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CHAPTER 12.4

Pediatric Cardiovascular Surgery

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Surgery for Atrial Septal Defect (Ostium Secundum)

Surgical Considerations

Description: Atrial septal defects (ASDs) are among the most common congenital cardiac defects. ASDs vary widely in size and location and are broadly classified as **ostium secundum, ostium primum, sinus venosus,** and **coronary sinus** types (Fig. 12.4-1). Ostium secundum defects—the most common (80%)—result from an incompletely formed or fenestrated septum primum covering the fossa ovalis. A L→ R shunt results in augmented pulmonary blood flow which, if left uncorrected, may → RV failure, atrial arrhythmias, pulmonary HTN, and rarely, pulmonary vascular occlusive disease (PVOD). In 1953, Gibbon successfully repaired an ASD using a pump-oxygenator and, in doing so, ushered in the era of open cardiac surgery.

Secundum ASDs that fail to close spontaneously should be closed electively in early childhood to avoid long-term complications. Currently, ASD closure is performed through a minimally invasive midline partial sternotomy approach on CPB. A right anterolateral thoracotomy through the 4th intercostal space also provides satisfactory exposure and provides female patients with better cosmesis. After CPB and cardioplegic arrest or ventricular fibrillation have been instituted, a right atriotomy is created and the ASD is visualized.

Repair is affected by direct suture closure or patch closure, using autologous pericardium or prosthetic material (e.g., Gore-Tex, Dacron). After the repair is completed and the atriotomy is closed, standard de-airing maneuvers are performed, the aortic cross-clamp is released, and CPB is discontinued. The chest is then closed in the standard fashion.

Variant procedure or approaches: Currently, a **device closure** of ASD in the cath lab is a standard procedure. A **robotic approach** to ASD closure is undergoing clinical trials. Patent foramen ovale (PFO) in adults is becoming a common indication for device closure to avoid risk of paradoxical embolism and stroke during surgery.

Usual preop diagnosis: ASD; ostium secundum defect; other variants

Summary of Procedures

Position	Supine or right lateral decubitus (thoracotomy)
Incision	Minimally invasive midline incision with limited right median sternotomy; right anterolateral thoracotomy (4th intercostal space)
Unique considerations	R→ L shunt → systemic embolization
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp or ventricular fibrillation time: 10–30 min Total: 2 h
Closing considerations	Routine closure with chest tube in pericardial space; temporary atrial pacing wire (older patients); possible left atrial line for monitoring
EBL	Minimal
Postop care	Extubation in OR or within 1st 4 h; 24-h monitoring in ICU

Mortality	Rare
Morbidity	Hemorrhage Postpericardiotomy syndrome Atrial arrhythmias
Pain score	6–8

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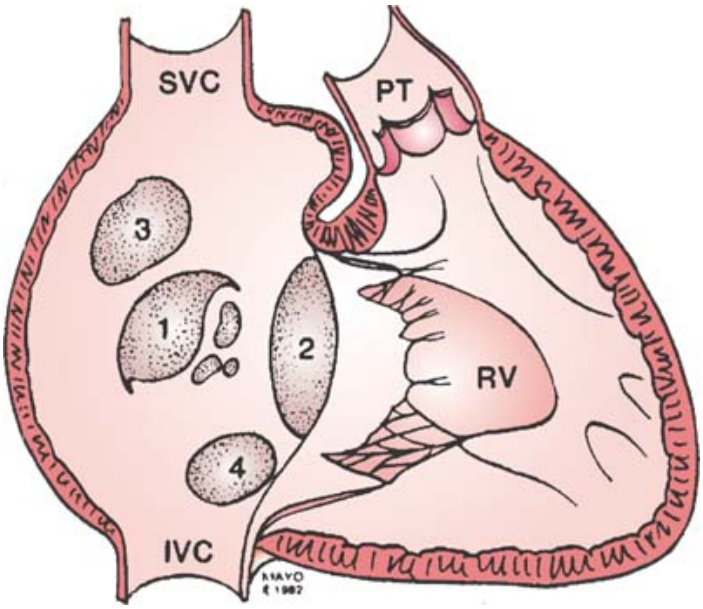

Figure 12.4-1. 1. Location of ASDs, numbered in decreasing order of frequency: 1 = secundum; 2 = primum; 3 = sinus venosus; 4 = coronary sinus type. IVC = inferior vena cava; PT = pulmonary trunk; RV = right ventricle; SVC = superior vena cava. (Reproduced with permission from the Mayo Foundation for Education and Research.)

Patient Population Characteristics

Age range	Neonate–adults
Male:Female	1:2
Incidence	10–15% of congenital heart defects
Etiology	Unknown; sometimes associated with syndromes such as Holt-Oram.
Associated conditions	ASD may coexist with nearly any of the other recognized congenital cardiac anomalies, but is more commonly associated with pulmonary stenosis (10%), partial anomalous pulmonary venous return (7%), VSD (5%), PDA (3%), and mitral stenosis (2%). ASDs are also part of other malformations, including tricuspid atresia, total anomalous pulmonary venous connection (TAPVC), and mitral atresia.

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Anesthetic Considerations

Preoperative

Although the secundum ASD is the most common, periop management varies little among all types of ASDs (including ostium primum, sinus venosus, and coronary sinus types). One

Pathophysiology	principle guideline in all L→R shunts is the estimation of shunt flow: $Q_p:Q_s$ This is determined by the amount of pulmonary blood flow (Q_p), size of the defect, and the PVR and SVR. Patients are generally asymptomatic until the $Q_p:Q_s$ ratio exceeds 3.0. As the shunt flow increases, Sx of pulmonary overcirculation may develop (e.g., failure to thrive, tachypnea at rest, and feeding difficulties). L→R shunt →↑ pulmonary blood flow (Q_p) →↑ PA pressures →↑ PVR → RV overload → RV failure. The larger the area of the defect, the greater the shunt.
Cardiovascular and Respiratory	Most infants with ASDs are asymptomatic. The ASDs are detected on cardiac auscultation (fixed splitting of S2). Young children may have h/o frequent URIs. Cyanosis is rare and indicates shunt reversal and development of pulmonary HTN. Surgical closure is advocated as these patients are at risk for paradoxical embolism, stroke, and bacterial endocarditis. Unlike ventricular septal defects (VSDs), ASDs rarely close spontaneously. Endovascular closure in the cath lab may be suitable for some patients, obviating the need for CPB. **Tests:** EKG: normal or RVH. CXR: cardiomegaly and ↑ pulmonary vascular markings. ECHO. TEE: essential for locating and estimating size of defect, other valvular abnormalities, and identification of pulmonary vein anatomy. Cardiac CT may be indicated for delineation of the pulmonary veins in sinus venosus ASD's because of the association of anomalous pulmonary venous return.
Laboratory	Hb/Hct; T&C
Premedication	Patients > 8 mo old may benefit from midazolam 0.5–0.7 mg/kg po 20 min before induction.

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Intraoperative

Anesthetic technique: GETA. Most of these patients are candidates for **fast-tracking**—the practice of extubation of cardiac patients in the OR or within 4 h of completion of surgery. To achieve this goal, the anesthetic technique must be adjusted to permit early extubation.

Criteria for fast-tracking include: patients > 4 mo of age, short CPB run, stable hemodynamic profile, minimal inotropic support, normal acid-base status, and adequate surgical hemostasis. Patients who are pacemaker-dependent are not suitable candidates for fast-tracking. Generally, patients selected for fast-tracking have undergone repair of a secundum ASD, VSD (no evidence of postop pulmonary HTN), bidirectional Glenn and Fontan procedures, or a RV → PA conduit change. For patients with a single ventricle, resumption of spontaneous ventilation decreases intrathoracic pressure and improves the transthoracic (PA-LA) pressure gradient. This increases venous return and improves CO.

Induction	Following inhalation induction using sevoflurane (O_2/N_2O), an iv is placed and intubation facilitated with rocuronium (1 mg/kg). After ET intubation, an arterial line is placed for close BP monitoring. **Fast-track:** Intravenous techniques are used for fast-tracking patients who are not candidates for regional anesthesia (e.g., parent preference, spine abnormality, age). Inhalation induction (sevoflurane ± N_2O) is followed by placement of a PIV and arterial line. Fentanyl (2–5 mcg/kg) and a muscle relaxant (e.g., rocuronium 1 mg/kg) are administered and the airway is secured. Then, either an infusion of remifentanil (0.1–0.4 mcg/kg) is started or single bolus iv methadone (0.2 mg/kg; max dose 20 mg) is administered. Propofol may also be used after separation from CPB and in transport to the ICU, where it is weaned and discontinued prior to extubation. **Non fast-track:** For patients who are not candidates for fast-tracking, the standard anesthetic is fentanyl 20–40 mcg/kg in divided doses with supplemental isoflurane before and during CPB. Additional midazolam (0.1–0.3 mg/kg) may be administered to

	prevent recall and awareness. Smaller children generally tolerate an inhalation induction with sevoflurane in 50% N_2O + O_2 After induction, iv access is established and rocuronium (1 mg/kg), followed by volatile anesthetic and supplemental iv fentanyl. An iv induction should be used in patients with significant pulmonary HTN and/or RV failure. IV induction is accomplished with etomidate 0.1–0.3 mg/kg or ketamine 0.5–1 mg/kg, followed by a muscle relaxant (e.g., vecuronium 0.1 mg/kg).	
Maintenance	In the maintenance of anesthesia, isoflurane may be used with iv techniques before, during, and after CPB. In patients in whom immediate postop extubation is planned, anesthesia is maintained using a volatile agent in air + O_2 N_2O is turned off after induction to avoid expansion of any intraop VAE. In patients with pulmonary HTN, RV failure and/or other medical conditions requiring postop mechanical ventilation, a high-dose narcotic technique may be appropriate (e.g., fentanyl 50–150 mcg/kg ± inhalation agent).	
Emergence	If the choice is made to extubate the patient receiving remifentanil in the OR, isoflurane is terminated during skin closure, muscle relaxant is reversed, and the remifentanil is turned off when the dressing is applied. When the patient meets standard extubation criteria (e.g., -25 cmH_2O NIF, spontaneous ventilation, adequate sat), remove the ETT and document spontaneous ventilation. If the patient is not extubated in the OR, transport the patient to the ICU sedated with remifentanil (0.1–0.4 mcg/kg/min) or low-dose propofol (25–75 mcg/kg/min), and extubate in the ICU. If the patient received methadone, standard criteria for extubation are used. Vasodilators are usually not required for BP control. O_2 bag, mask, airway equipment, and emergency drugs should be available during transport.	
Blood and fluid requirements	IV: appropriate for patient's size × 1–2 LR @ TKO Blood warmer for air bubbles. T&C 1 U PRBC.	**NB:** It is critical to avoid iv bubbles in all patients with intracardiac shunts. Meticulous attention must be paid to clearing the iv tubing and stopcocks of any bubbles to avoid paradoxical air embolism and possible stroke. Have blood available in the OR. Blood transfusion may be required in patients < 10 kg.
Monitoring	Standard monitors (see p. D-1). Cerebral oximetry T (two sites) Arterial line CVP line Urinary catheter ACT monitoring	ABG, electrolytes, and Hct should be checked as needed during the procedure. Place at least two O_2probes to ensure readings during critical times. Avoid dorsalis pedis and posterior tibial arterial lines (inaccurate 2° spasm post-CPB). Femoral arterial line may be used. Double lumen CVP in IJ preferable.
	TEE	TEE monitoring is used to assess anatomy and repair. On CPB, if the aorta is not cross-clamped, monitor for intra-cardiac air.
Positioning	and pad pressure points. eyes.	
Pre-CPB	Heparinization (200–400 U/kg) ACT > 480 sec. NMB. UO.	Heparin is added to the prime (1–3 U/mL). Confirm that antibiotics have been given 30–60 min before skin incision and repeat q 4–6 h per protocol.
CPB	Hypothermia adequate flow and pressure during CPB. Ventilation stopped face for venous congestion. ABG and ACT. UO. Blood should be available.	The patient is not actively cooled for secundum ASD closure. Temperature is allowed to drift to 33–34°C. Secundum ASD may be repaired using ventricular fibrillation instead of cardioplegia with aortic cross- clamping.

Transition off CPB	Rewarming Vasoactive infusions started at 32°C. Flush lines. Zero transducers. Suction ETT. Resume ventilation.	Surgeon will de-air heart, and this maneuver is monitored with TEE. Dopamine 3–5 mcg/kg/min. Observe inflation of both lungs.
Post-CPB	Reversal of heparin with protamine UO. ± Modified ultrafiltration (MUF)	1 mg of protamine will reverse 100 U of residual heparin (4 mg/kg may be needed in neonates). MUF uses the CPB machine to remove excess water and inflammatory mediators. It has been shown to transiently ↓ postop edema and improve cardiopulmonary function. This technique is not routinely performed at Stanford.
Complications	Air embolism (VAE) Supraventricular dysrhythmias Heart block Ventricular dysfunction	Potential for paradoxical embolization Likely 2° atriotomy

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Postoperative

Complications	Bleeding
Tests	ABG Electrolytes; Hct CXR

Suggested Readings

1. Allen HD, Driscoll DJ, Shaddy RE et al. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents*, 7th edition. Lippincott Williams & Wilkins, Philadelphia: 2007.

2. Bent ST: Anesthesia for left-to-right shunt lesions. In: *Anesthesia for Congenital Heart Disease*, 1st edition. Andropoulos S, Andropoulos R, eds. Blackwell Futura, CITY: 2005, 297–317.

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8. Torracca L, Ismeno G, Alfieri O: Totally endoscopic computer-enhanced atrial septal defect closure in six patients. *Ann Thorac Surg* 2001; 72(4):1354–7.

9. Wilson NJ, Smith J, Prommete B, et al: Transcatheter closure of secundum atrial septal defects with the amplatzer septal occluder in adults and children-follow-up closure rates, degree of mitral regurgitation and evolution of arrhythmias. *Heart Lung Circ* 2008; 17(4)318–24.

Surgery for Atrioventricular Canal Defect

Surgical Considerations

Description: Atrioventricular (A-V) canal defects comprise a spectrum of congenital cardiac anomalies stemming from the embryonic maldevelopment of endocardial cushions. This leads to the absence of septal tissue immediately above and below the level of the A-V valves and defects in the A-V valves in continuity with these septal defects. **Partial A-V canal defects** (or **ostium primum atrial septal defects**) involve the atrial septum and mitral valve, whereas **complete A-V canal defects** involve the atrial and ventricular septa and have a common atrioventricular valve. Intermediate or transitional A-V canal have varying degree of pathology between the above two common patterns. Pathophysiologically, these defects result in L → R shunting at the atrial and/or ventricular levels, leading to pulmonary HTN and CHF. A-V valvular insufficiency is also frequently observed with these defects, contributing to the early development of CHF.

Palliative repair of A-V canal defects consists of **pulmonary artery banding** (Fig. 12.4-2) to reduce excessive pulmonary blood flow. Palliation is rare and is reserved for very small infants with complicating conditions, such as RSV and other pneumonias. **Total correction** is now performed routinely, even in neonates. Repair consists of the closure of atrial and ventricular septal defects (VSDs) with closure of the cleft in the anterior leaflet of the mitral valve, and repair of associated defects, such as a patent ductus arteriosus (PDA) or secundum atrial septal defect (ASD). The septal defects may be repaired with either a **"two-patch technique,"** consisting of a Dacron or pericardial patch on the ventricular septum and pericardial patch on the atrial septum, or a single patch (pericardium) covering both ASDs and VSDs. A modified single patch technique where the A-V valve is plastered down to the ventricular septum is currently used by some surgeons.

Partial A-V canal defects: Exposure is obtained through a standard median sternotomy. CPB is instituted with aortic cross-clamping and cardioplegic arrest. Through a right atriotomy, the mitral valve cleft is sutured. A pericardial *(Print pagebreak 1207)* patch is placed across the top of the ventricular septum, around the coronary sinus orifice, and along the free edge of the superior portion of the ASD. The atriotomy is closed and the aorta unclamped after de-airing. The patient is rewarmed and weaned from bypass.

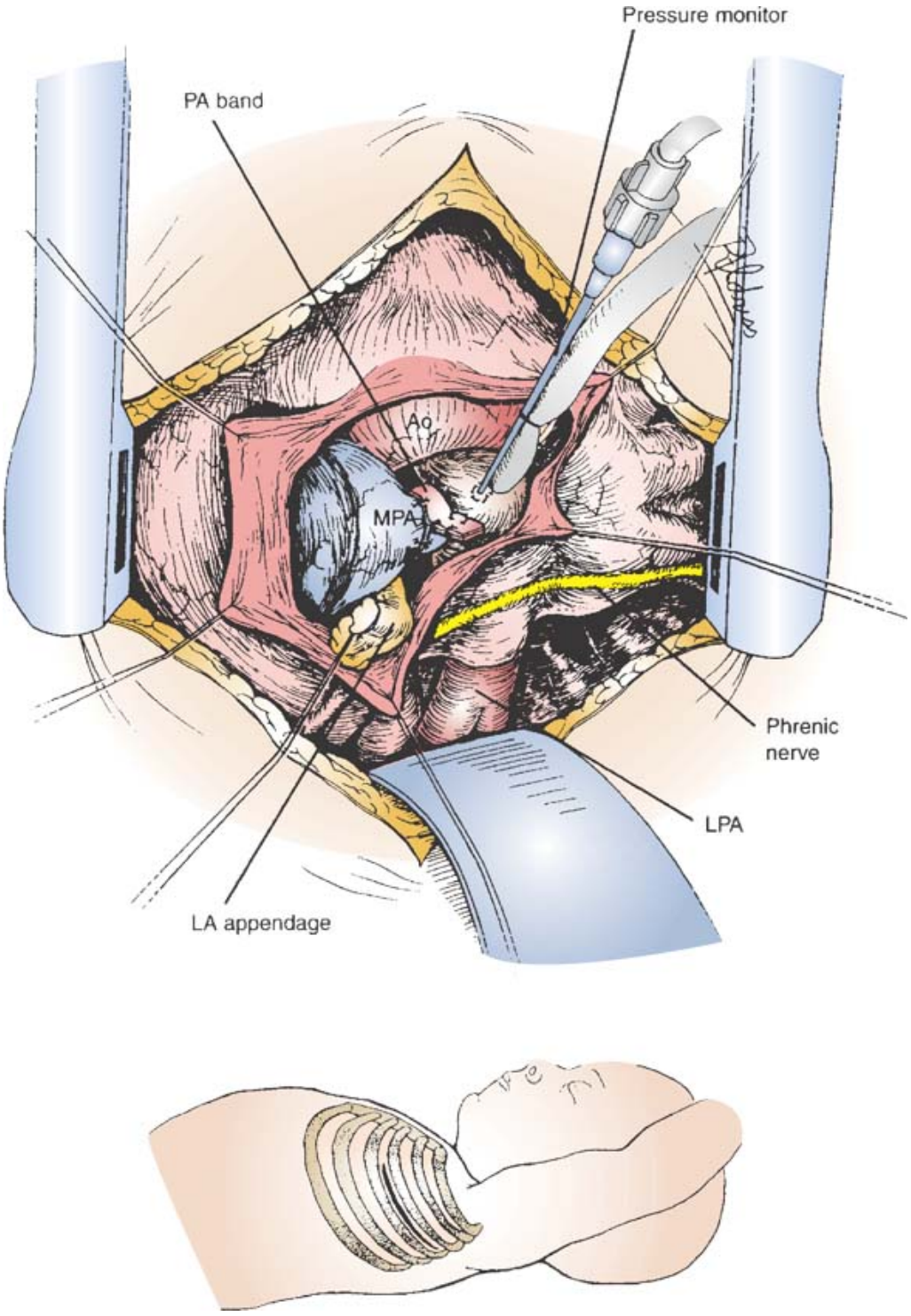

Figure 12.4-2. 2. Placement of pulmonary artery band. PA = pulmonary artery; Ao = aorta; MPA = main pulmonary artery; LPA = left pulmonary artery; LA = left atrial. (Reproduced with permission from Kaiser LR, Kron IL, Spray TL, eds: *Mastery of Cardiothoracic Surgery*. Lippincott-Raven, Philadelphia: 1998.)

Complete A-V canal defects: Following median sternotomy, CPB is instituted with aortic cross-clamping and cardioplegic arrest. In the **single-patch technique,** one patch is used to close both the ASD and VSD components. The anterior and posterior bridging leaflets often are divided and resuspended to the patch, thereby creating two separate valves. The cleft in the left AV valve is usually closed. If AV valve regurgitation is present, repair is undertaken. In the two-patch technique, two separate patches are used to close the ASD and VSD, and the common AV valve is sandwiched in between the two patches. Repair of AV valve is the same in either technique. Closure of the right atrium, de-airing, aortic unclamping, rewarming, and resuscitation of the heart are commenced, and CPB is discontinued. Closure proceeds in the standard fashion. In 5% of patients with complete A-V canal defects, tetralogy of Fallot (TOF) or left ventricular outflow tract obstruction (LVOTO) has to be addressed surgically.

Usual preop diagnosis: A-V canal defects (complete, intermediate, or partial); endocardial cushion defects

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Summary of Procedures

	Partial A-V Canal Defect	Complete A-V Canal Defect
Position	Supine	
Incision	Standard median sternotomy/right anterolateral thoracotomy	Standard median sternotomy
Unique considerations	None	Repair performed through a right atriotomy; no ventriculotomies required.
Antibiotics	Cefazolin 25 mg/kg q 4 h	
Surgical time	Aortic cross-clamp: 30–40 min Total: 1.5–2 h	Aortic cross-clamp: 60–80 min Total: 2.5–3 h
Closing considerations	Chest tube in pericardial space; temporary ventricular pacing wire; possibly right or left atrial line for monitoring	
EBL	Moderate	
Postop care	ICU for 1–2 d, with 12–24 h assisted ventilation; minimal need for inotropes.	1–3 d assisted ventilation; pulmonary HTN protocol, including hyperventilation and sedation; need for pulmonary vasodilators (e.g., NO) is uncommon; use of inotropes for adequate CO. Avoid excessive volume to prevent distension and mitral valve regurgitation.
Mortality	0–1%	1–2%
Morbidity	Transient heart block Atrial dysrhythmias Hemorrhage Infection	Pulmonary vasospasm Right heart dysfunction Mitral valve regurgitation Low CO Heart block/atrial dysrhythmias Residual VSD Hemorrhage
Pain score	6–10	6–8

Patient Population Characteristics

Age range	1–20 yr (usually 2–3 yr)	2–6 mo (occasionally older; rarely, adults)
Male:Female	1:1	
Incidence	0.5–1% of congenital heart defects	
Etiology	Down syndrome: Rare, in patients with partial A-V canal defect	Commonly associated with Down syndrome (75% in patients with complete A-V canal defect)
Associated conditions	Left A-V valve insufficiency	Pulmonary HTN, left A-V valve insufficiency, minor associated cardiac anomalies (e.g., PDA or ASD); major associated anomalies (e.g., TOF, LVOTO)

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Anesthetic Considerations

Preoperative

Pathophysiology	A-V canal defects may be either partial or complete. The partial A-V canal has no interventricular communication and has an ostium primum ASD. There is often a cleft in the anterior leaflet of the mitral valve → MR. The complete A-V canal has communication at both the arterial and ventricular levels with a single A-V valve that is regurgitant. Like all L → R shunt lesions, the degree of shunt depends on relative resistances in the systemic and pulmonary circuits. Patients with large L → R shunts are at risk of developing pulmonary HTN, requiring early surgical correction.
Respiratory	Look for pulmonary congestion or any infectious process. Pulmonary HTN can be present in patients with partial or complete A-V canal defects. **Tests:** CXR: enlarged heart + increased pulmonary markings
Cardiovascular	Complete A-V canal → CHF and biventricular failure. **Tests:** ECG: RAE, RVE; ECHO diagnostic; rarely cardiac cath may be indicated to assess PVR.
Down syndrome	A-V canal defects account for 40% of cardiac defects in the Down syndrome patient. These patients are at risk of atlantoaxial instability (20%); therefore, avoid excessive flexion-extension of the head and neck. Additional airway considerations (subglottic stenosis, large tongue, hypotonia) make these patients unsuitable for fast-tracking and early extubation.
Laboratory	See ASD, p. 1204.
Premedication	Midazolam 0.5–0.75 mg/kg po.

Intraoperative

Anesthetic technique: GETA

Induction	Typically, mask sevoflurane in air and O_2 If the iv is in place, ketamine 1–2 mg/kg or fentanyl 10 mcg/kg and rocuronium 1 mg/kg. Verify ETT position (see p. D-2). In children with Down syndrome, anticipate difficult airway and atlantoaxial instability. Avoid excessive neck flexion. In this population, The non-Down partial AV canal defects may be candidates for early or immediate extubation if the intracardiac defects are small and surgery uncomplicated.	
Maintenance	Air-oxygen with volatile anesthetic and narcotics is the standard. In patients requiring postop mechanical ventilation, fentanyl 50–100 mcg/kg total, rocuronium prn, and midazolam (0.1–0.2 mg/kg) ± volatile agent (FiO_2= 1) is used.	
Emergence	Patients undergoing repair of complete A-V canal defects are seldom suitable for fast-tracking. On conclusion of procedure, the patient is taken to PICU, intubated, ventilated, sedated, and monitored. Transport to ICU.	
Blood and fluid requirements	IV: 22–24 ga × 2 LR @ TKO Blood products See ASD, p. 1205.	See Table 12.4-2, p. 1226.
Monitoring	TEE	TEE is used to assess left A-V valve regurgitation, valvular function, residual shunt, and ventricular function.
CPB	Management of CPB is discussed in Intraoperative Considerations for TOF, p.	In the immediate post-CPB period, TEE exam is valuable for evaluation of ventricular function and residual mitral

	1205.	valve regurgitation.
	Air embolism	Avoid air embolism in A-V canal patients (shunt reversal → paradoxical embolization).
Complications	A-V block, ventricular dysfunction	A-V block may be temporary 2° edema and/or cardioplegic arrest. Permanent injury can occur to the conduction system at the A-V node or bundle of His with repair of the VSD. Temporary atrial and ventricular pacing wires are placed.
	Persistent bleeding Pulmonary HTN	See treatment for pulmonary HTN (see p. 430).

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Postoperative

Complications	Residual VSD/ASD AV valve regurgitation	
Tests	TEE	Analysis of post repair function is best done by TEE.

Suggested Readings

1. Allen HD, Driscoll DJ, Shaddy RE, et al., eds. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents*, 7th edition. Lippincott Williams & Wilkins, Philadelphia: 2007.

2. Bent ST: Anesthesia for left-to-right shunt lesions. In *Anesthesia for Congenital Heart Disease*, 1st edition. Andropoulos S, Andropoulos R, eds. Blackwell Futura, 2005, 297–317.

3. Castaneda AR, Jonas RA, Mayer JE Jr, et al: Atrioventricular canal defect. In *Cardiac Surgery of the Neonate and Infant*. WB Saunders, Philadelphia: 1994, 167–86.

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6. Vick WG, Titus JL: Defects of the atrial septum including the atrioventricular canal. In *The Science and Practice of Pediatric Cardiology*. Garson A, Bricker JT, McNamara DG, eds. Lea & Febiger, Philadelphia: 1990.

Surgery for Ventricular Septal Defect

Surgical Considerations

Description: Ventricular septal defect (VSD) is the most common congenital cardiac anomaly. VSDs can occur anywhere in the interventricular septum (Fig. 12.4-3). One of the most common locations is the perimembranous (conoventricular) in the region of membranous septum near the tricuspid and aortic valves. Supracristal (subarterial) defects are common in the Pacific Rim population. Muscular defects occur in the inlet, trabecular, or outlet muscular septum. A-V canal defects are present under the septal leaflet of the tricuspid valve. Physiologically, these defects result in L→ R shunting in proportion to the defect size. Untreated, this defect can → RV volume overload and CHF in infancy and irreversible pulmonary HTN later in life. As PVR rises, shunt reversal to a R→ L shunt can occur, producing hypoxemia and cyanosis; this is known as Eisenmenger's syndrome and occurs in 10% of

untreated, nonrestrictive VSDs (rare). Moderate-to-large VSDs that remain open > 6 mo of age should be closed. Severe, intractable CHF in infants refractory to medical therapy (e.g., diuretics, digoxin), or → PVR in infants > 6 mo are indications for earlier VSD repair.

Lillehei, et al performed the first successful VSD repairs using normothermic cross-circulation in 1955. Kirklin subsequently described successful VSD closure using extra-corporeal circulation in 1957. VSD repair is performed now through a median sternotomy on CPB with bicaval cannulation. Deep hypothermia (18°C) with circulatory arrest is used in neonates < 1800 g to facilitate repair. After CPB and cardioplegic arrest have been instituted, a right atriotomy is created and the VSD is visualized by retracting the tricuspid valve leaflets. The VSD is then closed with a *(Print pagebreak 1211)* patch (e.g., pericardium, Gore-Tex, Dacron), with care being taken not to place sutures through the nearby conduction fiber bundles or the aortic valve. After the repair is completed and the atriotomy is closed, standard deairing maneuvers are performed, the aortic cross-clamp is released, and CPB is discontinued. The chest is then closed in the standard fashion.

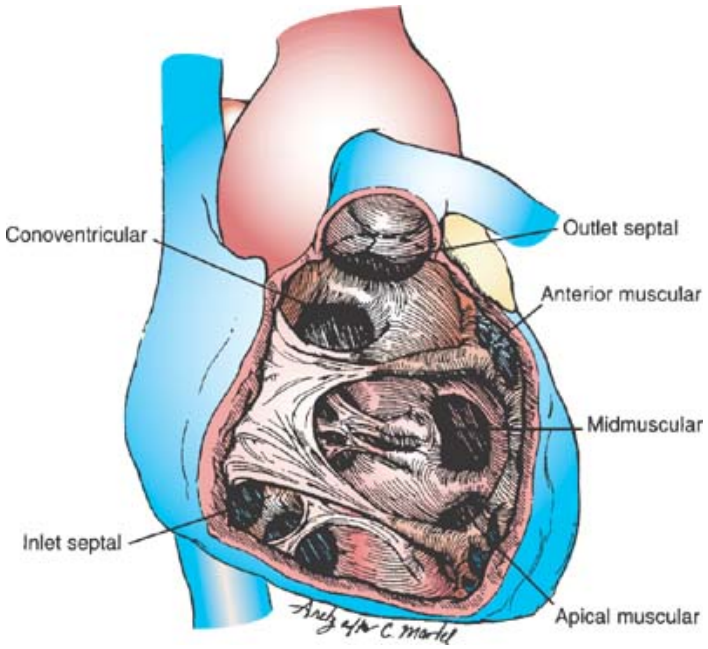

Figure 12.4-3. 3. The right ventricular free wall has been resected to show the VSDs: conoventricular = perimembranous; conal septal = outlet septal (subpulmonary); inlet septal = A-V canal type. Muscular (trabecular) defects may be midmuscular, anterior, or apical. The penetrating bundle is closely related to the inferior margin of the conoventricular defect and diverges away from this margin into the trabecular septomarginalis beneath the muscle of Lancisi. (Reproduced with permission from Kaiser LR, Kron IL, Spray TL, eds: *Mastery of Cardiothoracic Surgery*. Lippincott-Raven, Philadelphia: 1998.)

Variant procedure or approaches: Supracristal defects may be more easily exposed through a pulmonary arteriotomy, while some inferiorly located muscular VSDs may be better accessed through a right ventriculotomy. Multiple VSDs or "Swiss cheese" ventricular septum can be a challenging surgical problem. Many can be closed with the approaches described above. Some may require a combined surgical and device closure. Palliative pulmonary artery banding may be required in Swiss cheese muscular VSD, with the hope that many of the VSDs may close spontaneously. At a 2nd stage, the remaining VSDs are closed and the PA band is removed. Currently, device closure of muscular VSDs is experimental in the USA but is being used often in other countries.

Usual preop diagnosis: VSD

Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp: 15–45 min Total: 2–3 h

Closing considerations	Routine closure with chest tube in the pericardial space; temporary ventricular pacing wire; possibly right/left atrial and/or pulmonary arterial lines for monitoring
EBL	Minimal-to-moderate
Postop care	Extubation in OR or within several h; 24 h monitoring in ICU.
Mortality	≥ 1%
Morbidity	Hemorrhage Low CO Atrial dysrhythmias Complete A-V heart block Residual shunting; tricuspid regurgitation
Pain score	6–8

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Patient Population Characteristics

Age range	Neonate – adult
Male:Female	1:1
Incidence	20% of congenital heart defects; 2/1000 births
Associated conditions	PDA (6%); coarctation of the aorta (5%); congenital AS (2%); congenital mitral valve disease (2%); also part of other malformations, including TOF, TGA, DORV, tricuspid atresia; truncus arteriosus; others

Anesthetic Considerations

Preoperative

Pathophysiology	VSDs are classified by their location and degree of shunt flow. Large VSDs offer little resistance to flow and are nonrestrictive (RV = LV pressure). Small VSDs (restrictive) may close spontaneously in early childhood. Patients with large VSDs present with CHF and other signs of pulmonary over-circulation ($Q_p:Q_s > 3.0$). Cyanosis indicates pulmonary HTN and shunt reversal (Eisenmenger's syndrome).
Cardiovascular and respiratory	Infants may present with feeding difficulty and failure to thrive (FTT). Children with small VSDs may be asymptomatic, while children with larger VSDs may have ↓ exercise tolerance, fatigue, frequent URIs and Sx of CHF. A holosystolic murmur can be heard best at the left lower sternal border. Cyanosis is absent unless there is R → L shunting (suggesting pulmonary HTN). **Tests:** Obtain EKG, CXR, ECHO. If pulmonary HTN is suspected, cardiac catheterization is performed for measurement of PA pressures. If PA pressures are elevated, O_2 and NO responsiveness is tested. VSDs may be closed surgically or by a device in the cath lab if pulmonary HTN is not present.
Laboratory	Hct; electrolytes (children on diuretic Rx); others as indicated from H&P.
Premedication	See ASD (see p. 1204).

Intraoperative

Anesthetic technique: The anesthetic management of a patient with VSD (without pulmonary HTN and CHF) is similar to the patient with ASD. These patients may be candidates for fast-tracking and early extubation. Refer to fast-tracking in ASD section (see p. 1204). For patients with unrestrictive VSDs, avoid factors that ↓ PVR and ↑ shunt *(Print pagebreak 1213)* flow. This will compromise DBP and coronary perfusion. For patients who are not candidates for fast-tracking, the standard anesthetic is fentanyl 20–40 mcg/kg in divided doses with supplemental isoflurane before and during CPB. Additional midazolam (0.1–0.3 mg/kg) may be administered to prevent recall and awareness.

Blood and fluid requirements	IV: appropriate for patient size × 1–2 LR @ TKO Blood warmer T&C	Minimize administration of iv fluids. Both ventricles are volume-overloaded in large VSDs.
Monitoring	Standard monitors (see p. D-1). Arterial line Surgical LA line Urinary catheter ACT monitoring	A transthoracic LA line may be placed by the surgeon if there is concern about postop mitral valve and LV dysfunction.
	TEE	In the immediate post-CPB period, TEE exam is valuable for evaluation of residual VSD and ventricular function.
Pre-CPB	See ASD (see p. 1205).	
CPB	See ASD (see p. 1205).	Most VSDs are closed under mild/moderate hypothermic CPB (28–32°C).
Transition off CPB	See ASD (see p. 1205).	
Post-CPB	Maintenance of adequate filling pressure (LA = 5–10 mmHg)	
	Temporary pacemaker wires	Placed in patients with conduction disturbances; pacemaker wires are placed routinely, but may not be connected to a pacemaker.
	TEE Pulmonary HTN	for residual shunt and ventricular and valvular function.

Postoperative

Complications	Persistent bleeding Heart block	May occur particularly after perimembranous VSD closure
	Ventricular dysfunction/failure	Especially in patients with ventriculotomy Dx: TEE
Tests	ABGs Electrolytes; Hct; coags prn CXR	

Suggested Readings

1. Bent ST: Anesthesia for left-to-right shunt lesions. In *Anesthesia for Congenital Heart Disease*, 1st edition. Andropoulos S, Andropoulos R, eds. Blackwell Futura, 2005, 297–317.

2. Castaneda AR, Jonas RA, Mayer JE Jr, et al: Ventricular septal defect. In *Cardiac Surgery of the Neonate and Infant*. WB Saunders, Philadelphia: 1994, 187–201.

3. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Ventricular septal defects. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Baltimore: 1998, 264–85.

4. Knott-Craig CJ: Ventricular septal defects. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 687–96.

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(Print pagebreak 1214)

Surgery for Patent Ductus Arteriosus

Surgical Considerations

Description: Patent ductus arteriosus (PDA) usually is located between the proximal descending thoracic aorta and the main PA. Pathophysiologically, this results in L → R shunting and augmented pulmonary blood flow, which, if left untreated, may lead to pulmonary HTN and CHF. A relatively common congenital heart anomaly, comprising 12–15% of CHDs, PDA was first successfully ligated by Gross in 1938. Early administration of indomethacin may promote ductal closure in many premature infants, obviating surgical intervention; however, this mode of therapy generally is contraindicated in the setting of renal insufficiency or intracranial bleeding.

Surgical ductal closure is indicated for significant L → R shunting. The ductus usually can be exposed via a small, left, posterolateral thoracotomy in the 4th intercostal space or via the thoracoscopic approach. The ductus is identified and dissected with special care taken to avoid injury to the phrenic and left recurrent laryngeal nerves (Fig. 12.4-4). The ductus is interrupted with a surgical clip in neonates; in older children, the ductus is double- or triple-ligated or divided between vascular clamps, and the ends are oversewn. A small thoracostomy tube is placed, and the thoracotomy is closed. The thoracostomy tube is removed in the OR immediately after the chest is closed or a few hours later.

Variant procedure or approaches: Percutaneous coil embolization and **thoracoscopic clip ligation** are standard alternative approaches. A **robotic approach** is experimental.

Usual preop diagnosis: PDA

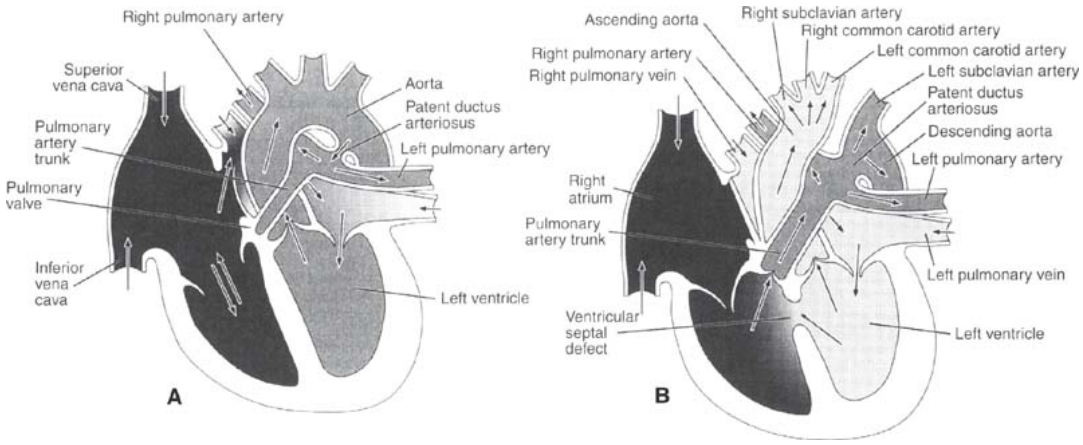

Figure 12.4-4. 4. PDA. **(A)** Ductal dependency for pulmonary blood flow in pulmonary valvular atresia. The ductus arteriosus must be open for blood to enter the pulmonary arteries; as the ductus arteriosus closes, pulmonary blood flow is lost, and the patient becomes cyanotic. **(B)** Dependence on the ductus arteriosus for perfusion of the distal aorta is shown in a patient with interrupted aortic arch. Left ventricular blood (oxygenated) is able to cross the VSD and enter the PA, where it mixes with right ventricular blood. The flow is then distributed to the branch PAs and across the ductus arteriosus to the descending aorta. (Reproduced with permission from Ungerleider RM, Plunkett MD, Gaynor JW: Congenital heart disease. In *Surgery of Infants and Children*
. Oldham KT, Colombani PM, Foglia RP, eds. Lippincott-Raven, Philadelphia: 1997).

(Print pagebreak 1215)

Summary of Procedures

Position	Right lateral decubitus

Incision	Small left posterolateral thoracotomy; 3rd or 4th intercostal space
Antibiotics	Cefazolin 25 mg q 4 h
Surgical time	30–60 min
Closing considerations	Routine closure ± left thoracostomy tube
EBL	Minimal
Postop care	24 h observation in ICU; extubation in the OR or within several h
Mortality	Rare
Morbidity	Hemorrhage Recurrent laryngeal nerve injury Chylothorax
Pain score	6–8

Patient Population Characteristics

Age range	1 mo–adulthood
Male:Female	1:2
Incidence	12–15% of congenital heart defects
Etiology	Failure of complete ductal closure

Anesthetic Considerations

Preoperative

Closure of PDA in the preterm infant is done at the bedside in the NICU. These patients are intubated, mechanically ventilated, hemodynamically unstable, and may require inotropic support. The premature infant requiring surgical closure of PDA typically presents because indomethacin Rx has failed or is contraindicated. They often have primary pulmonary disease and multisystem organ problems. In these infants, the symptoms of large L → R shunt and pulmonary overcirculation → cardiac and respiratory failure and ventilator dependence. The ductal runoff causes ↓ DBP and compromises coronary blood flow and other organ perfusion. Older patients may be eligible for endovascular closure in the cath lab, leaving a small percentage for open surgical closure in the OR. Surgical closure may be performed with video-assisted thoracoscopy (VAT) or via thoracotomy. The following discussion for anesthetic management covers open thoracotomy only. In older asymptomatic patients, risk of bacterial endocarditis necessitates closure.

Cardiovascular	Clinical presentation depends on the size of the ductus. An infant with a large ductus can present in CHF with tachypnea, tachycardia, diaphoresis, failure to thrive (FTT), and hepatosplenomegaly. A continuous systolic murmur heard best at the left upper sternal border and widened pulse pressure with bounding pulses may be present. **Tests:** CXR: Normal or cardiomegaly with ↑ PA size and ↑ pulmonary vascular markings. EKG: Normal or LVH/RVH. ECHO diagnostic: documents patency, evaluates left-sided cardiac chamber sizes and aortic arch anatomy, and estimates size of shunt.
Laboratory	Hct; others as indicated from H&P.
Premedication	Not indicated in premature infants. Midazolam 0.5–0.7 mg/kg po 20 min prior to induction for children > 9 mo having routine PDA closure.

(Print pagebreak 1216)

Intraoperative

Anesthetic technique: Most children presenting for elective closure of PDA via thoracotomy are candidates for thoracic epidural analgesia, while for those undergoing thoracoscopic closure, an epidural is not required. In patients > 25 kg, placement of a DLT for selective ventilation of the right lung will improve surgical access. In general these patients are candidates for early extubation in the OR (see fast-track in ASD, p. 1204).

Induction	At the bedside of the preterm infant, anesthesia can be provided with ketamine (1–2 mg/kg) and muscle relaxant (e.g., rocuronium 1 mg/kg iv). Small doses of fentanyl (2–5 mcg/kg) may be given for supplemental analgesia. During lateral decubitus positioning, careful attention to the airway and close monitoring of BP is necessary. Children presenting in the OR for elective operation tolerate an inhalation induction with sevoflurane in 50% N_2O + O_2 Once the patient is anesthetized, iv access is established, followed by oral intubation, with administration of a muscle relaxant (e.g., rocuronium 1 mg/kg).	
Maintenance	In patients for whom immediate postop extubation is planned, anesthesia is maintained with isoflurane in air + O_2	
Emergence	Patients are extubated awake. Post-thoracotomy, an effective regional anesthetic is helpful for fast-tracking.	
Epidural	Thoracic level T8-10 preferred. For neonates < 10 kg a caudal catneter can be advanced to thoracic levels. Initial bolus of bupivacaine (0.25%, 0.3 mL/kg) with hydromorphone (5–10 mcg/kg) ± clonidine (0.5–1 mcg/kg). Infuse bupivacaine (1/8%) at 0.3–0.5 mL/h during surgery. Check coagulation parameters prior to placing epidural. If < 10 kg, caudal approach can be used.	
Blood and fluid requirements	IV: 22–24 × 1–2 Continue iv dextrose. Blood warmer T&C for air bubbles.	Blood loss may become significant if the ductus is torn during ligation. Have blood available for rapid transfusion. Clear air bubbles from iv tubing and stopcocks (potential bidirectional shunt → paradoxical embolism).
Monitoring	Standard monitors (see p. D-1). ± Arterial line ± CVP line	In preterm infants monitoring of the preductal BP (right arm) and a postductal saturation monitor (usually lower extremity) are required because of possible inadvertent ligation of the descending aorta. Place BP cuff on preductal (right arm) and pulse oximeter on the foot to monitor flow in the ascending and descending aorta. Inadvertent aortic occlusion →↓ LE pulses/pressure/perfusion. Inadvertent PA occlusion →↓ SpO_2+ ↓ $ETCO_2$ Bradycardia may occur during manipulation of the ductus. The DBP will → postligation.
Positioning	and pad pressure points. eyes.	
Complications	Occlusion of aorta or PA Torn ductus → hemorrhage Residual PDA Lung trauma ± pneumothorax	

Postoperative

Complications	Recurrent laryngeal nerve injury Vagus nerve injury

Tests	Hct CXR	Other tests as indicated.

(Print pagebreak 1217)

Suggested Readings

1. Bent ST: Anesthesia for left-to-right shunt lesions. In *Anesthesia for Congenital Heart Disease*, 1st edition. Andropoulos S, Andropoulos R, eds. Blackwell Futura, 2005, 297–317.

2. Burke RP: Patent ductus arteriosus. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 657–62.

3. Castaneda AR, Jonas RA, Mayer JE Jr, et al: Patent ductus arteriosus. In *Cardiac Surgery of the Neonate and Infant*. WB Saunders, Philadelphia: 1994, 203–13.

4. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Patent ductus arteriosus. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Baltimore: 1998, 286–308.

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Surgery for Coarctation of the Aorta

Surgical Considerations

Description: Coarctation of the aorta is a congenital narrowing of the upper descending aorta, typically located at the juxtaductal region (Fig. 12.4-5). Coexisting intracardiac defects are not uncommon. Surgical repair of aortic coarctation, first performed by **Crafoord** in 1944, consisted of resection of the narrowed aortic segment, followed by an **end-to-end repair.** The same year, **Blalock** and **Park** proposed an alternative technique in which the left subclavian artery was divided distally and sutured into the descending thoracic aorta, creating a bypass. Subsequently, the use of an onlay prosthetic graft to widen the area of coarctation and the use of a **subclavian artery flap** were described by **Waldhausen. Prosthetic interposition tube graft repairs** have been described in patients with diffuse aortic hypoplasia. Patients with an associated intracardiac L → R shunt (e.g., VSD) may require concomitant pulmonary artery banding following repair of the coarctation.

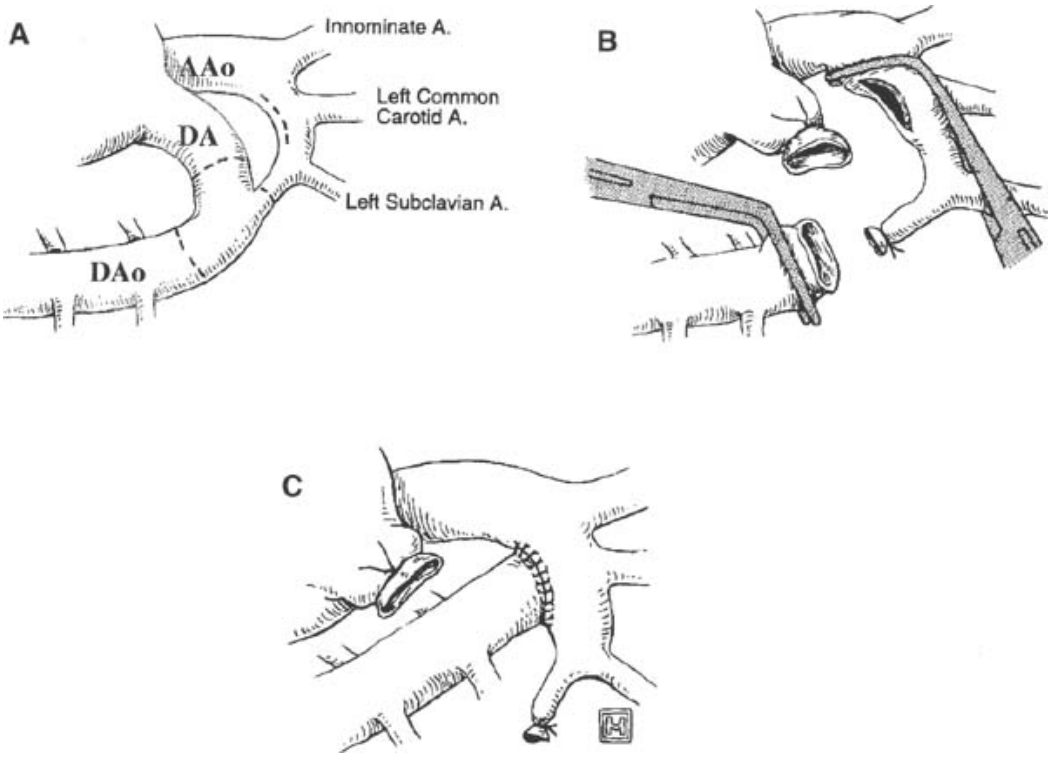

Figure 12.4-5. 5. Redrawn with permission from Younoszai AK, Reddy VM, Hanley FL, et al: Intermediate term follow-up of the end-to-side aortic anastomosis for coarctation of the aorta. *Ann Thorac Surg* 2002; 74(5):1631–4.

(Print pagebreak 1218)

In infants, a left posterolateral thoracotomy approach is used. The lung is retracted anteriorly and the pleura are incised vertically over the aorta, along the left subclavian artery and the descending aorta. The aortic arch, all the arch branches, and the descending aorta are thoroughly mobilized. The PDA/ligamentum arteriosus is ligated and divided. Vascular clamps are placed across the aortic arch and the descending aorta. The aortic arch clamp also partially occludes the distal ascending aorta. The aortic isthmus is ligated. The coarctated segment and all ductal tissue from the descending aorta are excised. An aortotomy is made in the aortic arch (may extend on to the distal ascending aorta) and the descending aorta is anastomosed to the aortic arch. Rarely if a subclavian flap angioplasty is performed, the distal left subclavian artery is ligated and opened longitudinally down into the aorta, across the coarctation and into the descending aorta. The subclavian flap is then turned down and anastomosed to the descending aorta across the coarctated segment. Patch aortoplasty is performed by creating a longitudinal aortotomy above and below the coarctation and suturing a generously sized patch onto the defect. Following repair, the aortic cross-clamps are released, and hemostasis is secured. A pleural drainage tube is placed and the pleura are sutured over the aorta, followed by standard closure.

Variant procedure or approaches: In adults and older children (teens), **balloon dilatation with stent placement** is an acceptable alternative.

Usual preop diagnosis: Coarctation of the aorta

Summary of Procedures

	Infant	Child or Adult
Position	Right lateral decubitus	
Incision	Posterolateral left thoracotomy, 3rd-4th interspace	
Unique considerations	Right upper limb arterial line. Avoid iv or arterial lines in left arm. Mild hypothermia and surface cooling (35°C).	Collateral circulation is adequate if distal aortic MAP > 50 mmHg. In rare cases of inadequate collateral circulation, left atrial-to-descending-aorta or femoral bypass should be considered. Mild hypothermia of 35° may be useful.

Antibiotics	Cefazolin 25 mg/kg q 4 h	
Surgical time	Aortic cross-clamp: 12–20 min Total: 1–1.5 h	Aortic cross-clamp: 10–15 min Total: 2–2.5 h
Closing considerations	Thoracostomy tube	
EBL	Minimal	1–2 mL/kg
Postop care	Early extubation in ICU; consider intercostal nerve block or epidural analgesia; careful control of MAP may require SNP or esmolol infusion.	
Mortality	< 1%	
Morbidity	Chylothorax Bleeding Infection Paraplegia: < 0.5%	
Pain score	8–10	8–10

Patient Population Characteristics

	Infant	Child or Adult
Age range	1 wk–1 yr	1 yr–adult
Male:Female	2:1; 1:1 for coarctations with associated defects	
Incidence	5–8% of congenital heart defects	5% of congenital heart defects
Etiology	Unknown. Ductal sling may be the single most contributory factor.	
Associated conditions	PDA (50%); VSD (36%); congenital mitral stenosis (7%); single ventricle (7%); TGA/VSD (7%)	Usually an isolated defect. When repaired at a later age, bicuspid aortic valve.

(Print pagebreak 1219)

Anesthetic Considerations

Preoperative

Pathophysiology	Coarctation is a narrowing of the aortic arch, most commonly occurring at the junction of the ductus arteriosus. The clinical presentation of the neonate or infant with coarctation depends on: (a) location and degree of obstruction, (b) rate at which high-grade stenosis develops, (c) patency of the ductus, (d) PVR, and (e) associated cardiac anomalies. Neonates with critical stenosis will demonstrate ductal-dependent aortic flow. With the ductus open and ↑ PVR, the neonate may be asymptomatic. R↑L ductal flow, enhanced by ↑ PVR, may → palpable lower extremity pulses, obscuring the Dx. As the ductus closes (after 4–10 d), lower torso hypoperfusion will become significant (acidemia, gut ischemia, cold lower extremities). Neonates may present in shock and require prompt resuscitation and surgery. Patients with mild-to-moderate stenosis and no associated abnormalities will be asymptomatic with upper extremity HTN and diminished lower extremity pulses.
Respiratory	**Tests:** CXR (cardiomegaly, characteristic aorta, PE) **Neonates:** Neonates present with “critical” coarctation and are

Cardiovascular	ductal-dependent. If the ductus is closing, PGE_1 may reopen the duct—or at least relax juxtaductal tissue—and improve perfusion. PGE_1 treatment may allow time for ECHO assessment of associated lesions. **Infants:** Present with upper extremity hypertension and murmur. Imaging studies such as MRI or CT scan may be needed to confirm diagnosis if gradient is not well detected by echo. **Older children:** Often present with HTN, HA, and diminished or absent femoral pulses. The indication for operation is similar to that of infants. Systemic HTN and LVH may not resolve following repair. **Tests:** ECHO; ECG; ABG; CXR—rib notching 2° collateral vessels. (This is a late finding.) Recently cardiac MRI or CT angio of the chest are used to diagnose coarctations and other associated anomalies.
Renal	**Tests:** BUN; Cr; electrolytes
CNS	PGE_1 is associated with apnea in the newborn. Generally, the neonate is intubated once the prostaglandin infusion is started.
Laboratory	Hct; T&C
Premedication	Not appropriate in severely ill infants. Otherwise, midazolam 0.5–0.7 mg/kg po (maximum of 20 mg). For children > 25 kg, lorazepam 1–2 mg po 2 h before surgery.

(Print pagebreak 1220)

Intraoperative

Anesthetic technique: GETA. For the neonate, fast-tracking is not appropriate.

Induction	In neonates, an iv induction with fentanyl 2–5 mcg/kg and rocuronium is appropriate. Avoid ↓ SVR and myocardial depression, while maintaining a normal HR. Inhalation induction (sevoflurane and O_2) is well tolerated in the older patients. In most infants and children, single-lumen ETT is sufficient. In children selective ventilation of the right lung with a DLT will facilitate surgery.
Maintenance	A narcotic technique (10–20 mcg/kg fentanyl ± volatile agent) is appropriate for critically ill neonates. Consider mild hypothermia (34–35°C) for CNS protection. The operation usually is via a left thoracotomy. The aortic arch, isthmus, and descending aorta are mobilized. 100 U/kg of heparin may be given. The ductus arteriosus is ligated, then the aortic arch and descending aorta are occluded. The anesthesiologist should observe the arterial line trace closely during proximal occlusion and immediately inform the surgeon of any damping. Upper and lower extremity cuff pressures are measured. When early postop extubation is planned, see fast-tracking (see p. 1204).
Emergence	Infants and children undergoing elective operation can usually be extubated in the OR. In young infants, systolic pressure consistently > 120 mmHg should be treated and an esmolol and or SNP infusion may have to be started in the OR. In neonates or infants with compromised cardiac function, extubation should be delayed until their clinical status has improved. Dopamine or milrinone may be required. A chest tube is left in place to monitor blood loss or the appearance of a chylous effusion.
Blood and fluid requirements	IV: appropriate for patient size × 1–2 LR @ TKO (D5 LR in neonates) Blood warmer T&C 1–2 U. Standard monitors (see p. D-1).
	As a rule, infants and children undergoing coarctation repair will not require transfusion. Infrequently, however, sudden and substantial blood loss can occur, necessitating rapid transfusion. Adequate iv access and immediate availability of blood is essential in this operation. BP must be monitored above and below the level of coarctation. A right radial arterial

	RUE arterial line ± CVP	line allows BP monitoring during occlusion of the aortic arch. If a percutaneous radial arterial line cannot be placed, a surgical cutdown should be performed or a right axillary arterial line should be placed.
Monitoring		
	BP cuffs × 2 (UE + LE)	Both upper and lower extremity cuff pressures should be monitored.
	Temperature	Monitor rectal temperature and maintain it below 350C
	NIRS	Both cerebral and somatic
		ABG, glucose, Hct at intervals.
Control of BP	Anesthetic depth	In neonates and infants, GA is usually sufficient to control BP during aortic cross-clamp; however, SNP may be necessary to control BP in some patients.
Aortic cross- clamping	± Heparin 100 U/kg Ductus arteriosus ligated Aorta cross-clamped × 2 arterial line for damping. Ischemia time = 10–20 min Maintain distal perfusion. Lactic acidosis →↓ SVR + ↓BP.	Aortic cross-clamping presents an acute afterload to the LV. In preparation for release of cross-clamp, decrease or eliminate volatile anesthetic. Have blood and other volume available. $NaHCO_3$ is frequently required (1–2 mEq/kg). ABG and treat accordingly.
Complications	Spinal cord ischemia HTN Bleeding Hypothermia Residual coarctation	

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Postoperative

Complications	Paraplegia	Incidence = 0.14–0.4%. The cause is likely 2° spinal cord ischemia.
	HTN Bleeding GI bleed Abdominal pain and ileus Recurrent laryngeal nerve injury Chylous effusion Residual or recurrent coarctation	Occurs infrequently. Attributable to mesenteric arteritis. Neonates are at risk for GI reperfusion injury following repair. These patients are kept npo for 12–24 h.
Tests	CXR Hct ABGs	

Suggested Readings

1. Allen HD, Clark EB, Gutgesell HP, et al, eds. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents*, 6th edition. Lippincott Williams & Wilkins, Philadelphia: 2001.

2. Castaneda AR, Jonas RA, Mayer JE Jr, et al: Aortic coarctation. In *Cardiac Surgery of the Neonate and Infant*. WB Saunders, Philadelphia: 1994, 333–52.

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Surgery for Tetralogy of Fallot

Surgical Considerations

Description: In tetralogy of Fallot (TOF), the RV infundibulum is maldeveloped, resulting in RV outflow tract obstruction (RVOTO), a large malalignment (overriding aorta) VSD, RVH (see Fig. 12.4-6) and, occasionally, an ASD (**pentalogy of Fallot**). TOF is the most common cyanotic congenital cardiac defect. Pathophysiologically, RVOTO leads to significant R → L shunting across a nonrestrictive VSD, which, in turn, leads to inadequate pulmonary blood flow and varying degrees of cyanosis. The variation in presentation depends primarily on the severity and type of RVOTO.

Patients with TOF were first treated palliatively beginning in 1944, with the introduction of the **Blalock-Taussig** (B-T) **systemic-to-pulmonary artery shunt.** This procedure augments pulmonary blood flow and, hence, systemic oxygenation. Currently, however, most cases of TOF are treated routinely with early complete correction between 3–12 mo of age. Neonatal repair is performed in infants who are severely cyanotic (SaO_2 < 80%), have ductal-dependent pulmonary blood flow, or have cyanotic spells. Initial palliation with the B-T procedure is now reserved *(Print pagebreak 1222)* for patients with severe pulmonary arterial hypoplasia and (by some surgeons) for an anomalous left anterior descending coronary artery originating from the right coronary artery.

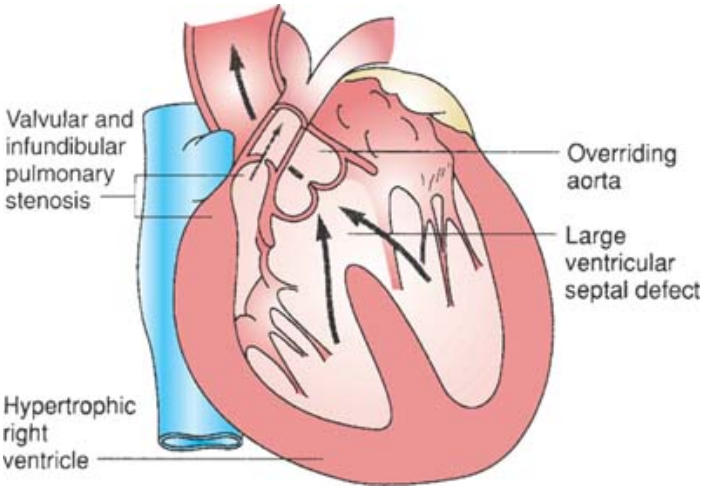

Figure 12.4-6. 6. Anatomic features of tetralogy of Fallot. The primary morphologic abnormality, anterior and superior displacement of the infundibular septum, results in malalignment VSD, overriding the aortic valve and obstructing RV outflow. RV hypertrophy is a secondary occurrence. (Reproduced with permission from Greenfield LJ, Mulholland MW, Oldham KT, et al, eds: *Surgery: Scientific Principles and Practice,* 2nd edition. Lippincott-Raven Publishers, Philadelphia: 1997.)

Surgical correction is performed through a standard median sternotomy on standard CPB with moderate hypothermia. During cooling, the modified B-T shunt, if present, is ligated and divided, followed by aortic cross-clamping, cardioplegic arrest, and topical cooling. The pulmonary valve and RVOT are accessed through a longitudinal pulmonary arteriotomy and the tricuspid valve (via a right atriotomy), respectively. RVOTO is treated as necessary by infundibular muscle resection, pulmonary valvotomy or valvectomy, and patch augmentation of the main PA or its branches. Autologous pericardium, Gore-Tex, or other type of

synthetic/biologic material may be used for patch widening the RVOT at any level, from the RV to the PAs. The primary goal in relieving the RVOTO is to save the pulmonary valve, avoid a trans-cannular patch, and minimize pulmonary valve insufficiency. The VSD, accessed through the tricuspid valve, is closed with a patch (pericardial/Dacron) in the standard fashion with care being taken to avoid injury to the bundle of His.

Variant procedure or approaches: In patients with severe hypoplasia or atresia of the RVOT, or in patients with an anomalous origin of the LAD coronary artery from the right coronary artery, a **Rastelli procedure** is performed. This operation consists of patch closure of the VSD and reconstruction of the RVOT in which a conduit, in the form of a cryopreserved homograft or valved prosthesis, is placed between the RV and the PA.

Usual preop diagnosis: TOF; cyanotic CHD; severe cyanotic spells; failure to thrive (FTT)

Summary of Procedures

	Neonate Repair	Infant and Adult Repair With Standard CPB
Position	Supine	
Incision	Standard median sternotomy	
Unique considerations	R → L shunt → systemic embolization	
Antibiotics	Cefazolin 25 mg/kg q 4 h	
Surgical time	Aortic cross-clamp: 30–60 min CPB: 90–120 min Total: 3–4 h	30–50 min 75–100 min 3–4 h
Closing considerations	Chest tube in pericardial (and possibly pleural) space, if opened; temporary ventricular pacing wires; routine right- and left-atrial lines for monitoring.	
EBL	Moderate	
Postop care	ICU × 2–3 d. 1–3 d assisted ventilation; inotropes for RV dysfunction	ICU for 1–2 d
Mortality	2%	
Morbidity	Junctional tachycardia Atrial dysrhythmias Hemorrhage Infection Low CO Stroke Heart block Residual VSD Transient, right-side CHF	
Pain score	8–10	8–10

(Print pagebreak 1223)

Patient Population Characteristics

Age range	Neonate-adult (usually < 6 mo)
Male:Female	3:2
Incidence	10% of congenital cardiac defects
Etiology	22g microdeletion is seen in some patients, who also may have DiGeorge syndrome; higher than expected prevalence of older maternal age at time of conception of affected children. A-V canal; PDA; previous systemic-to-pulmonary arterial shunt; anomalous left coronary artery from pulmonary artery

Associated conditions	(ALCAPA); metabolic acidosis (profound hypoxia); right-sided aortic arch

Anesthetic Considerations

Preoperative

Pathophysiology	The clinical presentation of TOF is dependent on the wide anatomic variation of this tetrad. The critical factor is the degree of RVOTO, which determines the amount of R → L shunting across the VSD. This obstruction is dynamic and can be related to RV infundibular spasm, and variations in pH, $PaCO_2$ and PaO_2 The onset of hypercyanotic episodes in an infant < 3 mo of age is the indication for complete surgical repair.
"TET" spells	Hypercyanotic episodes, or "TET" spells: These constitute medical and surgical emergencies; however, they are seen less frequently due to early surgical intervention. TET spells may present in early infancy and are related to infundibular spasm resulting in ↑ R→ L shunting. If untreated, these episodes can progress to unconsciousness, Sz, and death. For management of TET spells, see Table 12.4-1, p. 1224
Respiratory	Infectious/asthmatic pulmonary processes will complicate preop and postop cardiopulmonary function. Optimize pulmonary function preop. **Tests:** CXR: ↓ pulmonary vascular markings and characteristic heart shape.
Cardiovascular	CHF is not the usual presentation for an unrepaired Tet. An understanding of the patient's anatomic defects and their pathophysiology (e.g., is their RVOTO fixed or dynamic?) is essential for formulating the anesthetic plan. Avoid prolonged fasting to prevent profound decreases in preload on induction. **Tests:** ABG; ECG: for RVH, right axis deviation (RAD) ± complete or incomplete RBBB. CXR: heart size, "coeur en sabot" (elevation of apex 2° RVH, RAE). Cardiac cath and ECHO: Locate site of pulmonary outflow obstruction, VSD, bronchopulmonary collaterals, PDA, abnormal patency of previous surgical shunts (e.g., B-T shunt). Define aortic arch anatomy, abnormal coronary or subclavian arteries, tricuspid regurgitation/aortic insufficiency (TR/AI), ventricular function.
Hematologic	Polycythemia (typical with chronic O_2sat < 90%) →↑blood viscosity → thromboembolic events. Polycythemia →↓Plt + coagulopathy. Extreme polycythemia (> 70) may require isovolemic hemodilution intraop. **Tests:** CBC; Plt; bleeding time; coag profile
Laboratory	Electrolytes if on diuretics; otherwise, tests as indicated from H&P.
Premedication	As required to ↓ anxiety, avoid TET spells and facilitate separation from parents. Midazolam 0.5–0.75 mg/kg po 20 min before induction in a monitored setting.

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Table 12. 4-1. Treatment for TET Spells

Treatment Modalities	Effect
1. Knee-chest position	↑ SVR.
2. Abdominal compression	Simulates squatting. ↑ SVR.
3. Volume administration	↑ preload, opens RVOT.
4. O_2	Pulmonary vasodilator
5. Anesthesia and sedation	Negative inotropy ↑ reduced RVOT spasm.
6. Alpha-agonist (phenylephrine)	↑ SVR. Dose: 10 mcg/kg bolus and repeat.
7. Beta-blockers (esmolol)	Negative inotropy. Dose: 50 mcg/kg bolus.

Intraoperative

Anesthetic technique: GETA. Patients with TOF, pulmonary atresia, and MAPCAS (major aortopulmonary collaterals) requiring unifocalization, present a challenge for surgical and anesthetic management. Special periop requirements include: placement of cuffed ETTs is necessary. OLV also may be required to facilitate surgical exposure. Bronchial bleeding 2° extensive surgical dissection requires frequent ETT suctioning. A significant portion of the procedure is done off-CPB, and hemodynamic instability with ST segment changes may occur during surgical dissection. Closely monitor blood loss and replace mL/mL with PRBCs. Avoid air bubbles in iv tubing/stopcocks. In older patients presenting for conduit change or revisions, massive blood loss should be anticipated.

Induction	Typically mask induction with sevoflurane, O_2± air. If iv in place, ketamine (1–2 mg/kg) and/or fentanyl (10 mcg/kg) with rocuronium (1 mg/kg) ± volatile agent. Avoid ↓↓SVR. Verify ETT position (see p. D-2).	
Maintenance	For patients not undergoing unifocalization, anesthesia is maintained with isoflurane or sevoflurane. TET spells can occur 2° surgical manipulation. Hypercyanotic episodes can occur during surgical dissection for cannulation before CPB. The surgeon can provide direct aortic compression or urgently proceed to CPB. Refer to Table 12.4-1 for other treatment. For patients who are undergoing unifocalization, hypoxic pulmonary vasoconstriction is blunted by the use of volatile anesthetic agents, causing a further decrease in arterial saturation. As a result, we prefer to use total intravenous anesthesia with propofol/ketamine/midazolam infusion. Antifibrinolytics, such as Amicar, are routinely given to redo-patients to reduce blood loss.	
Emergence	Patients are transported to ICU monitored, intubated, and ventilated.	
Blood and fluid requirements	IV: 22–24 ga × 2 LR @ TKO T&C PRBC	Blood in OR before incision. For all repeat sternotomy patients, have blood available for immediate administration. Have massive transfusion system set up for older pateints.
Monitoring	Standard monitors Urinary catheter Arterial line (usually radial)	
	CVP line	Double-lumen CVP placed in IJ or femoral vein.
	TEE: Transthoracic LA land PA line: have transducers available.	During MAPCAS dissection and RVOT change with beating heart close monitoring of TEE for function and air.
Pre-CPB	Heparinization (4 mg/kg) ACT > 400 sec MAP 70 mmHg muscle relaxation. pupils. UO. Ventilation stopped. Hypothermia: 28°C to 32°C Hct 30 typical during CPB	In the pre-CPB period, crystalloid and/or 5% albumin is administered as needed to compensate for bleeding and 3rd-space losses.

CPB	adequate flow/pressure. anesthetic/NMB levels. pupils. face for venous congestion. ABG, electrolytes, UO, Hct, and ACT q 30 min.	Furosemide (0.5–1 mg/kg) for ↓ UO and hemo-concentration.
Transition off CPB	Rewarming Vasoactive infusions started at 32°C. Flush lines. TEE Zero transducers. Suction ETT. Resume ventilation. Observe lung inflation. Supplement NMB and deepen anesthesia. Rewarm to 36°C.	Surgeons de-air heart, which can be monitored by TEE. A measurement of the RV and LV pressure ratio helps to judge adequacy of repair of the RVOTO. TEE also is used to evaluate the anatomy, the repair, and pressures.
Post-CPB	bilateral breath sounds. ABGs, electrolytes, Hct (40). ACT (> 400). ECG: pacing may be necessary. Inotropic support (dopamine, milrinone, epinephrine, and CaCl) Rx pulmonary HTN (↓PVR): FiO_2= 1 $PaCO_2$ 35 Early use of inhaled nitric oxide (40 ppm) AV sequential pacemaker (heart block not uncommon). Transthoracic PA or LA line placed to monitor hemodynamics. Reverse anticoagulation. Rx coagulopathy	Infants are at risk for development of junctional ectopic tachycardia (JET). Avoid excessive warming > 37°C. Have amiodarone (5–10 mg/kg load) available. ↓ RV compliance is expected. RV afterload reduction is essential, initially through ventilatory efforts: clear unobstructed ventilation; do not allow wheezing (albuterol pm); use gentle hyperventilation on 100% O_2 Inotropic support of RV also typically is used—dopamine/initially and epinephrine reserved for severe RV dysfunction. Milrinone 0.5 mcg/kg/min and $CaCl_2$ 0–20 mg/kg/h are routine in infants. See Table 12.4-2 for blood product utilization. MUF uses the CPB machine to remove excess H_2O and inflammatory mediators (↓ edema and improved cardiopulmonary function).
Positioning	and pad pressure points. eyes.	
Complications	See Postoperative Complications, below.	

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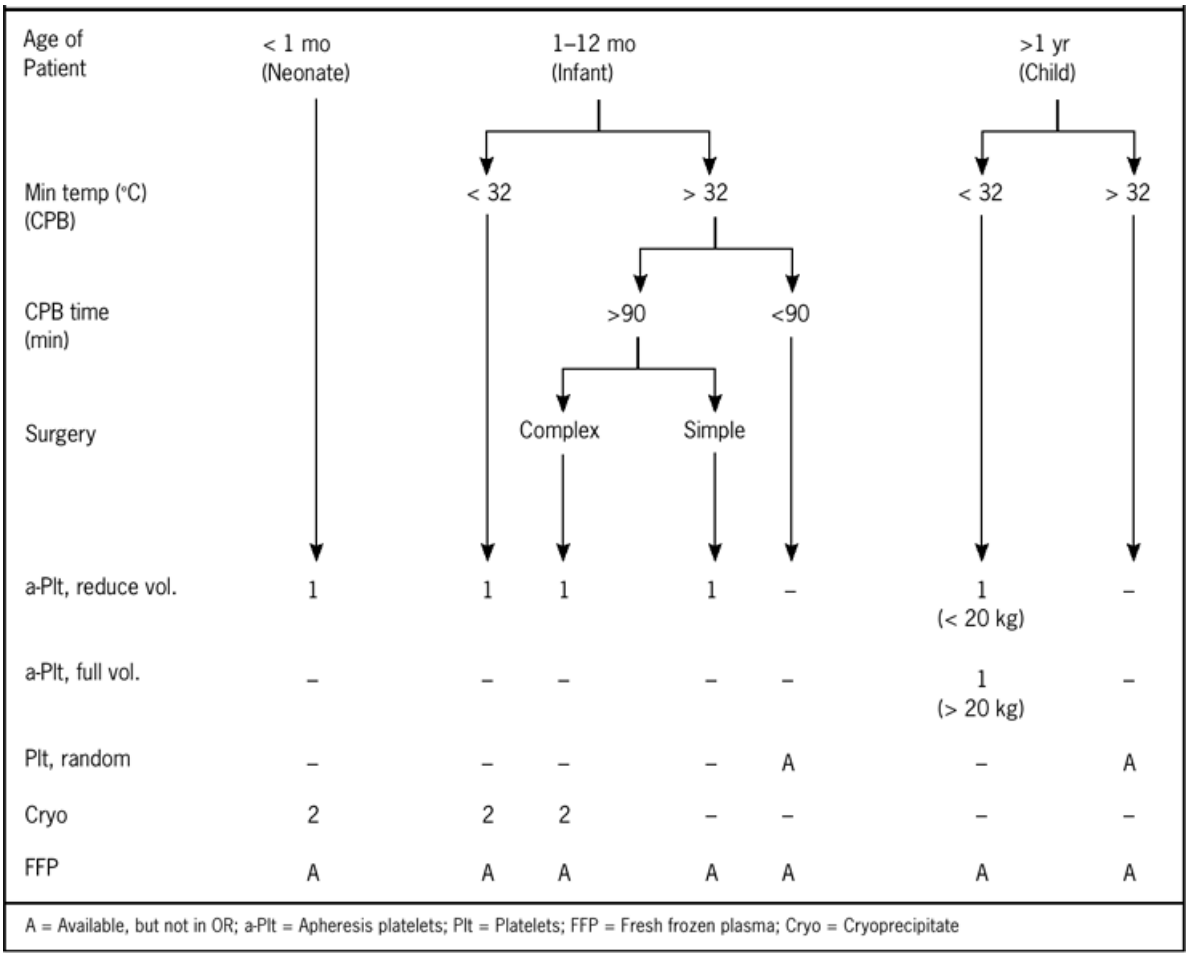
Table 12. 4-2. Blood Product Utilization [8]



A = Available, but not in OR; a-Plt = Apheresis platelets; Plt = Platelets; FFP = Fresh frozen plasma; Cryo = Cryoprecipitate

Postoperative

Complications	Persistent bleeding Persistent RVOTO Dysrhythmias (JET)	For neonatal repairs, a PFO is left surgically open to allow RV decompression via a R → L intraatrial shunt. This often will result in a ↓ systemic O_2sat. TEE will confirm the existence of intra-atrial shunting.
Tests	ABGs + electrolytes Coag profile CXR: line, ETT placement.	

(Print pagebreak 1227)

Suggested Readings

1. Allen HD, Clark EB, Gutgesell HP, et al., eds: *Moss and Adams' Heart Disease in Infants, Children, and Adolescents,* 6th edition. Lippincott Williams & Wilkins, Philadelphia: 2001.

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Surgery for Total Anomalous Pulmonary Venous Connection

Surgical Considerations

Description: Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac malformation in which the entire pulmonary venous return empties either directly into the systemic venous channels via anomalous veins or the right atrium (Fig. 12.4-7). An interatrial R → L shunt, in the form of a PFO or an ASD, is required to maintain systemic output and, hence, survival in the postnatal period. The objective of surgical correction is to redirect the entire pulmonary venous return to the left atrium.

TAPVC was first successfully repaired in 1956 by Lewis and Varco at the University of Minnesota by joining the pulmonary venous sinus to the left atrium and closure of the ASD. Although mortality for this lesion was initially quite high, particularly in infants with obstruction of the pulmonary veins, improvements in intraop and postop management have permitted successful correction in most neonates and infants. There are four TAPVC drainage patterns as defined by Darling, et al. These include **supracardiac** (45%), **cardiac** (25%), **infracardiac** (25%), and **mixed** patterns (5%) of venous drainage. The anomalous drainage in supracardiac TAPVC is usually by a left vertical vein into the innominate vein. In cardiac TAPVC, drainage is usually into the coronary sinus and occasionally into the right atrium. In infracardiac TAPVC, drainage is usually into the portal vein. The mixed type consists of combinations of the other three varieties of TAPVC.

When pulmonary venous drainage is obstructed, patients present with cyanosis severe pulmonary edema, pulmonary HTN, and ↓ CO. Symptomatic TAPVC is repaired at any age. TAPVC with obstruction is a true surgical emergency. Critically ill neonates with obstructed pulmonary venous return must undergo emergent correction after initial stabilization (i.e., intubation/ventilation, diuretics). Occasionally, preop ECMO may be required.

In most cases, the right and left pulmonary veins drain into a common pulmonary venous sinus, allowing for its anastomosis to the left atrium for a definitive repair. Through a standard median sternotomy, the aortic and venous cannulae are placed and CPB with cooling is initiated. The aorta is cross-clamped, immediately followed by cardioplegic arrest. The cardiac apex is lifted up, and the pulmonary veins are identified through the posterior pericardium. The left atrium is then opened transversely with extension onto the left atrial appendage, followed by the direct anastomosis of the pulmonary venous confluence to the left atrium. Finally, via a right atriotomy, the ASD or PFO is closed. The heart is de-aired, aortic cross-clamp is released, and the patient is rewarmed and separated from bypass. Successful outcomes are dependent on early and immediate correction of TAPVC with obstruction, before lung damage ensues. Other important factors include repair without residual obstruction and postop control of pulmonary HTN.

Variant procedure or approaches: Alternatively, repair of TAPVC may be accomplished through a right atriotomy and across the atrial septum, constructing the anastomosis between the pulmonary venous confluence and left atrium from within the left atrium or through a transverse sinus approach.

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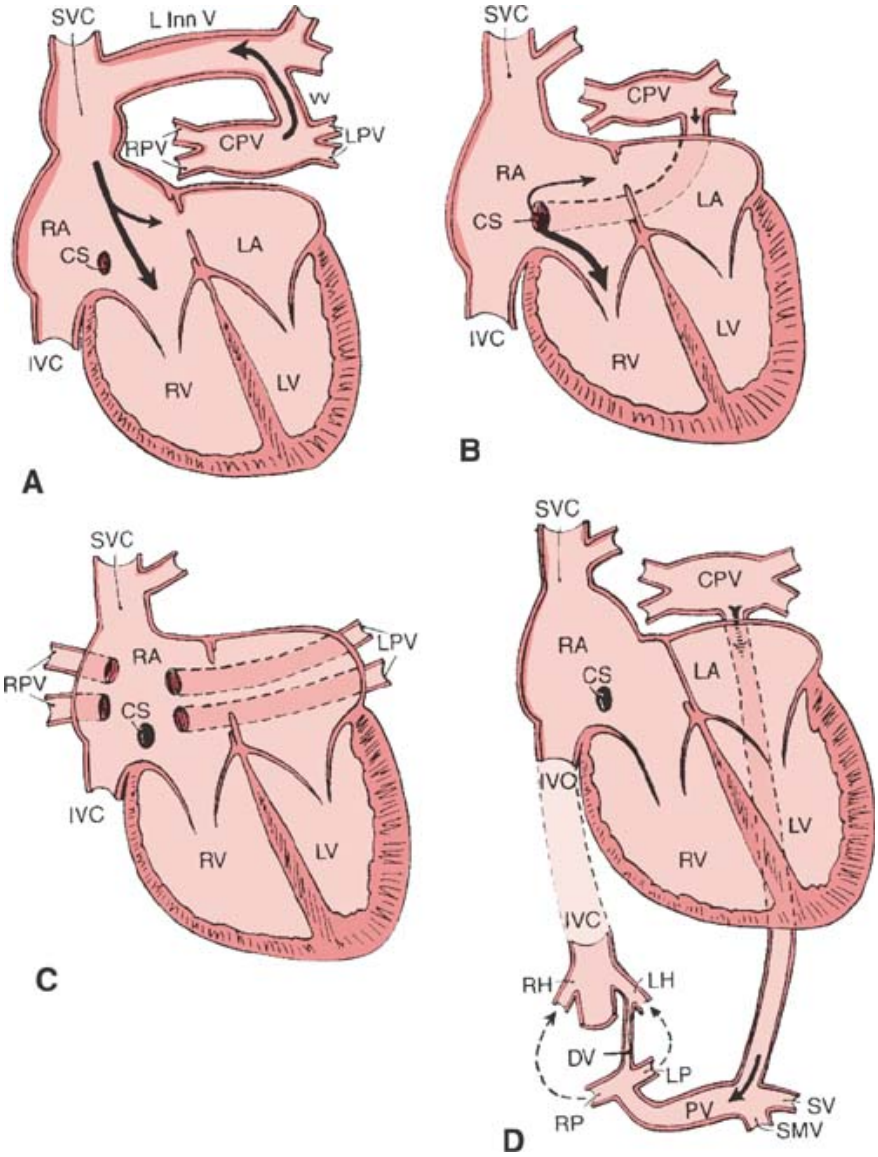

Figure 12.4-7. 7. Common forms of TAPVC: **(A)** TAPVC to the left innominate vein (L inn V) by way of a vertical vein (VV). **(B)** TAPVC to coronary sinus (CS). The pulmonary veins join to form a confluence designated as the common pulmonary vein (CPV), which connects to the coronary sinus. **(C)** TAPVC to right atrium (RA) The left and right pulmonary veins (LPV and RPV) usually enter the RA separately. (**D**) TAPVC to the portal vein (PV). The PVs form a confluence, from which an anomalous channel arises. (Reproduced with permission from Emmanouilides GC, Allen HD, Riemenschneider TA, et al: *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Philadelphia: 1998.)

Usual preop diagnosis: TAPVC; total anomalous venous return; total anomalous pulmonary venous drainage—all of these ± obstruction and either supracardiac, cardiac, or mixed types

Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
Unique considerations	Patients often require urgent or emergency surgery. Obligatory R → L shunting → risk of systemic embolization. Myocardial preservation using blood or crystalloid cardioplegia, in addition to topical myocardial hypothermia, is used.
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp: 30–60 min Circulatory arrest: occasionally, 30–45 min

	Total: 2–3 h
Closing considerations	A fine polyvinyl catheter inserted through the free wall of the right ventricle and advanced into the pulmonary trunk will allow monitoring for pulmonary HTN. A chest tube is inserted into the pericardial space; temporary ventricular pacing wires are placed. Left atrial lines normally needed.
EBL	Moderate
Postop care	2–6 d of assisted ventilation, sedation and hyperventilation; pulmonary vasodilators, including isoproterenol, PGE1, SNP; inotropes as needed for right heart dysfunction. Sometimes, if pulmonary injury is severe or pulmonary HTN is uncontrollable, ECMO may be indicated.
Mortality	2–10%, depending on presence of preop pulmonary venous obstruction and metabolic acidosis.
Morbidity	Pulmonary vasospasm: 25–40% Low CO: 10% Hemorrhage: 2–3%
Pain score	8–10

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Patient Population Characteristics

Age range	1 d–20 yr (usually < 1 mo)
Male:Female	4:1 in infracardiac type; equal distribution in other types
Incidence	0.5–2% of congenital heart defects
Etiology	Failure of fusion of the pulmonary vein evagination (from posterior left atrium) with the pulmonary venous plexus surrounding the lung buds.
Associated conditions	PDA present in nearly all infants within the first few wk of life and in about 15% of cases overall. VSD occasionally occurs (may be associated with TOF, DORV, interrupted aortic arch, and other lesions).

Anesthetic Considerations

Preoperative

Pathophysiology	In the supradiaphragmatic connection, all the pulmonary venous return drains into the right atrium via a vertical vein. When this vein or confluence is stenotic or obstructed, severe pulmonary HTN, cyanosis, pulmonary edema, and shock ensue. This is a true surgical emergency, requiring immediate intervention. An ASD or PFO is essential for survival.
Respiratory	In patients with obstructed TAPVR, expect severe pulmonary edema and pulmonary HTN. **Tests:** CXR: ↑ heart size, ↑ pulmonary vascularity, "figure-of-8," or "snowman" cardiac silhouette.
Cardiovascular	ECHO findings: large RV and small LA. Cardiac interventricular cath for balloon or blade septostomy in patients with restrictive ASD may be necessary to improve R → L shunting. **Tests:** ECG for right axis deviation (RAD), RAE and RVH.
Laboratory	ABG and Hct; other tests as indicated from H&P.

Premedication	Generally not indicated and not necessary in severely ill infants.
Transport of critically ill newborn	All critically ill neonates are intubated, ventilated, and often are on PGE_1 and/or dopamine infusions for hemodynamic stability. To facilitate transport of these patients, notify the nurse in advance, and request all drips transferred to OR syringe pumps. Maintain a dextrose-containing infusion during and after transport to ensure normoglycemia. Verify that the transport monitor has ECG, SaO_2 and invasive pressure transducers. Resuscitation medications and airway equipment must accompany the patient.

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Intraoperative

Anesthetic technique: GETA. Profound hypothermic CPB (16–18°C) with low flow may be required. Patients who weigh < 3.0 kg and the ICU ventilator are used in the OR, because this allows for maximum flexibility in ventilator support.

Induction	Infants are often severely ill and come to the OR intubated and ventilated, with invasive lines and inotropic support. Verify ETT placement. Patients are subject to rapid cardiovascular decompensation. Rocuronium (1 mg/kg) is given, as is fentanyl (1–3 mcg/kg) as tolerated. Volatile agents are rarely tolerated in obstructed TAPVC.	
Maintenance	Fentanyl (20–50 mcg/kg) in divided doses on CPB. Rocuronium prn, midazolam 0.1–0.2 mg/kg. FiO_2 = 1.0. Ventilation maneuvers to ↓ PVR. (See Post-CPB, p. 1225.)	
Emergence	Transport to ICU intubated and ventilated. If NO used in OR, must have delivery system ready for transport to ICU. There may be continued requirement for inotropic support.	
Blood and fluid requirements	IV: 22–24 ga × 2, taped securely LR @ TKO Continue iv dextrose in neonates. 5% albumin PRBC Blood warmer	Avoid air bubbles with R → L shunt—can cause systemic embolization of air. blood glucose. Avoid hyperglycemia, especially during profound hypothermic CPB. See Table 12.4-2 for blood product utilization.
Monitoring	Standard monitors (see p. D-1). Transthoracic LA, PA, RA lines	Severely ill infants usually present with invasive monitoring and ETT. correct placement of UAC and UVC preop.
Complications	Bleeding Refractory pulmonary HTN Biventricular failure Overdose of NO → methHb	Pulmonary HTN may be difficult to control. Ventilation maneuvers (e.g., ↓ $PaCO_2$, 100% O_2), vasodilators, and deep anesthesia to ↓ PVR. NO is started for refractory pulmonary HTN; if NO (40 ppm) is unsuccessful, ECMO may be instituted. In these instances sternum is not closed. Inotropic support (dopamine, milrinone, epinephrine, calcium chloride) metHb while on NO therapy.

Postoperative

Complications	See above.	
Tests	ABG Hct; electrolytes; coags Methemoglobin CXR	Only if on NO

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Surgery for Complete Transposition of The Great Arteries

Surgical Considerations

Description: Complete **transposition of the great arteries (TGA)** is a congenital cardiac defect in which the aorta arises from the RV and the PA arises from the LV (Fig. 12.4-8). TGA is associated with an intact ventricular septum (TGA/IVS) or a ventricular septal defect (TGA/VSD). Pathophysiologically, this discordant ventriculoarterial configuration results in systemic and pulmonary circulations placed in a parallel (normally in series) configuration. Thus, a L → R shunt in the form of an atrial septal defect (ASD), VSD, or patent ductus arteriosus (PDA) is required to permit oxygenated blood to enter the “right-sided” systemic circulation. The earliest surgical treatment for TGA was described by **Blalock** and **Hanlon** in 1950 with a procedure in which an **atrial septectomy** was performed, improving the mixing of pulmonary and systemic blood at the atrial level. In the 1950s, a variety of partial physiologic corrections were developed in which the pulmonary veins or the vena cava were transposed to the alternate atria. Palliative treatment was advanced by **Rashkind's** description of a **balloon atrial septostomy** in 1966. More complete physiologic correction was obtained by **atrial switch** operations, described by **Senning** in 1959 and **Mustard** in 1963, in which systemic and pulmonary venous return were baffled to the appropriate ventricles. Postop complications with the atrial switch operations, however, led to the development of the more “anatomic” **arterial switch** operations described by **Jatene, Yacoub,** and others beginning in the 1970s. By 1987, the arterial switch operation in neonates was widely accepted as the standard approach to TGA.

Total correction is now performed routinely in the first 2 wk of life. The heart is exposed through a standard median sternotomy and CPB is instituted. The aortic cross-clamp is applied, followed by cardioplegic arrest and induced hypothermia. The ascending aorta is transected just above the sinotubular junction at and the pulmonary trunk is transected just proximal to its bifurcation. Two buttons from the “neopulmonary artery” (former aortic root) containing the origins of the left and right coronary arteries are transposed and anastomosed to the “neoaorta” (former main pulmonary trunk). The distal aortic segment is swung beneath the PA bifurcation **(Lecompte maneuver)**. Pericardial patches are used to repair the defects resulting from excision of the coronary artery buttons, and any associated ASDs and/or VSDs are repaired. The distal aortic segment is anastomosed to the neoaorta and the distal bifurcated pulmonary artery segment is anastomosed to the neopulmonary artery. If hypothermia has been induced, a brief period of circulatory arrest facilitates repair of the ASD and/or VSD, if present. The aortic cross-clamp is removed and rewarming is begun during the neopulmonary arterial anastomosis. CPB is then discontinued and closure is routine.

Variant procedure or approaches: In children with TGA and VSD, additional cardiac anomalies are common. These include pulmonary stenosis, pulmonary atresia, straddling atrioventricular valves, hypoplasia of ventricular chambers, and coarctation or

interruption of aorta. Dynamic or structural subpulmonic obstruction is also more common in TGA and VSD than in TGA/IVS.

Rastelli procedure: The VSD is baffled to the aorta with construction of a RV→PA valved conduit and is the procedure of choice for patients with TGA/VSD and pulmonary atresia or significant valvular pulmonary stenosis. Often the VSD may have to be enlarged to prevent LVOT obstruction. Alternatively in some cases an aortic root translocation may be required.

Coronary arteries in TGA: Abnormalities in the origin and course of coronary arteries are common in TGA and, in the past, influenced the success of surgery. A simple rule that accounts for virtually all variations is that the coronaries arise from the sinuses of Valsalva, which face the pulmonary artery, and follow the shortest route to their ultimate destination. Only a small number pose a problem in switching the great arteries.

Usual preop diagnosis: Complete TGA; transposition of the great vessels; transposition ± VSD; ASD; PDA

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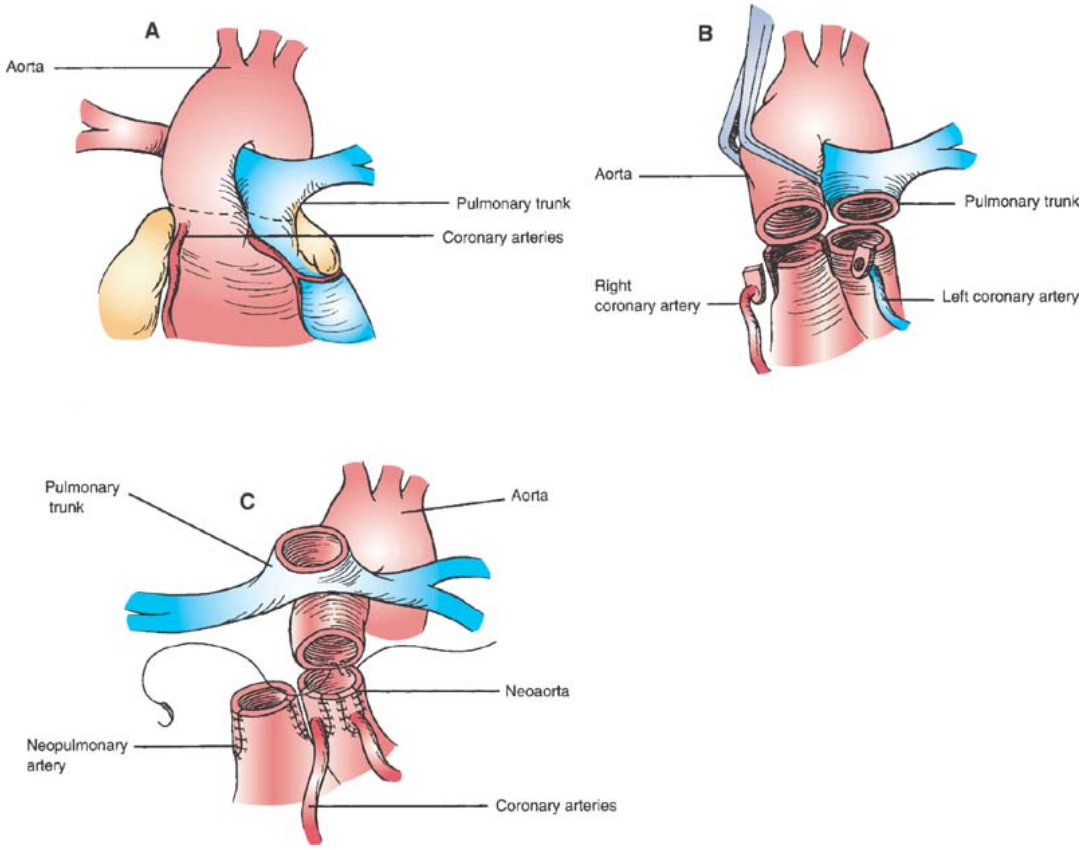

Figure 12.4-8. 8. TGA: **(A)** Typical anatomy of transposition of the great vessels, with the aorta anterior and slightly to the right of the pulmonary artery. The coronary arteries can be seen arising from the aorta. **(B)** Repair is best accomplished by an arterial switch procedure in infancy. The great vessels are transsected, and the coronary arteries are removed from the aorta and placed into the proximal pulmonary artery (neoaorta). **(C)** The distal aorta is brought behind the pulmonary artery bifurcation (Lecompte maneuver), and the neoaorta anastomosis is completed. (Reproduced with permission from Ungerleider RM, Plunkett MD, Gaynor JW: Congenital heart disease. In *Surgery of Infants and Children*. Oldham KT, Colombani PM, Foglia RP, eds. Lippincott-Raven, Philadelphia: 1997.)

Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
Unique considerations	Operation to be performed before LV pressure falls substantially 2° ↓PVR.
Antibiotics	Cefazolin 25 mg/kg q 4 h

Surgical time	Aortic cross-clamp: 45–90 min No circulatory arrest Total: 3 h
Closing considerations	Routine closure with chest tube in the pericardial space; temporary ventricular pacing wire; possible right or left atrial line for monitoring.
EBL	Moderate
Postop care	1–5 d of assisted ventilation; pulmonary HTN protocol consisting of hyperventilation and sedation and use of pulmonary vasodilators; inotropes for adequate CO.
Mortality	1–3%
Morbidity	Hemorrhage Atrial dysrhythmias ↓CO Coronary artery kinking and myocardial ischemia Pulmonary HTN
Pain score	8–10

(Print pagebreak 1233)

Patient Population Characteristics

Age range	1–21 d; ≥ 3–6 mo in TGA/VSD
Male:Female	2:1; for TGA/IVS, 3.3:1
Incidence	7–8% of congenital heart defects
Etiology	Usually no associated syndromes or other noncardiac abnormalities
Associated conditions	ASD; VSD; PDA; LVOTO; pulmonary atresia; coarctation of the aorta

Anesthetic Considerations

Preoperative

Pathophysiology	In TGA, the pulmonary and systemic circulations flow in parallel, unlike the normal series circulation. Deoxygenated blood returns to the right heart and is pumped to the aorta and systemic circulation without passing through the lungs for gas exchange. The oxygenated blood returns via the pulmonary veins and recirculates through the LV back into the lungs. Survival depends on mixing of pulmonary and systemic blood at some level (ASD, VSD, PDA). In the absence of a septal defect, a balloon atrial septostomy (Rashkind) may be required before surgery to promote mixing.
Respiratory	Some patients are maintained preop on PGE_1 to maintain ductal patency. PGE_1 can cause apnea; therefore, mechanical ventilation may be required. **Tests:** CXR: enlarged heart and pulmonary edema.
Cardiovascular	Patients with VSD have ↑ pulmonary blood flow, ↑ intercirculatory mixing → CHF. Marked cyanosis is seen in patients with TGA and intact ventricular septum, O_2sat depends on degree of mixing (Q_P/Q_S ratio). **Tests:** EKG: right axis deviation (RAD) and RVH. ECHO. Cardiac cath defines anatomy, coronary arteries, septal defects,

	PFO, PDA, bronchopulmonary collaterals, LVOT, pulmonic valve function and pressures, including PVR and LV:RV ratio. Balloon atrial septostomy (Rashkind-Miller) is required to improve mixing in some patients.
Laboratory	Hct; coags; other tests as indicated from H&P.
Premedication	Generally not necessary in children < 9 mo.
Transport of critically ill newborn	All critically ill neonates are intubated, ventilated, and often are on PGE_1 and/or dopamine for hemodynamic stability. To facilitate transport of these patients, notify the nurse in advance and request all drips transferred to OR syringe pumps. An air-O_2 blender should be available so the inspired O_2 concentration can be adjusted as needed. Verify that the transport monitor has EKG, SaO_2 and invasive pressure transducers. Resuscitation medications and airway equipment must accompany the patient. Patients can be transported on room air with gentle manual ventilation, taking care to avoid hyperventilation. Avoid oversedation and use of muscle relaxants before transport.

(Print pagebreak 1234)

Intraoperative

Anesthetic technique: GETA

Induction	Neonates with TGA and intact ventricular septum are placed on PGE_1 to maintain ductal patency and promote mixing of pulmonary and systemic blood. Anesthetic management is based on maintaining a balance between systemic and pulmonary blood flows. Excessive pulmonary blood flow →↓ systemic perfusion. IV induction with ketamine, fentanyl (e.g., 2–15 mcg/kg incrementally) and muscle relaxant (e.g., rocuronium 1 mg/kg) is the technique of choice. Inotropic infusion and PRBC transfusion may be required to support BP.
Maintenance	For patients with inadequate intercirculatory mixing and ↓ pulmonary blood flow, treatment should include ventilation maneuvers to ↓ PVR (see TOF, Post-CPB, p. 1225) and improve mixing. Deep GA helps to blunt reactive pulmonary HTN. Avoid ↓ HR →↓ CO in infants with limited myocardial reserve.
Emergence	Control aortic BP and treat any coagulopathy. Monitor LA pressures closely. for myocardial ischemia, which can be 2° coronary air emboli or at the implantation site of the coronaries. Transport to ICU intubated and ventilated.
Blood and fluid requirements	IV: 22–24 ga × 1–2 taped securely LR @ TKO Continue iv dextrose in neonates. 5% albumin Standard monitors (see p. D-1).
	Avoid air bubbles in patients with R → L and L → R shunts. See Table 12.4-2 for blood products utilization. Administer blood and products slowly, with close monitoring of LAP. Overtransfusion can cause systemic HTN, LA dilation, or pulmonary edema, producing additional suture line bleeding or mitral regurgitation. Severely ill infants often present with invasive monitoring and mechanical ventilation. Correct placement of UAC and UVC should be checked preop. Also verify position of ETT.

Monitoring	Transthoracic monitoring lines (RA, LA, or PA)	Placed routinely on rewarming, before weaning from CPB. These are for monitoring and administration of inotropes and other medications. The LV can be deconditioned may be dysfunctional and require inotropic support (dopamine, milrinone, epinephrine, and CaCl infusion). During weaning from CPB, it is critical to monitor LAP, as a reflection of left-sided filling, simultaneously with RA pressure monitoring.
	TEE	Abnormalities in coronary perfusion may manifest as changes in EKG rhythm and should be closely followed. TEE is useful both as a monitor of function and volume status. ECMO may be needed for severe ventricular dysfunction.
CPB	Management of CPB is discussed in Intraoperative Considerations for TOF, (p. 1225).	Low flow or moderate hypothermic CPB. Cool with pH stat management (T corrected ABG). Warm on reperfusion with alpha stat management.
Complications	Bleeding LV dysfunction and failure Myocardial ischemia Residual structural defects Dysrhythmias Pulmonary HTN	In patients with TGA and IVS, the LV may be inadequate to support the systemic circulation. The LV becomes deconditioned after the PVR drops in early infancy. The LV is then unable to function as the systemic ventricle. Inotropic support (dopamine and epinephrine) and afterload reduction (SNP or amrinone) may be necessary to separate from CPB. ECMO may be required for severe ventricular dysfunction.

(Print pagebreak 1235)

Postoperative

Complications	Hypothermia Bleeding Dysrhythmias Myocardial ischemia LV failure	Ischemia 2° coronary artery stenosis, stretching, spasm, or compression. Spasm can be treated with NTG and ↑ aortic pressure. If the now-anterior PA dilates (↓ PaO_2 ↑$PaCO_2$ ↓T, ↓ pH, excessive or too little PIP, or RV failure), the coronary arteries may be compressed. Measures to ↓ PA pressure are often essential.
Tests	ABGs; electrolytes; coag profile CXR TEE	line, chest tube placement; for pneumothorax.

Suggested Readings

1. Castaneda AR, Jonas RA, Mayer JE Jr, et al: D-transposition of the great arteries. In *Cardiac Surgery of the Neonate and Infant.* WB Saunders, Philadelphia: 1994, 409–38.

2. Castaneda AR, Mayer JE: Neonatal repair of transposition of the great arteries. In *Fetal and Neonatal Cardiology*. Long WA, ed. WB Saunders, Philadelphia: 1990.

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4. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Great artery anomalies. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Philadelphia: 1998, 501–14.

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6. Quaegebeur JM, Auteri JS: Transposition of the great arteries. In *Glenn's Thoracic and Cardiovascular Surgery*, 6th edition. Baue AE, ed. Appleton & Lange, Stamford: 1996, 1393–1407.

7. Spray TL: Transposition of the great arteries. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 785–99.

Surgery for Truncus Arteriosus

Surgical Considerations

Description: Truncus arteriosus is a rare cardiac anomaly in which there is a common aortopulmonary trunk originating from the base of the heart by way of a single, semilunar valve (truncal valve). This single great artery gives rise to the pulmonary, systemic, and coronary circulations. A nonrestrictive VSD is almost always found immediately below the truncal valve. An anatomic classification scheme proposed by **Collett** and **Edwards** in 1949 describes four types of truncus. In **type I** truncus (60%), a single arterial trunk gives rise to the aorta and main PA (Fig. 12.4-9A). In **type II** truncus (20%), the right and left PAs arise separately from the posterolateral aspect of the truncus. **Type III** *(Print pagebreak 1236)* truncus (10%) designates cases in which the two PAs also originate from the posterior truncus, but with widely separated orifices. In **type IV** truncus (10%), the PA branches are absent with pulmonary blood flow derived from aortopulmonary collaterals. Pathophysiologically, there is significant L→R shunting at the truncal and ventricular levels, leading to unrestricted pulmonary blood flow, CHF, and pulmonary HTN. Patients become symptomatic in infancy 2° CHF when the PVR ↓ and CO ↑. Untreated, 90% of infants die within 6 mo.

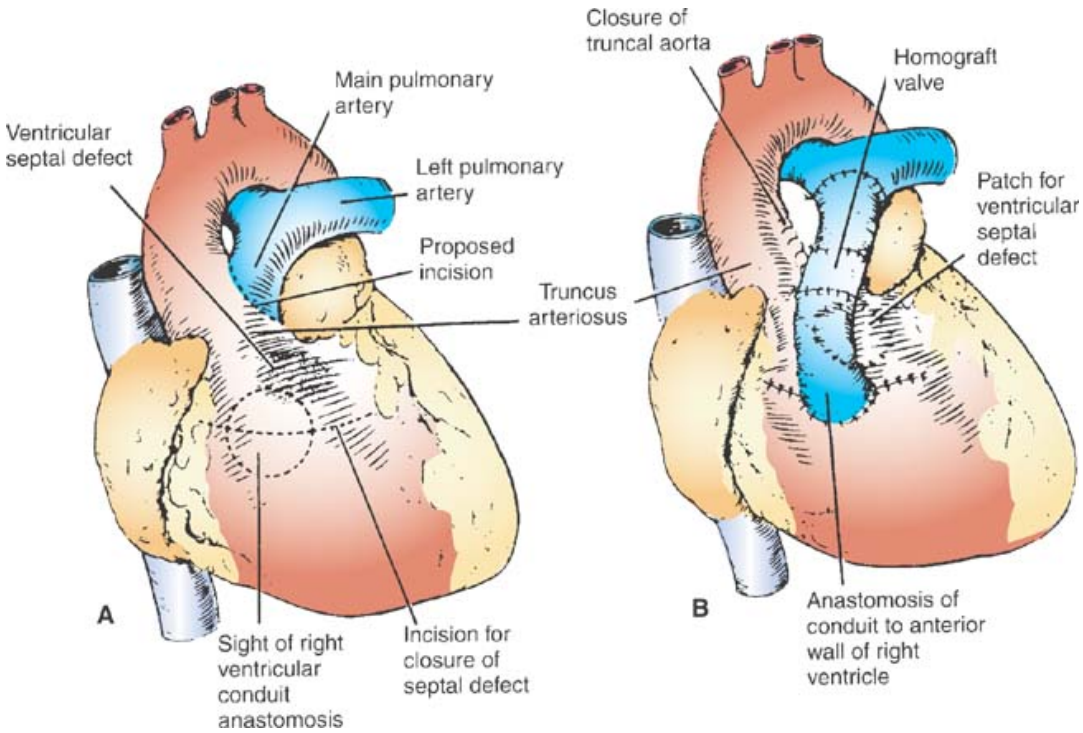

Figure 12.4-9. 9. Truncus arteriosus. (**A**) Cohen-Edwards (Type I). (**B**) Surgical correction involves the separation of

the main PA from the truncus, VSD patch closure and insertion of a conduit between the RV and the distal main PA. (Reproduced with permission from Way LW: *Current Surgical Diagnosis and Treatment,* 10th edition. Lange Medical Publications, City: 1973.)

The first total repairs of this congenital anomaly were reported in the early 1960s and included the insertion of **nonvalved artificial conduits** and **aortic allograft and valve conduits.** In the early and mid-1970s, repair was performed at younger ages, with abandonment of palliative banding of the PA branches. Currently, early primary repair is carried out during the first few weeks of life.

Through a median sternotomy, the truncus and PA branches are dissected and cannulation for CPB is performed. The PAs are snared to block pulmonary flow during perfusion. Hypothermia is induced and the aorta is cross-clamped. The PAs are separated from the main truncus, the resultant aortic defect is repaired with a patch, and a valved homograft (12–14 mm) is prepared. An end-to-end anastomosis of the distal homograft to the PA opening is performed, the VSD is closed through a right ventriculotomy, and the proximal end of the homograft is then anastomosed to the right ventriculotomy (Fig. 12.4-9B). The heart is de-aired, the aorta is unclamped, rewarming and resuscitation are performed, and CPB is discontinued.

There are several **special considerations** associated with this repair that warrant mention. The truncal valve may exhibit insufficiency, necessitating valve repair or replacement using a cryopreserved aortic or pulmonary homograft. In older infants, the PVR may be elevated and pulmonary hypertensive crises should be anticipated. In some patients, an associated interruption of the aortic arch has to be addressed and increases the complexity of the procedure. Coronary artery abnormalities are common in patients with truncus arteriosus and may contribute to their mortality.

Usual preop diagnosis: Truncus arteriosus (types I, II, III, IV)

(Print pagebreak 1237)

Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp: 30–90 min Total: 2.5–3 h
Closing considerations	Routine closure with chest tube in the pericardial space; temporary ventricular pacing wire; possible right or left atrial line for monitoring
EBL	Moderate
Postop care	1–5 d of assisted ventilation; pulmonary HTN protocol consisting of hyperventilation and sedation and use of pulmonary vasodilators. Inotropes for adequate CO.
Mortality	< 3%
Morbidity	Hemorrhage Atrial dysrhythmias Pulmonary vasospasm Truncal valve insufficiency ↓CO
Pain score	8–10

Patient Population Characteristics

Age range	3 wk–6 mo
Male:Female	1:1
Incidence	1.7–4.6% of congenital heart defects
Etiology	No known etiology Coexisting interrupted aortic arch or coarctation with PDA

Associated conditions	(10–20%); right aortic arch (16%); mitral valve anomalies (10%); moderate-size or large ASD (10%); DiGeorge syndrome

Anesthetic Considerations

Preoperative

Pathophysiology	A single, common aortopulmonary artery—the truncus—overlies the two ventricles and VSD. Depending on the configuration, the truncus gives rise to coronary and pulmonary arteries and ascending aorta. Complete mixing of systemic and pulmonary circulations occurs at the VSD and truncal valve, resulting initially in cyanosis. After birth, as PVR decreases, significant L → R shunting occurs at the level of the VSD. This causes pulmonary blood flow and CHF. If present, truncal valve insufficiency causes ventricular dilatation and low diastolic coronary perfusion, → myocardial ischemia. If untreated, 80% die of CHF in the 1st yr. Early total correction is indicated.
Airway	The facial anomalies (e.g., micrognathia) in DiGeorge syndrome may make intubation difficult.
Respiratory	Typically, these infants present with the respiratory Sx of CHF (dyspnea, tachypnea, hypoxemia). **Tests:** CXR: ↑ pulmonary markings and wide mediastinum.
Cardiovascular	Frequently, neonates are intubated and ventilated for hemodynamic support. Arterial saturation > 85% indicates excessive pulmonary blood flow. This may be controlled by the use of hypoxic gas mixtures (17–19% FiO_2), inspired CO_2 (3–5%), and PEEP. **Tests:** EKG. ECHO: Usually diagnostic. Identifies the origin of the PA from the common trunk and the presence (subtype A) or absence (subtype B) of a VSD. Evaluates truncal valve competency.
Endocrine	In DiGeorge syndrome, thymic hypoplasia occurs with hypoparathyroidism and symptomatic hypocalcemia. **Tests:** Ca^{++} levels; parathyroid hormone
Premedication	Not indicated.
Transport to OR	See Transport of the Critically Ill Newborn (see p. 1229).

(Print pagebreak 1238)

Intraoperative

Anesthetic technique: GETA

Induction	The goal for induction is maintenance of hemodynamic stability. If not intubated prior to induction, do not preoxygenate or hyperventilate. IV induction with ketamine (1–2mg/kg), fentanyl (2–4 mcg/kg) and muscle relaxant (e.g., rocuronium 1 mg/kg) is preferred. After ETT is secured, avoid hyperventilation and maintain arterial saturation 75–85%. Keep DBP > 20 mmHg to maintain coronary perfusion. Treat ↓ BP on induction with additional inotropes, transfusion of blood, or occlusion of a PA by the surgeon as soon as the chest is opened.

Maintenance	Fentanyl (20–50 mcg/kg). Rocuronium as needed ± midazolam 0.2 mg/kg ± volatile agent. FiO_2= 0.21. Avoid hyperventilation (↓ PVR →↑ shunt + CHF). PEEP may be useful to maintain PVR. The surgeon may leave a PFO to serve as a "pop-off" to preserve systemic perfusion when ↑ PVR and RV dysfunction occur. A transthoracic left atrial line is frequently placed for monitoring. In some cases, the sternum may be left open due to chest-wall and mediastinal edema and the presence of the anterior RV-PA conduit (Rastelli conduit).	
Emergence	Transport to ICU intubated and ventilated. Maintain narcotic and muscle relaxant infusions (e.g., fentanyl and vecuronium) for initial postop period (2–3 d) to minimize pulmonary hypertensive crises.	
Blood and fluid requirements	See Tetralogy of Fallot (TOF).	Continue dextrose infusion in neonates. See Table 12.4-2, p. 1226 for blood product utilization.
Infusions	Inotropic support	These patients require inotropic support (dopamine, milrinone, epinephrine, and CaCl infusion).
Monitoring	Standard monitors + LA, RA, PA lines TEE	Transthoracic lines TEE is helpful to assess function, truncal valve competency, RV function, and the direction of atrial shunt.
Positioning	See TOF (p. 1225).	
CPB	See TOF (p. 1225).	
Complications	Pulmonary HTN Truncal valve insufficiency Coronary artery insufficiency Residual lesion Bleeding	
	Pulmonary hypertensive crisis	Frequent in patients > 1 mo old. Rx: NO, sedation, PGE_1, hyperventilation.
	A-V block	Generally temporary, injury to conduction system unlikely because it is remote from VSD closure. Occasionally, a pacemaker may be required.
	Ventricular dysfunction Residual lesions	May require inotropic support. Residual VSD, severe truncal valve incompetence or obstruction, obstruction of coronary flow, LVOTO.

(Print pagebreak 1239)

Suggested Readings

1. Allen HD, Clark EB, Gutgesell HP, et al, eds. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents*, 6th edition. Lippincott Williams & Wilkins, Philadelphia: 2001.

2. Castaneda AR, Jonas RA, Mayer JE Jr, et al: Truncus arteriosus. In *Cardiac Surgery of the Neonate and Infant*. WB Saunders, Philadelphia: 1994, 281–93.

3. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Truncus arteriosus. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. EDITORS. Williams & Wilkins, Philadelphia: 1998, 434–41.

4. Kouchoukos NT, Blackstone EH, Doty DB, et al. Truncus arteriosus. In *Kirklin, Barrett, Boyes Cardiac Surgery*. Churchill Livingstone, Philadelphia: 2003, 1200–22.

5. Ramamoorthy C, Tabbutt S, Kurth CD, et al: Effects of inspired hypoxic and hypercapnic gas mixtures on cerebral oxygen saturation in neonates with univentricular heart defects. *Anesthesiology* 2002; 96:283–8.

6. Reitz BA, Yuh DD, eds: *Congenital Cardiac Surgery*. McGraw-Hill, New York: 2002.

7. Spray TL: Truncus arteriosus. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 759–70.

8. Tabbutt S, Ramamoorthy C, Montenegro LM, et al: Impact of inspired gas mixtures on preoperative infants with hypoplastic left heart syndrome during controlled ventilation. *Circulation* 2001; 104(Supp I):I159–64.

Surgery for Tricuspid Atresia

Surgical Considerations

Description: In tricuspid atresia, there is a developmental failure of the tricuspid valve, isolating the RA from the RV. There are three types of tricuspid atresia, based on the relationship of the great vessels to the ventricles, otherwise known as **ventriculoarterial concordance.** Type I, the most common (60–80%), consists of normal ventriculoarterial concordance. Type II (15–25%) consists of *d*-transposition, and Type III (3%) consists of *l*-transposition. In most cases of tricuspid atresia, the RV is hypoplastic, an ASD is present, and pulmonary blood flow is restricted 2° pulmonary stenosis or atresia. Together, these malformations lead to R → L shunting and varying degrees of cyanosis. Those patients with unobstructed pulmonary blood flow develop pulmonary overcirculation and CHF. Consequently, the initial palliative surgical management of this defect depends on the magnitude of pulmonary blood flow; the initial procedure may be either a modified **Blalock-Taussig systemic-to-PA shunt** or a **PA band.** The subsequent definitive surgical management consists of a bidirectional **Glenn shunt** and a modified **Fontan procedure.** Patients undergo these operations sequentially or, in rare cases, directly to the definitive Fontan operation.

The systemic-to-PA shunt, developed by **Blalock** and **Taussig** in 1944, was the first palliative treatment for this condition. Later, other shunts were introduced by **Potts** (descending aorta-to-left PA) and **Waterston** (ascending aorta-to-right PA). In 1958, Glenn described a shunt from the SVC to the right PA applied specifically to patients with tricuspid atresia. A modification of this shunt to a **bidirectional cavopulmonary anastomosis** was performed clinically by **Azzollina** in 1974 and has since gained widespread acceptance. In 1971, Fontan proposed a surgical repair for tricuspid atresia based on separation of the right and left circulations. Subsequent modifications to Fontan's original operation were designed to bypass the RV and direct systemic venous return to the pulmonary circulation. The most recent modifications divert vena caval blood directly to the PA—now referred to as a **total cavopulmonary connection** *(Print pagebreak 1240)* (modified Fontan). In 1988, **Laks** suggested leaving a small ASD between the right and left circulations, creating a R → L shunt for the purposes of augmenting systemic CO while still maintaining adequate oxygenation.

Modified Blalock-Taussig (B-T) shunt: In neonates with tricuspid atresia or other single ventricle variants with low pulmonary blood flow, a modified B-T shunt is the procedure of choice. The preferred approach is through a midsternotomy. The right PA and the right innominate and subclavian arteries are mobilized. The patient is heparinized and clamps are applied on the right PA and the innominate/subclavian arteries. Arteriotomies are performed and a Gore-Tex tube graft is interposed and anastomosed to the right PA and the subclavian/innominate arteries. The size of shunt (3–4 mm) is dictated by patient size and arterial anatomy. Alternatively, the procedure can be performed via right thoracotomy or through a left thoracotomy if the morphology dictates a left-sided shunt. A midline approach can be used for any variation of the shunt and also gives the option of using CPB in case of patient instability.

Pulmonary artery banding: In patients with tricuspid atresia or other variants of single ventricle and increased pulmonary blood flow, PA banding is performed to protect the pulmonary vascular bed, prevent pulmonary HTN, and prevent CHF. A midline sternotomy is the preferred approach. The main PA is exposed and a 2–3 mm wide strip of Silastic band is placed around the main PA and tightened to adjust the pulmonary blood flow. The goal is a SaO_2 of 80–85%.

Bidirectional Glenn shunt: A standard median sternotomy approach is used. If concomitant intracardiac repairs are not anticipated, the bidirectional Glenn shunt may be performed without CPB by placing a temporary shunt between the high SVC and the RA. If the patient has a preexisting Blalock-Taussig shunt on the right, it is divided after instituting CPB. A shunt on the left side may be left open, while the bidirectional Glenn shunt is created on the right. The SVC is divided and the cardiac end is oversewn. The remaining caval end is anastomosed end-to-side to the superior aspect of the right PA (Fig. 12.4-10A).

Fontan procedure: The heart is accessed via a standard median sternotomy. Previously placed systemic-to-PA shunts are dissected and occluded prior to bypass. Bicaval cannulation and CPB with hypothermia are used. The IVC is transected at its PA junction and a Gore-Tex tube graft is interposed end-to-end between the IVC and the inferior surface of the right PA. This operation, in conjunction with the previously performed bidirectional Glenn shunt, establishes a total cavopulmonary connection (Fig. 12.4-1B). After completing the anastomosis, the aortic cross-clamp is removed, the heart is rewarmed and the patient is weaned from CPB. Fenestration of the Fontan *(Print pagebreak 1241)* circuit may be desirable in high-risk patients, but is generally not required in patients with tricuspid atresia and good ventricular function. Our current standard is to perform the Fontan procedure without using cardiopulmonary bypass.

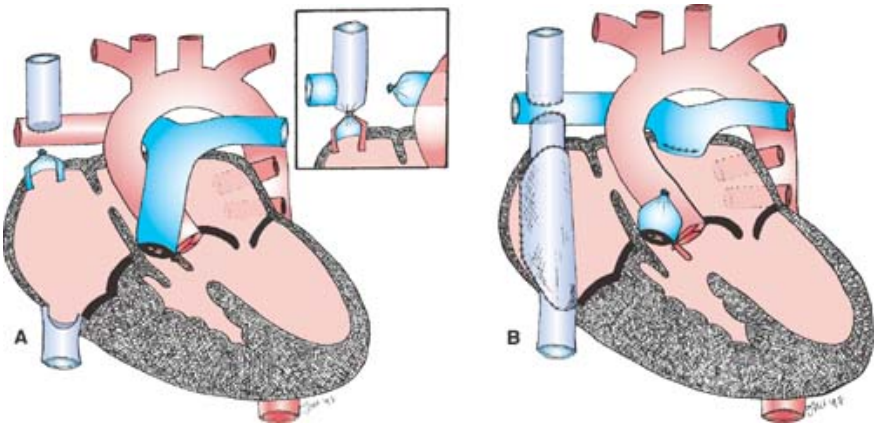

Figure 12.4-10. 10. Surgery for tricuspid atresia. **(A)** Bidirectional Glenn shunt. Inset shows classic unidirectional Glenn shunt. **(B)** Lateral tunnel Fontan operation. (Reproduced with permission from Emmanouilides GC, Allen HD, Riemenschneider TA, et al: *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents.* Williams & Wilkins, Philadelphia: 1998.)

Variant procedure or approaches: A RA → RV connection can be performed in patients with tricuspid atresia without pulmonary obstruction and an adequately functioning RV. This is accomplished with a direct anastomosis or a valved/nonvalved conduit (e.g., aortic homograft, Dacron graft).

Usual preop diagnosis: Tricuspid atresia after B-T shunt or PA banding; univentricular heart or single ventricle after shunt or band; cyanotic congenital heart disease

Summary of Procedures

	Bidirectional Glenn Shunt	Cavopulmonary Fontan Procedure
Position	Supine	
Incision	Standard median sternotomy	
Unique considerations	Particulate or gaseous emboli from iv lines can become systemic emboli.	Areas of PA stenosis need to be repaired concomitantly; subaortic obstruction needs to be considered if there is a restrictive VSD and the great vessels are transposed.
Antibiotics	Cefazolin 25 mg/kg q 4 h	
Surgical time	If a simple anastomosis is constructed, the aorta is not cross-clamped. Total: 1.5–2 h	Total: 3–4 h
Closing considerations	Chest tube in the pericardial space; temporary ventricular pacing wire; possible atrial monitoring line to assess ventricular filling pressure	Chest tube in the pericardial space; possible pleural tubes; temporary ventricular and atrial pacing wires; possible atrial line to correlate with CVP to determine the transpulmonary gradient
EBL	Moderate	

Postop care	Cardiac ICU; early extubation to minimize positive intrathoracic pressure	ICU or early extubation; inotropes for ventricular function, including low-dose dopamine, milrinone for afterload reduction. Maintain sinus rhythm, A-V pacing, if required.
Mortality	< 2%	2–4%
Morbidity	Bleeding	Pleural effusions
	Infection	–
	Atrial dysrhythmias	
	↑SVC pressure	–
	Upper extremity edema (SVC syndrome)	–
	↓CO	
	Cyanosis 2° ↓pulmonary blood flow	–
	Pulmonary arteriovenous fistula	Protein-losing enteropathy (late)
Pain score	8–10	8–10

Patient Population Characteristics

Age range	4 mo–1 yr	2–5 yr
Male:Female	1:1	
Incidence	1–3% of congenital heart defects	
Etiology	Unknown	
Associated conditions	TGA; AV valve insufficiency requiring repair or replacement; residual PA obstruction from previous procedures; dextrocardia; asplenia or polysplenia syndromes	

(Print pagebreak 1242)

Anesthetic Considerations

Preoperative

Pathophysiology

This is characterized by absence of the tricuspid valve and a hypoplastic RV with no communication between RA and RV. Survival in the neonatal period depends on presence of an interatrial communication. The degree of cyanosis reflects the size of the VSD and magnitude of RVOTO. Thirty percent of patients have CHF from pulmonary overcirculation 2° a large VSD and unobstructed RVOT and may require PA banding. Tricuspid atresia requires staged palliation, beginning with the establishment of adequate pulmonary blood flow. Patients undergo a balloon septostomy or a systemic-pulmonary artery shunt (B-T shunt) in the newborn period to augment pulmonary blood flow. Subsequently, at 3–4 mo of age, they undergo cardiac catheterization, and a palliative bidirectional Glenn shunt (SVC→ PA) is performed and the B-T shunt is ligated. The Glenn shunt reduces the volume load of the systemic ventricle. The final palliative procedure—the Fontan operation—is done between 1.5–3 yr of age, where the IVC is connected to the PA via an extracardiac conduit. The pulmonary and systemic circulations are now in series.

Identify any infectious or asthma-related problems. Optimizing

Respiratory	pulmonary function is imperative as the pulmonary blood flow will be supplied passively from the systemic venous return. **Tests:** CXR; O_2sat
Cardiovascular	**Tests:** EKG, ECHO, cath
Laboratory	Hct; others as indicated by H&P.
Premedication	Midazolam 0.5–0.75 mg/kg po 20 min before induction in children > 9 mo old

Intraoperative/Postoperative

For intraop and postop considerations for tricuspid atresia, see Anesthetic Considerations for Surgery for HLHS (see p. 1248).

Suggested Readings

1. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Tricuspid valve abnormalities. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Philadelphia: 1998, 369–84.

2. Kouchoukos NT, Blackstone EH, Doty DB, et al: Tricuspid atresia and management of single ventricle physiology. in *Kirklin, Barrett, Boyes Cardiac Surgery*. Churchill Livingstone, Philadelphia: 2003, 1113–76.

3. Mayer JE Jr: Tricuspid atresia single ventricle and the Fontan operation. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 848–57.

Surgery for Double-Outlet Right Ventricle

Surgical Considerations

Description: In double-outlet right ventricle (DORV), at least 50% of both of the great arteries arise from the RV. By necessity, there is a VSD that is usually large, but may in some cases be restrictive or, rarely, multiple and muscular. *(Print pagebreak 1243)* The VSD may be located primarily below the aorta (**DORV/subaortic VSD**) (Fig. 12.4-11A), below the pulmonary valve (**DORV/subpulmonic VSD,** or **Taussig-Bing DORV**) (Fig. 12.4-11B), below both great vessels (doubly committed, Fig. 12.4-11C), or remote from both great vessels (noncommitted, Fig. 12.4-11D). In DORV/subaortic VSD, oxygenated LV blood is directed to the aorta and deoxygenated systemic venous return from the RV is directed to the PA, resulting in minimal-to-no cyanosis. L → R shunting across the VSD, however, leads to pulmonary overcirculation, pulmonary HTN, and CHF. In DORV/subpulmonic VSD, oxygenated LV blood is directed to the PA and deoxygenated systemic venous return from the RV is directed to the aorta → pulmonary overcirculation and significant cyanosis. If early PA banding or primary correction is not performed in patients with pulmonary overcirculation, PVOD may result at an early age. In patients with some degree of pulmonary stenosis (usually infundibular), R → L shunting and cyanosis results; the natural history resembles that of patients with tetralogy of Fallot.

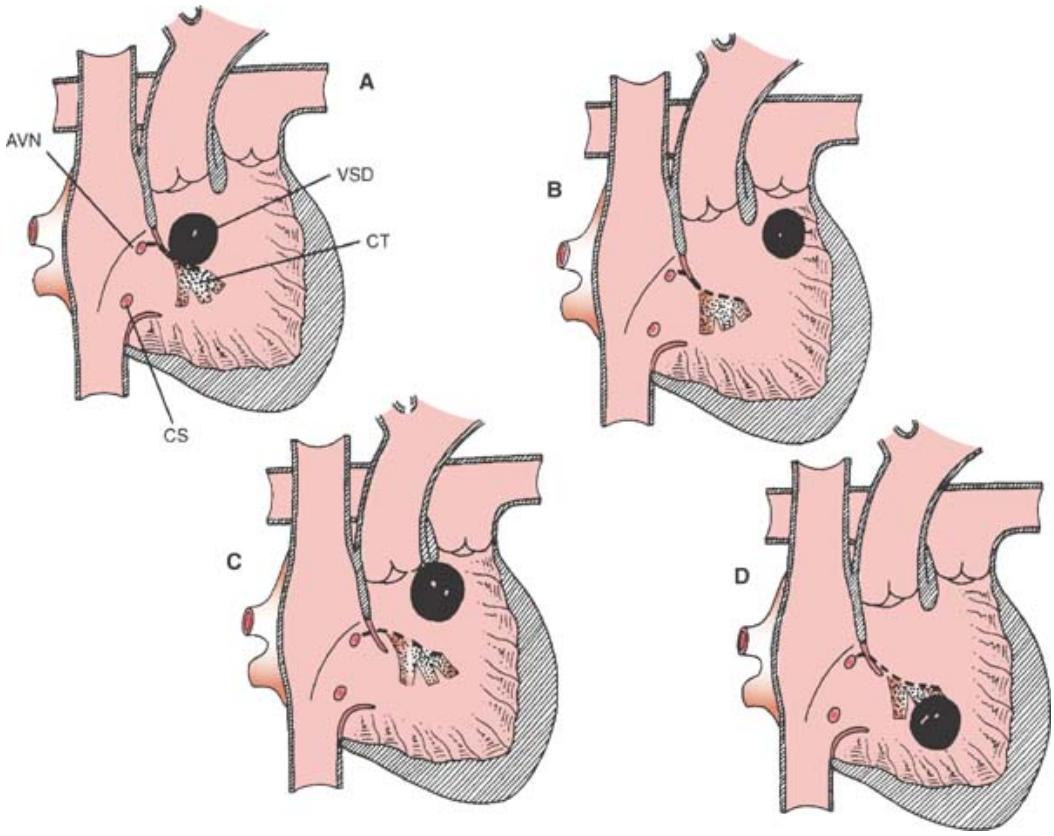

Figure 12.4-11. 11. Types of double-outlet right ventricle classified by the relationship of the VSD to the great arteries. The location of the atrioventricular node and conduction tissue is depicted. **(A)** Subaortic VSD. **(B)** Subpulmonary VSD. **(C)** Doubly committed VSD. **(D)** Noncommitted VSD. AVN = atrioventricular node; CS = coronary sinus; CT = conduction tissue. (Reproduced with permission from Kaiser LR, Kron IL, Spray TL, eds: *Mastery of Cardiothoracic Surgery*. Lippincott-Raven, Philadelphia: 1998.)

The first repairs of DORV were described by **Kirklin** in 1957. The Taussig-Bing DORV was first corrected in 1967. In recent years, management has been simplified for this variant by combining the **arterial switch operation** with an **intraventricular baffle to the pulmonary valve.** From a surgical standpoint, the anatomic variations and past attempts at correction may lead to unique intraop challenges. For example, if a previous Blalock-Taussig shunt has been performed, it needs to be controlled prior to initiating CPB. A previous pulmonary band may require removal and PA reconstruction.

DORV/subaortic VSD generally is repaired with an **intraventricular tunnel repair,** in which LV blood from the VSD is channeled through the RV to the aorta. Through a median sternotomy, CPB is instituted, and previous shunts are ligated. The aorta is cross-clamped and cardioplegia is administered. After performing a right atriotomy, the interior of the RV is inspected through the tricuspid valve; a right ventriculotomy may be necessary for adequate exposure. A tunnel constructed of pericardial or Dacron patch is then placed between the VSD and the subaortic infundibulum. Augmentation of the RVOT, in the form of an outflow patch or extracardiac RV → PA conduit, is often necessary if the outflow tract is obstructed or if the RVOT is encroached upon by the intraventricular tunnel. The heart is then rewarmed and resuscitated, CPB is discontinued, and standard chest closure is begun.

(Print pagebreak 1244)

DORV/subpulmonic VSD (Taussig-Bing DORV) can be corrected with an arterial switch operation and VSD closure under CPB (see Surgery for Complete Transposition of the Great Arteries, p. 1231). DORV with significant pulmonary stenosis repair resembles that for the TO. DORV with doubly committed VSD is similar to DORV/subaortic VSD. DORV with uncommitted VSD poses a surgical challenge. A two-ventricle repair requires intracardiac baffles. Alternatively, a single-ventricle approach is taken.

Usual preop diagnosis: DORV ± pulmonary stenosis; s/p PA band or modified Blalock-Taussig shunt; Taussig-Bing type of DORV; CHF

Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp: 30–80 min Total: 2–3.5 h
Closing considerations	Routine closure with chest tube in the pericardial space; temporary ventricular pacing wire; possible LA line for monitoring
EBL	Minimal
Postop care	ICU for 1–4 d of controlled ventilation; possible pulmonary HTN protocol in cases of unrestricted pulmonary blood flow (see description of management for A-V canal defect, p. 1206).
Mortality	2%
Morbidity	Hemorrhage ↓CO Atrial dysrhythmias RV dysrhythmias Intraventricular tunnel obstruction Baffle leakage; residual RVOTO Heart block
Pain score	8–10

Patient Population Characteristics

Age range	3–24 mo
Male:Female	1:1
Incidence	1–3% of congenital heart defects
Etiology	No specific correlations or associated conditions
Associated conditions	Pulmonary stenosis; complete A-V canal defect; coarctation of the aorta; interruption of the aortic arch; straddling tricuspid valve

(Print pagebreak 1245)

Anesthetic Considerations

Preoperative

Pathophysiology	Both great vessels arise from the RV and there is a single, large VSD, whose location may vary (subaortic, subpulmonic). The pathophysiology varies according to the VSD location and other concomitant cardiac anomalies, such as aortic stenosis (AS) or pulmonary stenosis (PS). Those with subaortic VSD present with minimal or no cyanosis, but may have pulmonary overcirculation and CHF. In infants with PS in addition to subaortic VSD, the physiology is similar to TOF. In DORV with subpulmonic VSD, deoxygenated venous return from RV is directed to the aorta, leading to cyanosis. The oxygenated LV output is directed through the VSD into the PA, leading to pulmonary overcirculation. This physiology resembles TGA. The surgical approach may consist of VSD closure, or TOF or TGA repair.

Anesthetic management: Is dictated by specific surgical repair. See sections on VSD, TOF, and TGA, for Anesthetic Considerations (p. 1212, 1223, 1233).

Suggested Readings

1. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Great artery anomalies. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents.* Williams & Wilkins, Philadelphia: 1998, 501–14.

2. Kanter KR: Double-outlet ventricles. In *Mastery of Cardiothoracic Surgery.* Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 771–84.

3. Kouchoukos NT, Blackstone EH, Doty DB et al. Double outlet right ventricle. In *Kirklin, Barrett, Boyes Cardiac Surgery.* Churchill Livingstone, Philadelphia: 2003, 1509–40.

Surgery for Hypoplastic Left Heart Syndrome

Surgical Considerations

Description: Hypoplastic left heart syndrome (HLHS) represents a spectrum of left-sided cardiac malformations centered around a markedly hypoplastic LV, and an atretic or hypoplastic aortic valve and ascending aorta; the mitral valve is also usually hypoplastic or atretic (Fig. 12.4-12). Consequently, the RV supports both the pulmonary and systemic circulations. The pulmonary venous return (PVR) enters the RA via an ASD, and the admixture of systemic and pulmonary venous return is delivered into the RV and main PA. Systemic flow is delivered into the aorta by way of a typically large PDA. Cyanosis, CHF, and systemic hypoperfusion result as PVR decreases and pulmonary blood flow increases at the expense of systemic blood flow.

Until the mid-to-late 1980s, patients born with this anomaly usually died within the first 2 wk of life, with survival beyond 6 wk being very rare. A palliative surgical treatment was reported by Norwood in 1983. This treatment of HLHS comprises staged operations, with the first stage designated as the **Norwood operation.** The procedure is directed toward establishing effective CO from the RV to the systemic circulation and to support the pulmonary circulation with a systemic-to-pulmonary arterial shunt. An adequate atrial septal communication is essential.

Stage I (Norwood): A midline sternotomy approach is used. The innominate artery (alternatively, the main PA or ascending aorta, if it is of adequate size) and RA appendage are cannulated and CPB with cooling is instituted. The arch vessels and proximal descending thoracic aorta are dissected. Deep hypothermia with low-flow CPB technique is used. During arch reconstruction, cerebral blood flow is maintained, although many surgeons prefer total circulatory arrest. The ductus arteriosus is then divided and the pulmonary end is oversewn. Next, the main PA is divided and the distal PA is oversewn. An aortotomy is created, extending from the interior aspect of the aortic arch through the lateral aspect of the ascending aorta to the level of the transected pulmonary trunk. The entire aortic arch complex is then augmented, creating a “neoaorta” from the proximal portion of the transected main PA, which is anastomosed to the ascending aorta and arch. A cryopreserved homograft patch is used to facilitate the repair. The RA is *(Print pagebreak 1246)* opened and the atrial septum is excised to maximize the size of the interatrial communication. The heart is de-aired and rewarming is started with full CPB. If circulatory arrest was used, the cannulae are reinserted, and rewarming begins. Finally, a B-T systemic-to-PA shunt is created to provide pulmonary blood flow. The patient is weaned from CPB, a pericardial drainage tube is placed, and the chest is closed in standard fashion.

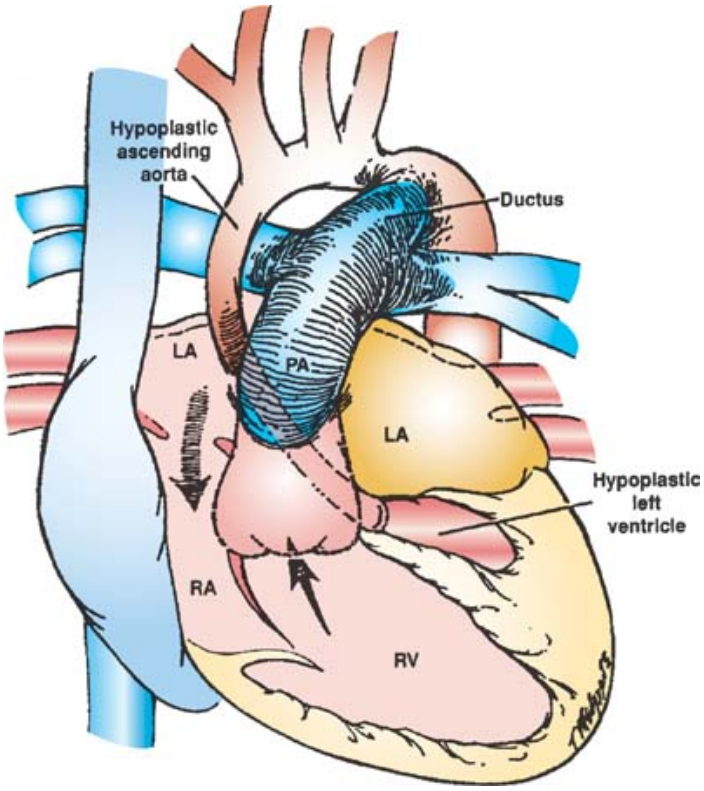

Figure 12.4-12. 12. The anatomic features of hypoplastic left heart syndrome. The right atrium (RA), right ventricle (RV), and main pulmonary artery (PA) are larger than in neonates with normal circulation. LA = left atrium. (Reproduced with permission from Kaiser LR, Kron IL, Spray TL, eds: *Mastery of Cardiothoracic Surgery* . Lippincott-Raven, Philadelphia: 1998.)

Currently, a modification of the Norwood procedure strongly advocated by Japanese surgeons is being adopted by many surgeons worldwide, including us. This involves abandoning the modified B-T shunt in favor of RV-PA shunt ± valve. This modification eliminates the diastolic run-off and preserves coronary blood flow. The major advantage is a stable early postop course. The long-term implications of right ventriculotomy in a systemic RV are not known at the present time.

Stage II (bidirectional Glenn procedure): As the PVR normally falls in the weeks after the first-stage operation, excessive pulmonary blood flow from the B-T shunt may → RV volume overload and CHF. The 2nd-stage procedure for HLHS is intended to reduce the volume load on the single ventricle, while maintaining adequate pulmonary blood flow. This operation consists of a bidirectional cavopulmonary (Glenn) shunt (see Surgery for Tricuspid Atresia, p. 1239), and usually is performed in children 4–6 mo old.

Stage III (Fontan procedure): The 3rd and final stages of the Norwood procedure usually are performed about 6–12 mo after the 2nd stage and consist of the completion of the Fontan procedure (see Surgery for Tricuspid Atresia, p. 1239). It is designed to divide the systemic venous return from the pulmonary venous return by establishing continuity from the IVC to the confluence of the PA and SVC.

Variant procedure or approaches: Successful cardiac transplantation for this condition was performed by **Bailey** in 1985. Cardiac transplantation is associated with a lower operative mortality; however, a significant number of neonates on the waiting lists do not receive donor hearts. Moreover, cardiac transplantation is associated with lifelong immunosuppression and its associated risks.

Usual preop diagnosis: HLHS; mitral and aortic atresia

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Summary of Procedures

Position	Supine
Incision	Standard median sternotomy
	Hypothermia (< 18° C) ± circulatory arrest; manipulation of

Unique considerations	PVR and SVR, both preop and postop, is extremely important, with the FiO_2 and ventilation being crucial in regulating pulmonary blood flow. For very high pulmonary blood flows and O_2 sats > 85%, consider the addition of N_2 to lower the FiO_2 to < 20%; use of afterload reduction to manipulate SVR and ↓ ventricular work.
Antibiotics	Cefazolin 25 mg/kg q 4 h
Surgical time	Aortic cross-clamp: 30–60 min Circulatory arrest (if used): 30–45 min Total: 2.5–3.5 h
Closing considerations	Sternum often left open, with chest tube in the pericardial space, with closure delayed until POD 2–4; temporary ventricular pacing wire; possible left atrial line for monitoring
EBL	Minimal-to-moderate
Postop care	2–7 h of controlled ventilation; moderate need for inotropic drugs; balancing PVR and SVR to optimize pulmonary blood flow and systemic CO.
Mortality	10–20%
Morbidity	Hemorrhage Infection ↓CO RV dysfunction Inadequate/excessive pulmonary blood flow Tricuspid regurgitation
Pain score	8–10

Patient Population Characteristics

Age range	5–30 d
Male:Female	2:1
Incidence	3–5% of congenital heart defects
Etiology	Often associated with genetic disorders, but no single associated defect
Associated conditions	CNS defects (29%); microencephaly (25–27%); VSD (5%); TGA

Anesthetic Considerations

Preoperative

Pathophysiology	HLHS is characterized by a hypoplastic or atretic aortic and mitral valves associated with hypoplasia of the LV, ascending aorta, and aortic arch. Pulmonary venous return enters the LA and then passes into the RA via an ASD or the PFO. Rarely, pulmonary venous return is via TAPVC to the RA. The systemic circulation is supplied entirely through the ductus arteriosus. Perfusion of the ascending aorta, coronary arteries, and transverse arch is retrograde via the ductus. Maintenance of adequate systemic perfusion relies on ductal patency and is achieved with continuous administration of PGE_1 Maintaining ↑ PVR by ↓ FiO_2 to 0.17–0.20, or by adding CO_2 to breathing gas mixture, may be necessary. As the ductus closes, the systemic perfusion is compromised, → ischemia, acidosis, and eventually death.

Cardiovascular	Survival is dependent on maintaining ductal patency, and adequacy of mixing at the atrial level and a balance between PVR and SVR to ensure adequate pulmonary and systemic perfusion. **Tests:** EKG; RAE/RVH; ECHO: assessment of tricuspid or common AV valve regurgitation. Severe TR is a contraindication for 1st stage repair.
Neurological	Preop head ultrasound to r/o intracranial pathology (e.g., hemorrhage). Periop neurological monitoring, including cerebral oximeter can be useful, either both hemispheres or left cerebral hemisphere and flank (somatic oxygenation)
Laboratory	BUN; Cr; Hb/Hct; Plt count
Premedication	Not indicated.
Transport to OR	See Transport of the Critically Ill Newborn (p. 1229).

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Intraoperative

Anesthetic technique: GETA

Induction	On arrival in the OR, monitors are applied and ventilation should be adjusted to maintain arterial O_2sat = 75–85%. Avoid hyperventilation (→↑pulmonary blood flow). Arterial sat > 85% will cause systemic hypoperfusion. See Truncus Arteriosus, p. 1238.	
Maintenance	Fentanyl (20–40 mcg/kg) with muscle relaxant (Rocuronium 1 mg/kg) in divided doses on CPB. Volatile anesthetics on CPB. Midazolam (0.1–0.2 mg/kg) as tolerated.	
Emergence	Transport to ICU intubated, ventilated, and monitored.	
Blood and fluid requirements	See Tetralogy of Fallot (TOF) (p. 1224). See Table 12.4-2, p. 1226, for blood product utilization.	Blood loss can be due largely to dilutional coagulopathy and multiple suture lines.
Infusions	See Truncus Arteriosus.	
Monitoring	See Truncus Arteriosus (p. 1238).	
CPB	See TGA (p. 1235).	
Transition off CPB and Post-CPB	See TOF (p. 1225).	After Stage I (Norwood) surgical repair, the blood flow continues to be supplied in a parallel fashion from the single ventricle. At Stanford, a small homograft conduit is placed from the RV to PA (Kashimoto modification) instead of the traditional modified B-T shunt. This prevents the diastolic run-off of the B-T shunt and improves coronary perfusion. Optimal PaO_2 30–50 mmHg with MAP 40–50 mmHg.

Postoperative

Complications	Bleeding Pulmonary overcirculation Myocardial dysfunction and failure Aortic arch obstruction Pulmonary HTN CNS injury

Suggested Readings

1. Allen HD, Clark EB, Gutgesell HP, et al eds. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents*, 6th edition. Lippincott Williams & Wilkins, Philadelphia: 2001.

2. Emmanouilides GC, Allen HD, Riemenschneider TA, et al: Left ventricular outflow abnormalities. In *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents*. Williams & Wilkins, Philadelphia: 1998, 501–14.

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3. Jacobs ML, Norwood WI: Hyperplastic left heart syndrome. In *Pediatric Cardiac Surgery: Current Issues*. Jacobs ML, Norwood WI, eds. Butterworth-Heinemann, Stoneham: 1992, 182–92.

4. Jacobs ML, Norwood WI: Hypoplastic left heart syndrome. In *Glenn's Thoracic and Cardiovascular Surgery*, 6th edition. Baue AE, ed. Appleton & Lange, Stamford: 1996, 1271–81.

5. Jacobs ML: Hypoplastic left heart syndrome. In *Mastery of Cardiothoracic Surgery*. Kaiser LR, Kron IL, Spray TL, eds. Lippincott-Raven, Philadelphia: 1998, 858–66.

6. Kirkham FJ: Recognition and prevention of neurological complications in pediatric cardiac surgery. *Pediatr Cardiol* 1998; 19:331–45.

7. Kouchoukos NT, Blackstone EH, Doty DB, et al: Aortic atresia and other forms of hypoplastic left heart physiology. In *Kirklin, Barrett, Boyes Cardiac Surgery*. Churchill Livingstone, Philadelphia: 2003, 1377–1400.

8. Nicolson SC, Steven JM, Jobes DR: Hypoplastic left heart syndrome. In *Pediatric Cardiac Anesthesia,* 3rd edition. Lake CL, ed. Appleton & Lange, Stamford: 1998, 337–52.

9. Ramamoorthy C, Tabbutt S, Kurth CD, et al: Effects of inspired hypoxic and hypercapnic gas mixtures on cerebral oxygen saturation in neonates with univentricular heart defects. *Anesthesiology* 2002; 96:283–8.

10. Reitz BA, Yuh DD, eds: *Congenital Cardiac Surgery*. McGraw-Hill, New York: 2002.

11. Sano S, Ishino K, Kado H, et al. Outcome of right ventricle to pulmonary artery shunt in first stage palliation of hypoplastic left heart syndrome–a multi-institutional study. *Ann Thorac Surg* 2004; 78(6):1951–7.

12. Tabbutt S, Ramamoorthy C, Montenegro LM, et al: Impact of inspired gas mixtures on preoperative infants with hypoplastic left heart syndrome during controlled ventilation. *Circulation* 2001; 104(Supp I):I159–64.