). Panel of four-chamber magnetic resonance imaging showing transposition of the great arteries status post atrial baffle procedure. The pulmonary veins are baffled to the right ventricle and the vena cavae are both baffled to the left ventricle. The resulting circulation is a systemic right ventricle connected to the aorta and a subpulmonary left ventricle connected to the pulmonary artery. Courtesy of Michael Taylor, MD.

Clinical Pearl

Between 20% and 50% of patients are pacemaker-dependent by age 50 years, with many requiring implantable cardioverter defibrillators. Lead placement is backwards in these patients and can result in confusion, since leads are seen on the left side of the heart. The atrial lead is placed in the left atrial appendage and ventricular leads in the LV (subpulmonary atrium and ventricle), unlike the typical lead placement in the RA appendage and RV of a normal heart.

How are baffle complications treated?

Whenever possible, transcatheter procedures such as balloon dilation, with or without stent placement, are used to treat baffle stenosis. In many cases of superior limb stenosis (i.e., SVC), the procedure is complicated by the presence of pacing leads entering the SVC. To place a stent, the pacing wires either have to be removed with laser therapy and then replaced, or the leads can be left in place and "jailed" by the stent. There are rare scenarios in which "jailing" leads may be indicated such as when the patient is being listed for heart transplant or if removing the leads poses prohibitive risk. Baffle leaks are treated with device closure. Surgical revisions are rarely necessary and have been plagued by a high operative mortality.

Although baffle-related complications may be difficult to identify by TTE, other noninvasive imaging techniques such as cardiovascular magnetic resonance imaging (CMRI) or cardiac computed tomography (CT) may offer improved visualization. In cases when the diagnosis remains unclear, such as the patient presented in the scenario, TEE performed during cardiac catheterization may confirm the diagnosis of a baffle complication and allow simultaneous treatment with a structural intervention.

How well does the RV function as the systemic ventricle?

Systemic RV function is largely preserved until the fourth decade of life, after which it begins to deteriorate. Causes of RV failure are multifactorial, including differences in

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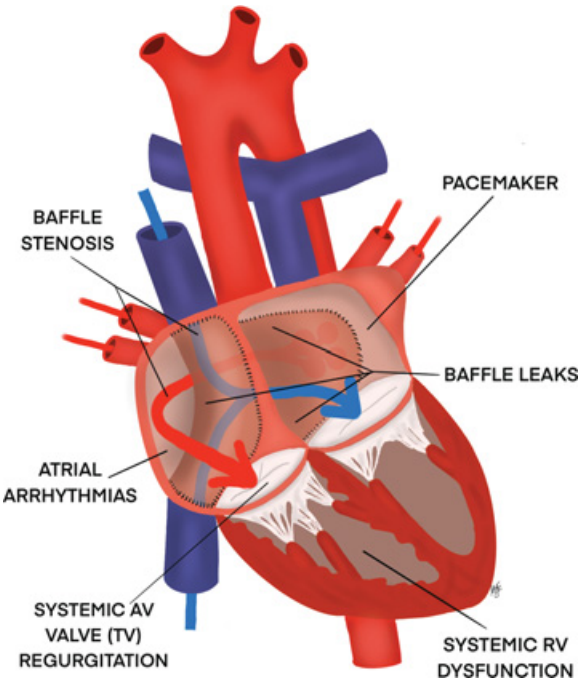

Figure 22.4 **Common complications of a Mustard repair for d-TGA.** Courtesy of Michael Eisses, MD.

geometric shape, coronary perfusion, and myocardial fiber orientation compared to the morphologic LV, with the end result that the RV is disadvantaged as a systemic ventricle. Also, the presence of TR significantly exacerbates systemic RV dysfunction, creating a cycle of worsening regurgitation and RV dysfunction.

Clinical Pearl

Despite the fact that the RV is not well suited to perform as the systemic ventricle, RV function is mostly preserved until the fourth decade of life. Tricuspid regurgitation is poorly tolerated in patients with a systemic RV.

How is RV function assessed?

With TTE, RV function is usually described in qualitative terms, ranging from normal to severely depressed function. However, the use of quantitative methods such as tricuspid annular plane systolic excursion (TAPSE), fractional area change, ejection fraction, and tissue Doppler may provide more objective assessment of RV function. The caveat is that echocardiography results are confounded by the frequent presence of significant TR, which can falsely infer good ventricular function as the aforementioned RV measures are preload and afterload dependent. These techniques are also plagued by the difficulty of evaluating function because of the geometry of the RV (whether it be a normal RV or systemic RV). Three-dimensional strain echocardiography shows promise but has not been validated. Cardiac magnetic resonance imaging and/or cardiac CT are commonly used to provide a more reliable quantitative assessment of ventricular function using volume analysis and can provide an evaluation of anatomy. However, a significant number of patients are not eligible for CMRI due to the presence of a pacemaker.

Clinical Pearl

Transthoracic echocardiography can overestimate right ventricular function in the presence of TR.

What is the survival and functional status of patients after atrial switch procedures?

Historically, survival has been 80%–90% at 20 years and 68% at 40 years in patients with a Mustard procedure. Survival rates are marginally better in those with a Senning procedure. Most adults are in New York Heart Association Class I or II and are able to work and function quite normally. About half of deaths are sudden, suggesting an arrhythmogenic mechanism, with rapid atrial flutter or ventricular arrhythmias thought to be the likely mechanisms.

What is the recommended follow-up for patients who have undergone an atrial switch procedure?

According to the 2018 American Heart Association/American College of Cardiology guidelines for the management of adults with CHD, follow-up is recommended annually, with TTE to be performed every 2–3 years. Echocardiography is also the primary diagnostic imaging modality in the event of new symptoms. Cardiac magnetic resonance imaging, cardiac CT, and cardiopulmonary exercise testing are also recommended every few years. Cardiac catheterization is performed on an as-needed basis. The incidence of complications, especially sinus node dysfunction with accompanying TR and systemic ventricular dysfunction, increases with age, mandating closer follow-up in older patients.

Despite these recommendations, in reality follow-up is often lacking due to a large number of factors, including poor transfer and coordination of care with appropriate ACHD specialists, lack of availability and expertise of ACHD in the community, insurance and financial issues, and poor patient compliance. A surprising proportion of

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ACHD patients do not possess an adequate understanding of their cardiac disease or details of their repair and thus may struggle with self-advocacy and understanding the need for routine surveillance and care.

Unless the patient is in extremis, it is generally recommended that the ACHD patient with anything more than repaired simple CHD (ASD, VSD, or PDA) be followed by an ACHD cardiologist and cared for by ACHD specialists, including anesthesiologists, intensivists, and interventional radiologists.

In addition to the patient's CHD, it is equally important in the ACHD population to consider evaluation for acquired heart disease or other systemic diseases.

Clinical Pearl

Adult congenital heart disease patients frequently lack appropriate follow up surveillance and care. They should also be evaluated for acquired heart disease or other systemic diseases when indicated.

What key findings can be suggestive of potential complications?

Exercise tolerance is probably the best indicator of overall function in patients after an atrial switch procedure. Many symptoms develop gradually with decreasing cardiac function, so it is crucial to follow up with pointed questions, especially since poor conditioning can lead to similar symptoms. An overall decrease in exercise tolerance may be related to any aspect of abnormal anatomy from ventricular dysfunction to baffle complications. Specifically, paroxysmal dyspnea and orthopnea may point to pulmonary venous baffle obstruction or PH, and palpitations suggest atrial or ventricular arrhythmias. Syncope is very concerning, as it can be a symptom of malignant atrial or ventricular arrhythmias. Patients with ICDs should be asked if it has discharged; the device should be interrogated, and the history reviewed.

Clinical Pearl

Exercise tolerance is the best indicator of overall cardiac function. Poor exercise tolerance may be related to any of several important complications of an atrial switch and should be evaluated before anything but emergent surgery.

On physical examination, a low heart rate suggests sinus node dysfunction, and facial plethora may be related to obstruction of the SVC systemic venous baffle. Cyanosis is unusual and may suggest a combination of baffle stenosis and a baffle leak or PH. The location of surgical scars may help reconstruct the surgical history if the patient is a poor historian. (See Table 22.1.). The presence of hepatomegaly may be related to inferior baffle obstruction or PH. The cardiac examination should focus on rate, rhythm, and the presence and timing of murmurs, which can help distinguish atrioventricular valve regurgitation or a residual VSD (pansystolic) from the ejection murmur more consistent with pulmonary or aortic valve stenosis. However, heart sounds can be misleading, since the aortic valve is the more anterior of the two great vessels. A loud, split S2 may be normal and not a result of PH.

Table 22.1 Surgical Procedures and Scars Associated with d-TGA

Scar Location	Surgical Procedure
Midline	Atrial switch procedure
Left thoracotomy	Coarctation repair
Right thoracotomy	Surgical septostomy
Pacer in left subclavian pocket	Normal venous anatomy – normal pacer placement
Pacer in right subclavian pocket	Presence of left SVC or thrombosed right subclavian vein

An electrocardiogram (ECG) should be performed or available. Right ventricular hypertrophy and right axis deviation are expected findings in patients with a systemic RV. Rate and rhythm can help determine sinus node function. If a pacemaker is present, paced beats should be noted, and formal pacemaker evaluation may be indicated if concerns arise from the history and physical examination. A chest radiograph will demonstrate sternal wires. In pacemaker-dependent patients, leads enter the left (subpulmonary) ventricle. Variable degrees of cardiomegaly and RV hypertrophy are usually present and consistent with a systemic RV.

Clinical Pearl

Red flags on physical examination may include lack of sinus rhythm, cyanosis, facial plethora, a pansystolic murmur (likely due to TR), and hepatomegaly.

Anesthetic Implications

What are the anesthetic considerations in patients who have undergone an atrial switch procedure?

The majority of ACHD patients are cared for in centers without specific expertise in CHD, so it is important for the

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anesthesiologist to have an understanding of the patient's disease and the previously described potential complications. A picture is worth a thousand words. Whenever possible, a copy of the patient's catheterization report or even a hand-sketched picture of the patient's heart often allows the anesthesiologist and other providers unfamiliar with the anatomy and repair to mentally reconstruct the direction of blood flow.

Clinical Pearls

Early consultation with an ACHD cardiologist is optimal for a patient with anything more than simple CHD; remote consultation may be necessary if no local provider is available. The most recent catheterization report with a picture of the patient's heart or a hand-drawn sketch of the anatomy and repair is extremely helpful to understand the direction of blood flow. Attaching it to the patient's bed or chart is very useful for all providers.

When transfer to a specialized center is not possible, the timing of the preoperative assessment should be planned to allow sufficient time before the procedure to obtain records from the patient's cardiologist. If records are not available, then a focused history and physical examination with an emphasis on exercise capacity can help classify the patient's condition. Even if the focused examination suggests good exercise capacity it is ideal to request a TTE in order to confirm the diagnosis and rule out any unexpected findings, since patients may not provide reliable histories. This sequence is suggested if no prior cardiology visits or TTE are available for reference. High-risk patients include those with limited exercise capacity, dyspnea (especially at rest), palpitations, presence of an ICD, RV dysfunction, TR, and evidence of PH. Elective surgery should be deferred pending a cardiology consultation in all high-risk patients.

If the patient is deemed stable, low-risk, and undergoing a low-risk procedure, then anesthesia should proceed in a standard fashion similar to that for any patient, with these caveats:

- Systemic RV function may not be as good as described in studies, especially in the presence of TR.
- The presence of TR is not as well tolerated in patients with a systemic RV.
- Pacemaker and ICD function may require reprogramming (similar to any patient with a pacemaker or ICD), and there should be a low threshold for placing defibrillator pads, even in the absence of a history of tachyarrhythmias, especially if the location of the surgical site will limit access to the chest.

A low threshold for utilization of monitors such as TEE should be considered as long as the echocardiographer understands the anatomy. If volume status and ventricular function are the primary objectives of monitoring, then it is possible to "ignore" the intraatrial anatomy and just focus on ventricular function and volume status using standard views, such as a four-chamber view or transgastric mid-papillary view, with the caveat that the RV is the systemic ventricle. The RV will appear more hypertrophied and dilated and the LV will appear compressed compared to normal anatomy. However, the technique for "eyeballing" function should be similar to that in a normal patient. Ideally, if the provider is able, the additional assessment of TR will permit a more accurate grading of "true" RV function as described previously.

Clinical Pearl

Transesophageal echocardiography can be used to monitor ventricular function and volume status even by those with only basic TEE training by ignoring intraatrial anatomy and with the caveat that the RV is the systemic ventricle.

What are the anesthetic considerations in high-risk patients presenting for noncardiac surgery?

A high-risk patient having a low-risk procedure should be managed like other high-risk cardiac patients with similar pathology. For example, an atrial switch patient with poor RV function and TR is analogous to a patient with severely decreased LV function and MR. A patient with an atrial switch and either PH or atrial arrhythmias requires management similar to that for patients with normal anatomy and similar pathology. The need for invasive monitoring will depend on both the procedure and the particular hemodynamic risks for that patient. There are no specific considerations for arterial access, and arterial lines should be placed for indications similar to those in other patients. There are special considerations for central venous access, as discussed in the next question. In general, there should be a low threshold for postoperative admission to intensive care, even after a low-risk procedure, especially in non-ACHD hospitals, given the lack of familiarity of most providers with the disease.

When a high-risk patient requires a high-risk procedure, the optimal scenario is to have it performed in a specialized center with cardiologists, cardiac anesthesiologists, and intensivists familiar with ACHD. All the considerations previously mentioned apply; in addition, the ability to perform and interpret intraoperative TEE should be available.

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Options for advanced resuscitation with mechanical circulatory support (MCS) devices must be discussed in the context of the patient's anatomy. The choice of MCS device must consider the presence of intraatrial baffles and a systemic RV, which is morphologically distinct from an LV and may affect technical aspects of device placement. The preoperative visit should include determination of candidacy for MCS and whether the patient is a transplant candidate, as well as discussion of an advanced care directive.

What considerations exist when placing central venous lines in patients with an atrial switch?

If central venous access utilizing neck vessels is being considered due to patient or surgical factors, one must take into account the likely higher than normal risk of complications that could occur with line placement in a patient with a baffle. The use of fluoroscopy may facilitate placement, and the line should be placed by a proceduralist familiar with atrial baffle anatomy and knowledge of the expected course of the wire and catheter. The following factors should be considered prior to line placement:

1. ***Knowledge of the primary lesion***: For example, if the atrial switch was performed for d-TGA, both superior (internal jugular or subclavian vein) and inferior (femoral) venous pressure measurements should be identical, since they reflect systemic venous pressures, that is, central venous pressure (CVP), whereas, if a hemi-Mustard is performed for l-TGA, an upper body CVL measures Glenn pressure, that is pulmonary artery pressure, and a femoral venous line measures CVP, which are not the same. (See Figure 22.5A and B.)
2. ***Knowledge of the presence of baffle stenosis or leaks***: Known baffle stenosis can make line placement difficult and distort measurements. Baffle obstruction can be manifest by higher than expected pressure readings and a lack of phasic appearance if the line is superior to the obstruction. If there is resistance to advancement of the wire or catheter while placing a central venous line in the neck, there should be a low threshold to abandon placement, especially because obstruction of the superior limb of the atrial baffle is not uncommon, as discussed previously. Line placement through the femoral vein is also an option and avoids entering the heart and baffles.

Placing a central line in the neck in a patient with a baffle leak can result in inadvertent catheter misplacement or migration into the pulmonary venous baffle, which can theoretically extend the tear. In addition, pressure measurements will reflect pulmonary venous pressure, not systemic venous pressure. Also given that a leak usually results in a left-to-right shunt, intracardiac saturations (RA, PA) will be unreliable.

3. ***The purpose of CVL placement***: If the line is being placed solely for volume resuscitation or medication infusion, then access via the femoral veins or a rapid infusion catheter (RIC) placed in a large upper extremity vein may be ideal, since it avoids the need to enter the heart. If the line is being placed for CVP measurement and/or medication infusions, femoral access may also be ideal, since it often allows accurate CVP measurement while avoiding the risk of entering the atrial baffles. When a PA catheter (PAC) is necessary to measure PA pressures or for cardiac output monitoring, the accuracy of some measurements (mixed venous or PA saturation and cardiac output) depends on the absence of baffle leaks or significant regurgitation of the subpulmonary atrioventricular valve (MR in the case of d-TGA and TR in the setting of l-TGA). Pulmonary artery catheter placement can be challenging, since it can be difficult to enter the PA from the LV, arguing for placement by an experienced provider using fluoroscopy. Pulmonary artery catheters should be used only when absolutely indicated for patient management.

What are the anesthetic considerations for cardiac catheterization procedures in a patient with an atrial switch?

In this population, a cardiac catheterization is typically performed to diagnose and treat a baffle problem, to diagnose PH and confirm its etiology, or as part of a pretransplant cardiac evaluation. Cardiac catheterizations may also be done as part of an electrophysiology procedure to treat arrythmias or to place or repair transvenous pacing or ICD systems.

Anesthetic management of ACHD patients in the catheterization laboratory depends on patient-related factors such as the diagnosis and severity of disease, the history of surgical procedures, and additional comorbidities. Many ACHD patients have significant anxiety and poor coping skills during medical procedures and prefer general anesthesia because that is what they received during their prior pediatric procedures. In addition, the need for TEE to guide device placement may mandate the use of general endotracheal anesthesia. When intracardiac echocardiography is used, the procedure can be performed with sedation alone, although even then a general anesthetic with a supraglottic device or endotracheal tube is frequently required for patient safety because of the duration or complexity of the case.

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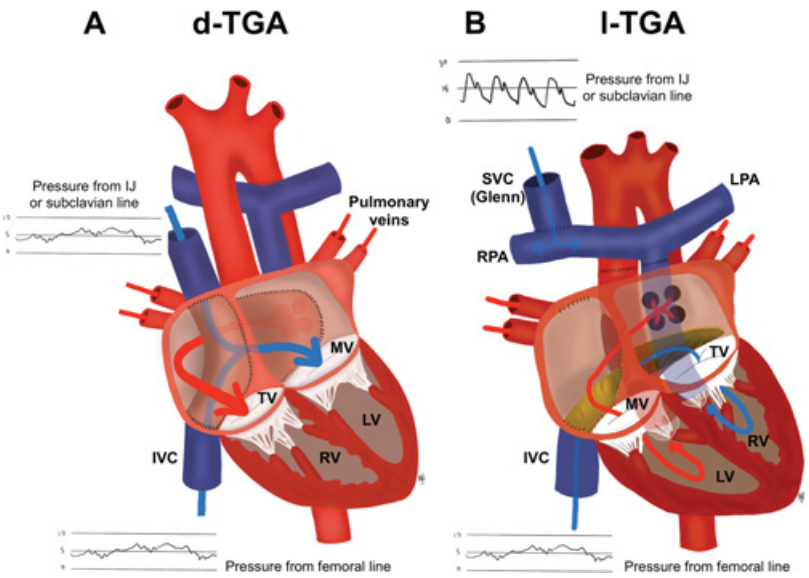

Figure 22.5 Central venous pressures. Depending on the original anatomy and the presence of a complete Mustard or hemi-Mustard, venous pressures from a central line in the neck versus a central femoral line may be different. In (A), both SVC and IVC pathways should reflect central venous pressure, whereas in (B), the IVC reflects central venous pressure and the SVC reflects PAP. IVC, inferior vena cava; LPA, left pulmonary artery; LV, left ventricle; MV, mitral valve; PAP, pulmonary artery pressure; RAP, right atrial pressure; RPA, right pulmonary artery; RV, right ventricle; SVC, superior vena cava; TV, tricuspid valve. Courtesy of Michael Eisses, MD.

Regarding vascular access, a peripheral intravenous catheter is all that is required for induction unless the patient's clinical status is precarious and necessitates a preinduction arterial line. The cardiologist usually places femoral arterial and venous lines for the majority of procedures, and the venous line can be accessed by large-bore extension tubing if needed. Placing separate invasive lines may be warranted if the patient requires pre- or postprocedural arterial blood pressure monitoring or ongoing central venous access. Replacing a relatively large femoral venous sheath used for the catheterization with a smaller CVL for use in the postoperative period may be complicated by unacceptable bleeding around the smaller line, necessitating placement at an alternate site. A more permanent CVL in the upper body can be performed with fluoroscopy at the conclusion of the procedure.

The procedure begins with a diagnostic catheterization of the systemic venous chambers, including the superior and inferior baffles and pathways, as well as the LV and PA. Saturation measurements in these locations are used to determine systemic and pulmonary cardiac outputs and assess for the presence of shunts. The patient should be maintained on room air in order to use the Fick equation for cardiac output and shunt calculations. Room air also allows baseline measurements of cardiac pressures. A left heart catheterization is performed in a retrograde fashion when indicated. When there is a concern for a baffle leak or obstruction, the complete TEE is performed concurrently with the catheterization in order to evaluate the baffle, help localize pathology, and guide the procedure when indicated.

The postoperative disposition of the patient varies with the severity of the patient's disease, and the procedure performed. In general, if the procedure is limited to a diagnostic catheterization, most patients are discharged home. If a therapeutic procedure with device placement is performed, the patient is usually admitted for observation and repeat TTE prior to discharge.

How can the baffle be evaluated using TEE?

A physician sonographer with advanced training in echocardiography should perform the examination and help guide the interventionalist. A general description of the echocardiographic evaluation is provided in the text that follows, since it helps non-CHD echocardiography trained providers understand the anatomy. When imaging a patient with an atrial switch using TEE, the optimal views include the mid-esophageal four-chamber view and an orthogonal (perpendicular) plane. Landmarks are used to identify the pathway being evaluated. (See Table 22.2.)

The four-chamber view provides qualitative assessment of biventricular and atrioventricular valve function and looks similar to that of a normal patient except the LV often appears flattened by a dilated and hypertrophied systemic RV. (See Figures 22.6 and 22.7.) The tricuspid and mitral valves are in their normal position and should be evaluated for regurgitation. In this view, the baffle and pathways are usually well seen. The more posterior pathway is the pulmonary venous baffle. The entire pathway can be seen by identifying the pulmonary veins using 2D and Doppler techniques and then following this pathway

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Table 22.2 Landmarks Used to Identify the Systemic and Pulmonary Venous Pathways in Patients with d-TGA and Atrial Switch Procedures

Landmarks Used to Identify Pathway			
Systemic venous pathway	Find IVC and SVC and follow over to left	Pacer and defibrillator leads travel in pathway	Pathway in closest proximity and opening to the MV
Pulmonary venous pathway	Use color flow Doppler to find pulmonary vein (LUPV is easiest) and then follow pathway to the right.	The most posterior pathway	Pathway in closest proximity and opening to the TV

IVC, inferior vena cava; LUPV, left upper pulmonary vein; MV, mitral valve; SVC, superior vena cava; TV, tricuspid valve.

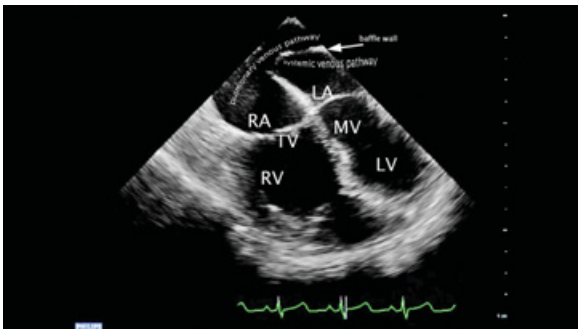

Figure 22.6 TEE ME four-chamber view. The ventricles appear in their usual position but note that the RV is larger than the LV, although the LV is slightly more dilated than expected because this patient has a significant baffle leak causing a left-to-right shunt similar to an ASD (causing LV dilation instead of RV dilation). In addition, there is right ventricular hypertrophy (the RV wall thickness is >5 mm). A baffle is seen in the atrium (labelled baffle wall). The pulmonary venous pathway is most posterior and is seen to empty into the TV. The systemic venous pathway is that portion of the LA in continuity with the MV. ASD, atrial septal defect; LA, left atrium; LV, left ventricle; ME, mid-esophageal; RV, right ventricle; TV, tricuspid valve.

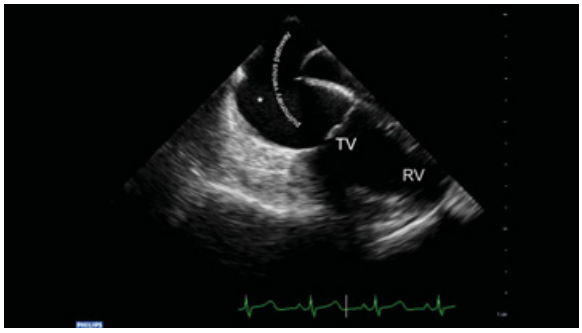

Figure 22.7 The image above is rotated to visualize the RV. The pulmonary venous pathway is seen to empty into the TV and RV. The asterisk marks a common location of obstruction in this pathway. RV, right ventricle; TV, tricuspid valve.

rightward toward the TV. In the four-chamber view, the systemic venous baffle is seen on the right side of the screen, anterior to the pulmonary venous baffle. It empties into the MV and LV. To follow the course of the systemic venous baffle, the probe is inserted into the stomach from the four-chamber view and turned to the right. The liver and the IVC are imaged enabling identification of the inferior portion of the superior venous pathway (SVP), and as the probe is withdrawn it is followed over to the left. (See Figure 22.8.)

When imaging in a plane orthogonal to the four-chamber view, the probe is rotated from right to left (counterclockwise). (See Figure 22.9.). Images depend on the cutting plane relative to the baffle. Starting with the probe rotated all the way to the right, the IVC drains to the inferior limb of the SVP. With further leftward rotation the superior (from SVC) and inferior (from IVC) limbs of the SVP are seen on either side of the pulmonary venous pathway (PVP), which courses from posterior to anterior. This view is analogous to the mid-esophageal (ME) bicaval view with the SVC on the right of the screen and the IVC on the left. With further leftward rotation, the RV and aorta are visualized, and both limbs of the SVP are seen to merge in the center. A portion of the PVP is seen posterior to the merger, and another is seen close to the TV. Then, as the probe is rotated counterclockwise, the SVP is seen to merge completely. The RV and aorta disappear as the LV and PA are visualized. During rotation, the most posterior pathway and the one closest to the TV is always the PVP. The SVP is always closest to the mitral valve. In addition, as the probe is rotated, both ventricles, outflow tracts, and semilunar valves are seen in the long axis. As in a normal patient, the RV and tricuspid valve are first seen. The aorta originates from the RV. With further counterclockwise rotation the LV, MV, and PA are visualized.

Aliasing during color-flow Doppler examination is highly suggestive of baffle obstruction. Baffle leaks are occasionally large enough to be visible on 2D imaging, but caution is warranted to not confuse echo dropout with a defect. Echo dropout is created when the echo beam and structure of interest are almost parallel, resulting

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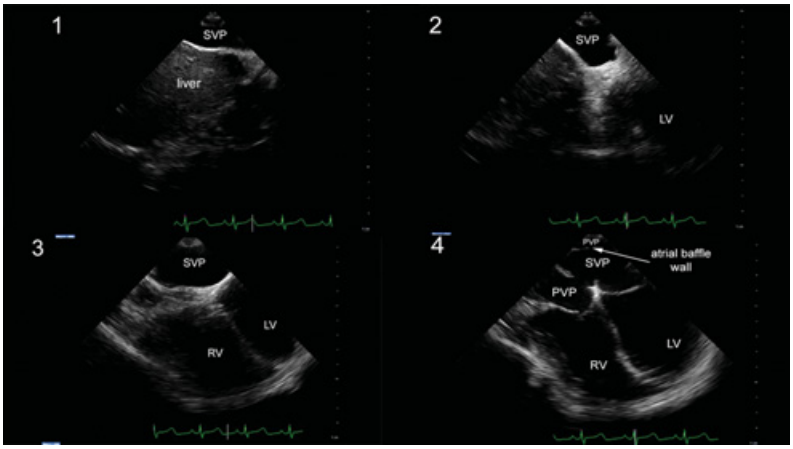

Figure 22.8 Sequence of TEE images during withdrawal from TG to ME levels and a sweep to the left at 0°. (1) TG position at the level of the liver. The IVC is visualized and identifies the SVP. (2–3) The pathway is kept in the center of the image as the probe is slowly withdrawn to the ME level and rotated to the left. (4) With the probe all the way to the left, the SVP opens to the MV. It is anterior to the PVP and is separated from it by the atrial baffle wall. IVC, inferior vena caval; ME, mid-esophageal; PVP, pulmonary venous pathway; SVP, superior venous pathway; TG, transgastric.

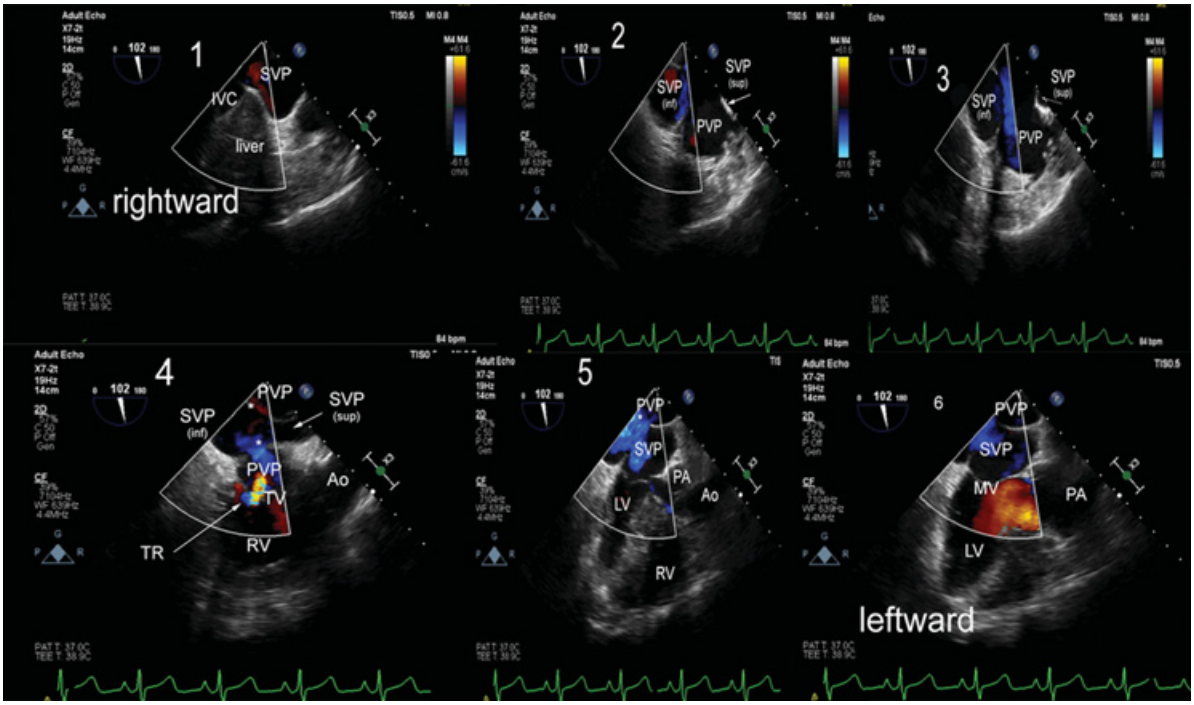

Figure 22.9 TEE color flow Doppler 102° rotation (orthogonal) sweep from the right to the left (counterclockwise). The right of the screen is superior, the left inferior, the bottom anterior, and the top posterior. (1) The IVC is located and allows identification of the inferior portions of the SVP. (2–3) The probe is withdrawn to the ME level and turned leftward. Both limbs of the SVP are seen on either side of the PVP. (4) The first ventricle visualized during the rotation is the RV. The RV, TV, and Ao are seen. Both limbs of the SVP are seen to merge. The PVP is seen posterior and closest to the TV valve. Mild to moderate TR is seen. Two baffle leaks are seen in this image (asterisks). (5) With further leftward rotation both ventricles are seen. The pathway closest to the MV is the SVP. The more posterior baffle leak is now seen between the PVP and SVP (asterisk). Note, the shunt here is left-to-right, which is the predominant overall direction of the shunt in this patient. (6) With the probe rotated all the way to the left, the LV and PA are seen, and no further shunt is visible. Ao, aorta, Inf, inferior; IVC, inferior vena cava; ME, mid-esophageal; PA, pulmonary artery; PVP, pulmonary venous pathway; RV, right ventricle; Sup, superior; SVP, superior venous pathway; TR, tricuspid regurgitation; TV, tricuspid valve.

in minimal return of echo signals and a void in the picture. A defect on 2D imaging, especially when it is an area parallel to beam, should always be verified in another view and with color-flow Doppler. (See Figure 22.10.) When inconclusive, agitated saline can be used to verify a communication. Other supportive findings include dilation of the systemic venous atrium and ventricle (in this case the LA and LV).

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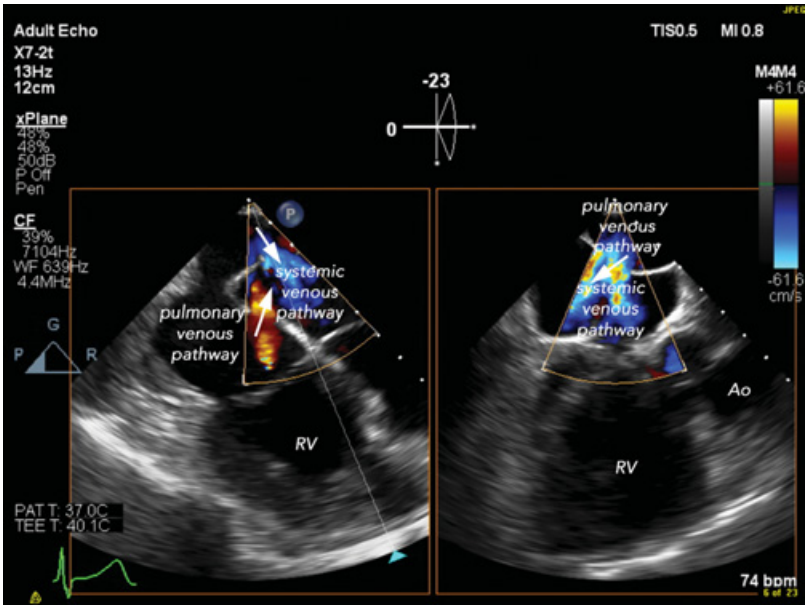

Figure 22.10 A TEE color flow Doppler ME four-chamber view and an orthogonal cut at the level of the posterior leak. This is easily performed using a TEE probe capable of biplane (X-plane) imaging. Both leaks are seen in the left-sided image, and the direction of flow is left-to-right (arrows). Although the dimensions are not measured in these images, they are quite large and measure at least 1 cm in this plane (based on the dots on the side of the image which are 1 cm apart). The right-sided image cuts through the more leftward and posterior defect only (see blue triangle at the bottom of the image and the location of the cutting plane). Ao, aorta; CFD, color flow Doppler; ME, mid-esophageal; RV, right ventricle.

Transcatheter procedures are used to treat baffle stenosis and leaks and have a high success rate. (See Figure 22.11.) TEE and fluoroscopy are used to guide the proceduralist and assess the results.

The majority of ACHD patients with atrial switch procedures initially had d-TGA, although atrial switch procedures are still occasionally performed for other complex lesions. It is necessary to understand the anatomy of the repair as well as the original lesion in order to appreciate all the anesthetic implications. Most patients remain a high-risk subgroup to anesthetize and given the complexity of the repair are best cared for in specialized centers.

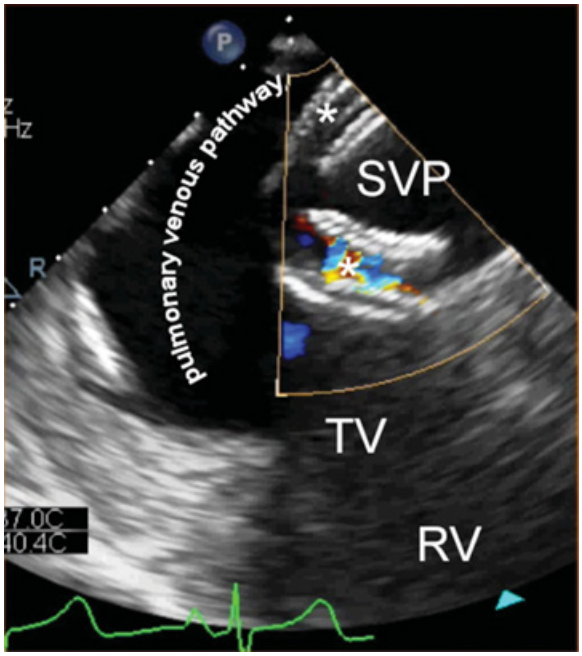

Figure 22.11 An ME four-chamber color flow Doppler view with the probe turned toward the right. This is a zoomed imaged similar to Figure 22.6. Two septal occlusion devices are visible (marked with an asterisks).

Suggested Reading

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