

Pulmonary Atresia, Ventricular Septal Defect, Major Aortopulmonary Collaterals

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Pulmonary atresia with ventricular septal defect (PA/VSD) and major aortopulmonary collateral arteries (MAPCAs) is a cyanotic CHD on the spectrum of tetralogy of Fallot (TOF) with ultimate pulmonary obstruction in the form of valve atresia; it is therefore also referred to as tetralogy of Fallot with pulmonary atresia. The PA tree is much more likely to be abnormal in PA/VSD than in TOF.

There is a wide variability in the anatomy of patients with PA/VSD/MAPCAs. On one end of the spectrum, a patient will have well-formed confluent branch PAs supplied by a ductus arteriosus and no MAPCAs, while at the other end of the spectrum, the patient will have no native PAs and all pulmonary supply will be dependent on MAPCAs. The main PA (MPA) segment may be absent, may present as a fibroelastic cord without a lumen, or may have varying degrees of hypoplasia. The native branch PAs may be confluent or discontinuous, and may be normal, hypoplastic, or absent. Ductal origin of one or both branch PAs may be present. Segmental and subsegmental arteries may likewise be present or absent.

MAPCAs arise from the descending thoracic aorta (or the brachiocephalic vessels) as vestiges of the primitive PA supply and, in the absence of native PA branches, comprise the sole source of blood flow to some lung segments. In this disease, some lung segments may have native PA supply, other segments may only have MAPCA supply, and yet others may have dual supply from both MAPCAs and native PA branches. The ductus arteriosus may be present or absent; rarely, bilateral PDAs are present. The RV is typically normal or dilated in size.

As with other conotruncal defects, there is a strong association between PA/VSD and DiGeorge (22q11 deletion) syndrome. A right aortic arch is a common associated anatomic variant.

Pathophysiology and Clinical Presentation

The patient symptomatology and clinical presentation is dictated by the amount of pulmonary blood flow (Q_p) through the MAPCAs and the native PAs (via a PDA or communication with MAPCAs). In infants, Q_p can be highly dynamic due to changes in PVR, changes in the nature of the MAPCAs, and ductal closure (if applicable).

All neonates with a diagnosis of PA/VSD are admitted to the CICU or NICU and their management will vary based on the source and amount of pulmonary flow. Patients that are dependent on a PDA for adequate pulmonary blood flow are treated with PGE until a procedure is performed to secure a stable source of pulmonary blood flow. Patients that have robust MAPCAs may develop symptoms of pulmonary overcirculation, especially as PVR drops in the first few weeks of life, and will thus require diuretic therapy. If there is no PDA and MAPCAs are inadequate, the patient will be cyanotic and will require an early procedure to improve pulmonary blood flow. Patients that

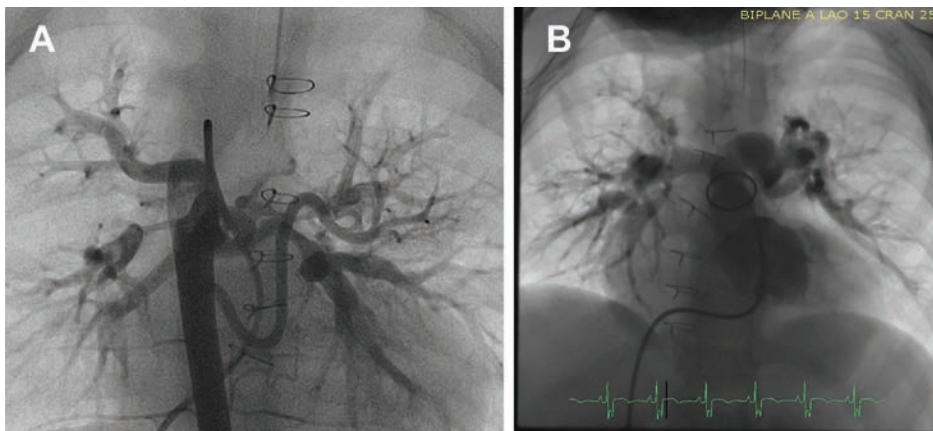


Figure 17-1. Cardiac catheterization on PA/VSD/MAPCAs. A) Catheterization showing multiple MAPCAs arising from the descending aorta. B) Catheterization showing MAPCAs unifocalized into the branch PAs with good overall pulmonary arborization.

develop stenosis of MAPCAs prior to unifocalization will experience a decrease in total Qp resulting in worsening hypoxemia or if they presented with overcirculation, improvement in symptoms.

The physical exam findings will depend on the overall Qp and status of the MAPCAs. A continuous murmur may be heard over lung segments supplied by MAPCAs. Infants with pulmonary overcirculation will develop failure to thrive, a gallop, pulmonary edema, and hepatomegaly. Hypoxemic patients will present with the expected sequelae including cyanosis and clubbing.

Diagnosis

- **CXR.** The cardiac silhouette takes on a boot shape, as seen in TOF. The quality of the pulmonary vascular markings is reflective of PA or MAPCA supply.
- **ECG.** RA enlargement and predominance of right-sided forces is common. Findings of left-heart enlargement (i.e., LA enlargement, biventricular hypertrophy) are seen in patients with large amounts of pulmonary blood flow.
- **Echocardiogram.** The appearance of the cardiac anatomy is similar to that in TOF, with a large VSD typical of conotruncal defects and aortic override. PA anatomy can be delineated to some degree but the images are frequently limited to the proximal native PAs (if present) and proximal MAPCAs. The presence or absence of a PDA should be documented. Aortic arch anatomy and coronary anatomy should also be assessed.
- **Cardiac catheterization (Figure 17-1).** Cardiac catheterization is performed on every patient with PA/VSD and MAPCAs prior to surgical intervention. Angiography is useful to demonstrate the number, size, and distribution of MAPCAs. Selective injection of each individual MAPCA is particularly helpful for mapping the pulmonary segmental supply. In the absence of a ductus arteriosus, pulmonary-vein

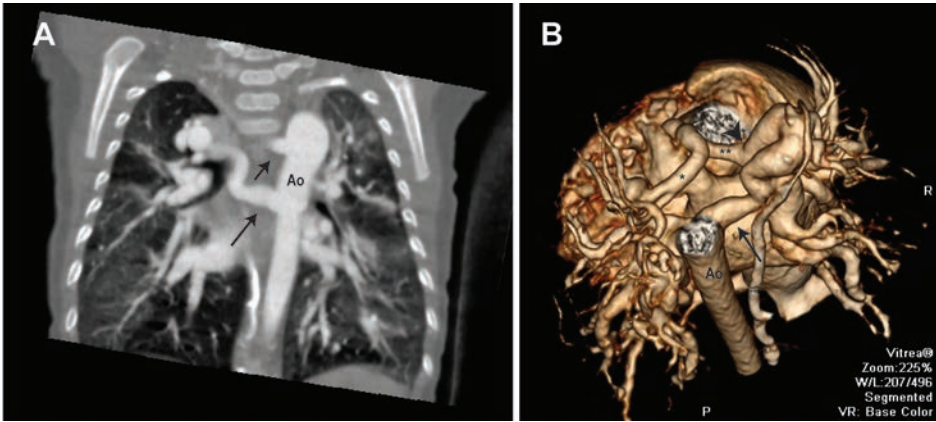


Figure 17-2. CTA of a patient with PA/VSD/MAPCAs. A) Coronal view showing 2 large MAPCAs (arrows) arising from the descending thoracic aorta (Ao). B) 3D reconstruction showing a large MAPCA (arrow) arising from the descending thoracic aorta and bifurcating into several large branches. One of the branches anastomoses to the left PA (arrowhead). * denotes the right PA and ** denotes the left PA.

wedge angiography is very important to define the presence and quality of the native PAs, as many patients will have diminutive but confluent branch PAs that go unrecognized with echocardiogram or CTA. The blood supply of all pulmonary segments must be accounted for. Lung segments with dual supply can be identified and associated MAPCAs intervened on. In some patients, especially those between stages of intervention, cardiac catheterization may also be useful to assess pressures within various MAPCAs. It is sometimes difficult to calculate Q_p , the relationship between pulmonary and systemic blood flow ($Q_p:Q_s$), and PVR due to the presence of multiple sources of pulmonary blood flow.

- **CTA (Figure 17-2).** Very useful to demonstrate the nature, course, and distribution of MAPCAs and native PAs. CTA is particularly useful as a complement to cardiac catheterization for surgical planning as it shows the course of the MAPCAs and their relationship with surrounding structures (e.g., trachea, bronchi, atria).

Management Strategy

The ultimate goal in the therapeutic pathway for most patients with PA/VSD/MAPCAs is to achieve a biventricular circulation with a single PA tree that arises from the RV and supplies all pulmonary segments. In some patients, a fully unifocalized PA tree supplying all lung segments is not achievable secondary to poor quality of PAs or MAPCAs. An abundance of severely stenotic pulmonary vessels or a limited number of pulmonary segments included in the pulmonary circulation may represent excessive afterload for the RV, precluding VSD closure.

Due to the heterogeneity of this lesion, management is individualized for each particular patient. However, the overall philosophy of management at TCH follows some general guidelines (Figure 17-3):

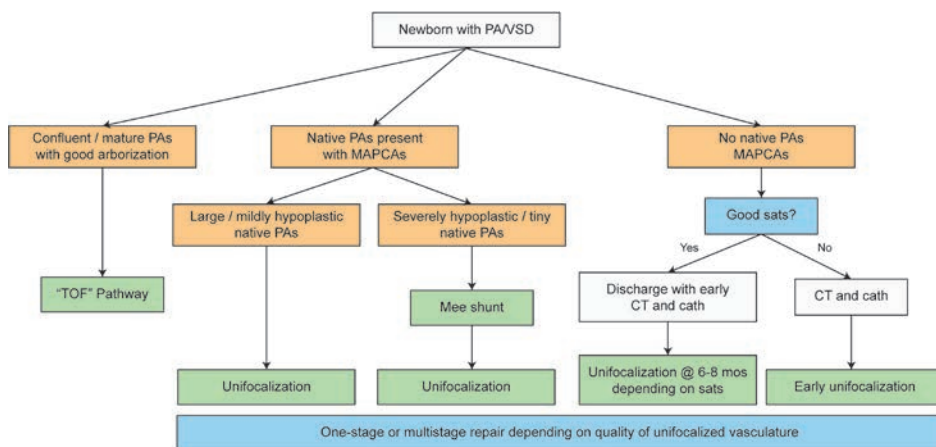


Figure 17-3. General algorithm for management of patients with PA/VSD/MAPCAs.

- Patients with well-formed confluent branch PAs supplied by a PDA.** These patients are analogous to patients with tetralogy of Fallot. They will be maintained on PGE and will then undergo a procedure to secure a stable source of pulmonary blood flow. Options include placement of a PDA stent or creation of a modified Blalock-Taussig-Thomas shunt (mBTTS) (see Chapter 38). Later in infancy, patients will undergo biventricular repair with a Rastelli procedure (see below). In some circumstances, the patients may undergo a single-stage repair with a Rastelli procedure at the newborn stage.
- Patients with hypoplastic branch PAs and MAPCAs.** A large proportion of patients with PA/VSD/MAPCAs will have diminutive branch PAs that may not be seen on CTA due to the lack of contrast flow into them. This is why a good pulmonary-venous angiography in the cardiac catheterization lab is very important. The overall strategy for these patients is to promote the growth of the confluent branch PAs in order to use as a scaffold to unifocalize relevant MAPCAs. These patients generally present with oxygen saturations that can vary from the 70s to the 90s, depending on the quality of the MAPCAs. As such, it is not usually necessary to perform any procedure during the newborn period. Patients are usually discharged home and undergo assessment of PAs and MAPCAs with a combination of cardiac catheterization and CTA. If the branch PAs are diminutive, a central shunt (Mee shunt) is performed (see below), usually at 3-6 months of age. An mBTTS may be used in patients that have slightly larger PAs in order to avoid overcirculation but still promote PA growth. The PA anatomy is reassessed later in infancy and the patient is then put forward for surgical repair in the form of a Rastelli procedure with or without unifocalization (see below).
- Patients with no native central PAs and sole MAPCA supply to the lungs.** This is the most challenging group of patients with PA/VSD/MAPCAs. Despite the lack of visualized central PAs, many patients will actually have diminutive central

PAs that were not seen on preoperative imaging but may be usable as part of the surgical strategy. After full assessment of MAPCAs with CTA and angiography, patients are taken to the OR, usually in late infancy. If diminutive central PAs are found on mediastinal exploration, a Mee shunt may be performed. Otherwise, all unifocalizable MAPCAs are brought together to create a central confluence that is then connected to either an mBTTS or a nonvalved conduit arising from the RV. Patients are then reassessed later in life with cardiac catheterization and CTA, and depending on the pulmonary arborization, the decision is made whether to proceed with a Rastelli procedure or not.

Anesthetic Considerations

Patients with PA/VSD/MAPCAs require frequent anesthetics for diagnostic studies, cardiac catheterizations, and surgical procedures. These patients are managed under general anesthesia with endotracheal intubation and mechanical ventilation due to the little pulmonary reserve secondary to the low Q_p.

Preoperative Considerations

- **Difficult airway.** Patients with 22q11 deletion syndrome have micrognathia with the potential of a difficult airway. In addition, the trachea is usually short and mainstem bronchial intubation is possible.
- **Cyanosis.** It is important to assess for polycythemia as part of the preoperative evaluation. A hematocrit >65% is linked to impaired microvascular perfusion, hyperviscosity syndrome, and thrombosis.
- **Increased bleeding risk.** Cyanosis is associated with an increased bleeding risk due to thrombocytopenia and factor deficiencies.

General Anesthetic Considerations

The speed of inhalation induction of anesthesia is slower in patients with right-to-left shunting due to the low Q_p. On the other hand, IV induction bypasses the first-pass pulmonary uptake, making IV induction faster. Due to the decreased Q_p, there are lung areas that are ventilated but not perfused, causing a ventilation/perfusion (V/Q) mismatch. The use of ETCO₂ monitoring to assess ventilation may become unreliable due to the mismatch and lead to respiratory acidosis. As such, it is important to monitor arterial blood gases in longer procedures.

Spasm of MAPCAs during surgical manipulation and/or catheter interventions is possible and can lead to profound cyanosis. Stopping the stimulation, deepening anesthesia, treating acidosis, and increasing BP usually relieves the spasm.

Procedure-Specific Anesthetic Considerations

- **Cardiac catheterization.** Interventional rehabilitation of PA branches is associated with 2 dreaded complications for the anesthesiologist: intrapulmonary bleeding and lung reperfusion injury. Intrapulmonary bleeding is easily diagnosed by red blood in the airway and decreased lung compliance. Lung reperfusion injury also manifest with decreased lung compliance and mostly pink, frothy secretions. Most of the time, these complications are self-limited and can be medically managed with

100% FiO₂ and an increase in PEEP. Occasionally, one-lung ventilation is needed to protect the healthy lung. The last resort, if unable to ventilate and oxygenate, is the use of rescue ECMO.

- **Unifocalization procedures.** Unifocalization procedures performed through a thoracotomy require lung isolation techniques when feasible (>10 kg). This can lead to alterations in dead space and respiratory acidosis. The use of regional anesthetic techniques (i.e., thoracic epidural or paravertebral blocks) are useful adjuvants to general anesthesia in order to minimize postoperative pain.
- **Complete repair and PA augmentation.** The risk of bleeding is high in these procedures due to repeated surgeries, multiple suture lines, and underlying cyanosis. Antifibrinolytic use and point-of-care coagulation testing are useful to minimize the use of blood products. Postrepair RV dysfunction is common despite the use of inotropic therapy (milrinone and low-dose epinephrine) to wean off CPB. iNO should be available, especially in high-risk patients. In addition to TEE, direct PA pressure measurement is usually performed at the surgical field.

Catheter-Based Intervention

As mentioned above, diagnostic catheterization plays an important role in defining the anatomy of the MAPCAs, the supply of each of the different pulmonary segments, and the anatomy of the PAs (via pulmonary venous wedge angiography).

Catheter-based therapy in PA/VSD/MAPCAs is focused on ensuring adequacy of pulmonary blood flow and PA architecture. Prior to surgical intervention, areas of dual supply that involve small MAPCAs may be identified and can be occluded.

Ductal stenting may be appropriate in patients with a PDA and clinical evidence of inadequate Q_p during a trial of ductal closure (see Chapter 38).

MAPCAs may become stenotic before or after unifocalization. Angioplasty or stenting of unifocalized MAPCAs is performed when stenosis is clinically significant, as this can result in cyanosis (if the VSD remains patent) or RV hypertension (if the VSD has been closed). A discussion with the surgical team is paramount to discuss what the best strategy for stenotic MAPCAs may be for a particular patient. Many patients require serial catheterizations to “rehabilitate” stenotic unifocalized MAPCAs. Cutting-balloon angioplasty is sometimes used in stenotic lesions that are particularly resistant to standard balloon angioplasty.

Patients who receive a conduit in the RV-PA position frequently require catheter-based interventions (angioplasty, stenting, or transcatheter valve placement) on the conduit.

In specific clinical scenarios, for patients possessing rare anatomic variants with patent MPA segments and isolated valve atresia, pulmonary valve perforation and valvuloplasty may be useful.

Surgical Intervention

As mentioned above, patients with this disease are managed in a multidisciplinary fashion and the therapeutic approach is individualized based on anatomy and physiology.

A series of surgical procedures can be performed on these patients, depending on the overall strategy (Figure 17-3).

Aortopulmonary Shunts

Patients that have diminutive native PAs can benefit from growth of those PAs by augmenting flow with the use of an aortopulmonary shunt. The shunt that we commonly use for patients with significantly hypoplastic PAs is the Mee shunt. This shunt involves detaching the main PA from the heart and creating an end-to-side anastomosis between the distal main PA and the ascending aorta. The procedure may be performed through a median sternotomy or a lateral thoracotomy. The pericardium is opened and the optimal location for the shunt on the ascending aorta is marked with a fine suture. The proximal main PA is ligated and the vessel is divided after controlling the branch PAs with fine tourniquets. A side-biting clamp is placed on the previously selected site on the ascending aorta, an incision is performed and enlarged with an aortic punch, and the anastomosis is performed with either a running fine suture or a set of interrupted sutures. The branch PAs are released and the sidebiting clamp is removed from the aorta. It is important to keep an adequate orientation of the branch PAs in order to avoid twisting of the diminutive PAs, in particular the right PA. The location of the shunt is usually located on the posterior aspect of the aorta and slightly to the left.

A Mee shunt is not a good alternative for patients with only mild or moderate hypoplasia of the PAs since they may become quite overcirculated. These patients are better served by an mBTTS (see Chapter 38).

Unifocalization

An important consideration in the workup of patients with MAPCAs is which MAPCAs are relevant and which MAPCAs are not. In general, the purpose is to achieve a pulmonary vascular bed that includes all or as many pulmonary segments as possible. A combination of CTA and cardiac catheterization usually allows to define whether each of the different 19 pulmonary segments is supplied by a native PA, only by a MAPCA, or have “dual supply”, meaning supply from both a native PA and a MAPCA. In general, MAPCAs that solely supply pulmonary segments should be unifocalized (unless they are very small and unifocalization is not technically feasible). MAPCAs that provide dual supply to pulmonary segments may be unifocalized (if they are large and accessible) or occluded (either in the cath lab or surgically). As a rule of thumb though, large MAPCAs are usually important sources of pulmonary blood supply and one should think twice before occluding them. It is important for the imaging (cardiac catheterization or CTA) to be recent (<3 months) in order to accurately reflect the status of the MAPCAs.

Unifocalization of MAPCAs can be performed either as part of a complete repair or as a separate procedure via either a median sternotomy or a thoracotomy. Ideally, MAPCAs are unifocalized into a native PA scaffold after allowing the native PAs to grow. However, MAPCAs may need to be unifocalized into a shunt or into a confluence that is supplied by a shunt or a conduit from the RV. The decision regarding the most optimal strategy is made depending on the individual characteristics of the patient and how this operation fits on the overall surgical and interventional roadmap designed for the particular patient. Another consideration is whether temporary occlusion of

the involved MAPCAs will be feasible without the need for CPB.

A preoperative CTA is very useful to define the particular location and course of MAPCAs and the relationship between the MAPCA and surrounding structures. If the procedure is performed as part of an intracardiac repair, it is useful to perform as much of the dissection as possible prior to administering heparin and instituting CPB. However, complete

dissection of the MAPCAs may be challenging due to the location of the vessels, usually behind the cardiac mass and the airway. Due to the size of the MAPCAs, it is desirable to control them with fine tourniquets immediately after CPB is instituted in order to reduce the usually massive amount of pulmonary venous return. If the MAPCAs have not been completely dissected on institution of CPB, the body is kept at normal temperature to allow the heart to eject the blood that is returning through the pulmonary veins (cooling decreases the effectiveness of the heart's contractility and can lead to cardiac distention). Once all MAPCAs are controlled, the patient is cooled down for the intracardiac portion of the procedure.

Rastelli Repair

The ultimate goal of the treatment strategy for these patients is a biventricular repair. The Rastelli repair involves the creation of a baffle between the LV and the aorta (therefore closing the VSD) and placement of an RV-PA conduit to allow the RV to eject blood into the lungs. The Rastelli procedure may be performed as part of a large operation involving unifocalization of MAPCAs or as a separate procedure.

The Rastelli procedure is performed through a median sternotomy with regular aortobicaval cannulation, using moderate hypothermia (25-28 °C). After cardioplegic arrest, a right atriotomy is performed and the left heart is vented through the atrial septum. Working through the atriotomy, the VSD is closed by creating an autologous pericardial (or Dacron®) baffle between the LV and the aorta. It is important to create the baffle in a way that allows it to bulge in order to prevent LVOT obstruction. This is achieved by advancing more on the patch and less on the heart tissue. A right ventriculotomy is then performed for placement of the conduit. Performing the VSD closure via the atriotomy allows the conduit to be placed more lateral on the heart and away from the sternum. The ventriculotomy should be created close to the shoulder of the heart (the obtuse margin) and away from the LAD and any large conal branches. It is important to excise RV muscle in order to allow unimpeded flow from the RV into the conduit. Options for conduits include cryopreserved aortic and pulmonary homografts, bovine jugular vein conduits (Contegra®), and composite porcine-valved Dacron® conduits (Hancock® conduits). Our preference for small children has been to implant cryopreserved homografts. In these cases, a small hood is created proximally between the RV and the homograft, using homograft tissue, autologous pericardium, or Gore-Tex® material. The placement of the RV-PA conduit can be performed either after ASD

TCH experience with PA/VSD/MAPCAs (1995-2013)

Number of patients: 107 (does not include patients without MAPCAs)

- Single ventricle palliation: 9 (8%)
- Single-stage repair: 17 (16%)
- Staged repair: 61 (57%)
- Awaiting intervention: 20 (19%)

closure and removal of the cross-clamp (with the heart beating), or prior to removal of the cross-clamp (with the heart stopped), depending on the expected difficulty and the length of the cross-clamp period. It is not customary for us to leave an ASD in place. An LA line and peritoneal dialysis catheter are routinely placed.

PA pressures are routinely measured after weaning off CPB. Some degree of pulmonary hypertension is not unusual. However, if PA pressures are significantly increased despite the use of iNO and there is no evidence of mechanical obstruction of the conduit or the branch PAs, one has to assume that the pulmonary vascular bed is not adequate enough to support a biventricular circulation at this time. It may be necessary to create a VSD fenestration or remove the VSD patch (and possibly place a PA band) in order to avoid RV failure.

Postoperative Management

The postoperative management depends on the procedure performed.

- **Aortopulmonary shunts.** Based on the size of the shunt, SVR, PVR, and intravascular volume-controlling strategies will be key to optimizing Qp:Qs. Special attention must be placed on appropriate pain control in patients with thoracotomies, as these are more painful than sternotomies and have a higher risk for atelectasis and possible longer ventilatory support requirements. See Chapter 38 for specific postoperative management of patients with aortopulmonary shunts.
- **Unifocalization.** It is important to know the size and anatomy of MAPCAs in order to provide adequate management to optimize Qp. Long, tortuous MAPCAs unifocalized into small PAs will need PVR-optimizing maneuvers such as deep sedation, consideration of neuromuscular blockade, achievement of FRC, oxygen supplementation, and iNO. Volume resuscitation may be useful in some instances. If not all MAPCAs have been unifocalized, the use of vasopressors to increase the driving pressure into the lungs (systolic BP) may be needed.
- **Full repair (Rastelli procedure, PA plasty).** RV systolic and diastolic dysfunction is common in the early postoperative stages. Medical strategies to support systolic function (milrinone and/or epinephrine infusions) and reduce afterload (achieving FRC, oxygen supplementation, and iNO) are needed. These patients also tend to benefit from lower heart rate and increased filling to manage their RV diastolic dysfunction. Manipulation of PVR and SVR for cardiopulmonary interactions and ventricular interdependence will be key the first 24–48 hours postoperatively. Most patients will remain sedated the first night postoperatively. LAP allows adequate fluid management over the first few days. Peritoneal dialysis should be started on the first postoperative night.